WEBVTT

NOTE duration:"01:03:17"

NOTE recognizability:0.865

NOTE language:en-us

NOTE Confidence: 0.8215359625

00:00:03.950 --> 00:00:07.911 To see you again. So.

NOTE Confidence: 0.8215359625

00:00:07.911 --> 00:00:12.066 There are already 52 participants,

NOTE Confidence: 0.8215359625

00:00:12.070 --> 00:00:14.908 so I'm going to go ahead

NOTE Confidence: 0.8215359625

00:00:14.908 --> 00:00:16.800 and start introducing you.

NOTE Confidence: 0.8215359625

00:00:16.800 --> 00:00:21.528 Uhm? So welcome everyone for

NOTE Confidence: 0.8215359625

00:00:21.528 --> 00:00:24.096 the pathology grand rounds.

NOTE Confidence: 0.8215359625

00:00:24.100 --> 00:00:27.940 Today's speaker is doctor Jason Hornick.

NOTE Confidence: 0.8215359625

00:00:27.940 --> 00:00:31.480 He's a professor of pathology at

NOTE Confidence: 0.8215359625

00:00:31.480 --> 00:00:35.020 Brigham and Women's Hospital in Boston,

NOTE Confidence: 0.8215359625

00:00:35.020 --> 00:00:38.068 or Doctor Hornick grew up in

NOTE Confidence: 0.8215359625

00:00:38.068 --> 00:00:42.805 California and then came up north to

NOTE Confidence: 0.8215359625

00:00:42.805 --> 00:00:46.915 do his college at Amherst College.

NOTE Confidence: 0.8215359625

00:00:46.920 --> 00:00:49.566 He went back to do MD PhD.

NOTE Confidence: 0.8215359625

00:00:49.570 --> 00:00:50.508 In California,

NOTE Confidence: 0.8215359625

00:00:50.508 --> 00:00:53.791 and then he must have missed the

NOTE Confidence: 0.8215359625

00:00:53.791 --> 00:00:56.126 seasons because he came right

NOTE Confidence: 0.8215359625

00:00:56.126 --> 00:00:59.960 back to Boston where he did his

NOTE Confidence: 0.8215359625

00:00:59.960 --> 00:01:04.880 residency and stayed on and his.

NOTE Confidence: 0.8215359625

00:01:04.880 --> 00:01:08.779 He completed his entire residency at Brigham.

NOTE Confidence: 0.8215359625

00:01:08.780 --> 00:01:11.180 He knew exactly what he wanted.

NOTE Confidence: 0.8215359625

00:01:11.180 --> 00:01:12.282 He did,

NOTE Confidence: 0.8215359625

00:01:12.282 --> 00:01:16.139 and AP only residency and a fellowship,

NOTE Confidence: 0.8215359625

00:01:16.140 --> 00:01:19.828 which was three months each in soft tissue.

NOTE Confidence: 0.8215359625

00:01:19.830 --> 00:01:21.070 Him path,

NOTE Confidence: 0.8215359625

00:01:21.070 --> 00:01:23.550 jiwan pathology and general

NOTE Confidence: 0.8215359625

00:01:23.550 --> 00:01:27.039 pathology and this might seem mad.

NOTE Confidence: 0.8215359625

00:01:27.040 --> 00:01:30.166 But all of this exposure shaped

NOTE Confidence: 0.8215359625

00:01:30.166 --> 00:01:32.194 his academic career anyway.

NOTE Confidence: 0.8215359625

00:01:32.194 --> 00:01:34.978 He rounded it off with one.

NOTE Confidence: 0.8215359625

00:01:34.980 --> 00:01:38.868 Year of GI pathology.

NOTE Confidence: 0.8215359625

00:01:38.870 --> 00:01:42.678 I'm not sure how that helped his career.

NOTE Confidence: 0.8215359625

00:01:42.680 --> 00:01:45.602 His doctor Hartnick made his name

NOTE Confidence: 0.8215359625

00:01:45.602 --> 00:01:48.820 as a soft tissue pathologist.

NOTE Confidence: 0.8215359625

00:01:48.820 --> 00:01:51.622 He is an outstanding educator and

NOTE Confidence: 0.8215359625

00:01:51.622 --> 00:01:54.933 a frequent speaker at national and

NOTE Confidence: 0.8215359625

00:01:54.933 --> 00:01:57.649 international meetings and courses.

NOTE Confidence: 0.8215359625

00:01:57.650 --> 00:02:02.216 He is codirector of surgical pathology

NOTE Confidence: 0.8215359625

00:02:02.216 --> 00:02:06.591 update course at Brigham and Women's

NOTE Confidence: 0.8215359625

00:02:06.591 --> 00:02:10.076 and of the diagnostic pathology

NOTE Confidence: 0.8215359625

00:02:10.076 --> 00:02:13.189 update course for the US CAP.

NOTE Confidence: 0.8215359625

00:02:13.190 --> 00:02:16.110 Doctor Hornik is very generous of his time.

NOTE Confidence: 0.8215359625

00:02:16.110 --> 00:02:18.210 He wears many hats,

NOTE Confidence: 0.8215359625

00:02:18.210 --> 00:02:20.310 director of Surg Path,

NOTE Confidence: 0.8215359625

00:02:20.310 --> 00:02:23.565 medical director of Histology and

NOTE Confidence: 0.8215359625

00:02:23.565 --> 00:02:25.518 the immunohistochemistry lab.

NOTE Confidence: 0.8215359625

00:02:25.520 --> 00:02:30.108 His cheer of the QA committee and his

NOTE Confidence: 0.8215359625

00:02:30.108 --> 00:02:33.432 also on several cancer focused committees

NOTE Confidence: 0.8215359625

00:02:33.432 --> 00:02:37.519 at the Brigham and Women's Hospital.

NOTE Confidence: 0.8215359625

00:02:37.520 --> 00:02:38.674 He nationally,

NOTE Confidence: 0.8215359625

00:02:38.674 --> 00:02:42.713 he served on the education and committee.

NOTE Confidence: 0.8215359625

00:02:42.720 --> 00:02:45.611 All of us cap and is currently

NOTE Confidence: 0.8215359625

00:02:45.611 --> 00:02:48.200 on the Board of Directors.

NOTE Confidence: 0.8215359625

00:02:48.200 --> 00:02:51.966 He's been a panelist and moderator on

NOTE Confidence: 0.8215359625

00:02:51.966 --> 00:02:55.860 several of the educational sessions.

NOTE Confidence: 0.8215359625

00:02:55.860 --> 00:02:58.710 His written guidelines, his chair,

NOTE Confidence: 0.8215359625

00:02:58.710 --> 00:03:01.122 dicey committee for CAP,

NOTE Confidence: 0.8215359625

00:03:01.122 --> 00:03:04.137 and he developed the proficiency

NOTE Confidence: 0.8215359625

00:03:04.137 --> 00:03:06.928 testing for thousands of IFC

NOTE Confidence: 0.8215359625

00:03:06.928 --> 00:03:09.016 labs around the country,

NOTE Confidence: 0.8215359625

00:03:09.020 --> 00:03:12.278 our lab being one of those.

NOTE Confidence: 0.8215359625

00:03:12.280 --> 00:03:16.306 He is involved with the agencies

NOTE Confidence: 0.8215359625

00:03:16.306 --> 00:03:20.334 sarcoma staging and the cancer genome.

NOTE Confidence: 0.8215359625

00:03:20.334 --> 00:03:25.170 Atlas is a sarcoma analysis working group.

NOTE Confidence: 0.8215359625

00:03:25.170 --> 00:03:28.434 He also finds time to write board questions

NOTE Confidence: 0.8215359625

00:03:28.434 --> 00:03:31.570 for the American Board of Pathology.

NOTE Confidence: 0.8215359625

00:03:31.570 --> 00:03:32.182 Uhm?

NOTE Confidence: 0.8215359625

00:03:32.182 --> 00:03:35.242 Doctor Hornick has developed and

NOTE Confidence: 0.8215359625

00:03:35.242 --> 00:03:38.647 described many IHC markers that

NOTE Confidence: 0.8215359625

00:03:38.647 --> 00:03:41.227 diagnose genetic abnormalities,

NOTE Confidence: 0.8215359625

00:03:41.230 --> 00:03:44.590 and many of these are actually

NOTE Confidence: 0.8215359625

00:03:44.590 --> 00:03:46.830 have affected our practice,

NOTE Confidence: 0.8215359625

00:03:46.830 --> 00:03:50.850 so these are significant practice changers.

NOTE Confidence: 0.8215359625

00:03:50.850 --> 00:03:54.441 Some of a short list would be I and

NOTE Confidence: 0.8215359625

00:03:54.441 --> 00:03:58.380 I won that detects smart B1 genetic

NOTE Confidence: 0.8215359625

00:03:58.380 --> 00:04:02.760 aberrations start six that detects the

NOTE Confidence: 0.8215359625

00:04:02.760 --> 00:04:06.580 translocation in solitary fibrous tumors.

NOTE Confidence: 0.8215359625

00:04:06.580 --> 00:04:09.720 Back story for sinonasal sarcomas.

NOTE Confidence: 0.8215359625

00:04:09.720 --> 00:04:12.008 Panther Gabino history chemistry,

NOTE Confidence: 0.8215359625

00:04:12.008 --> 00:04:16.729 which we use on head and neck tumors.

NOTE Confidence: 0.8215359625

00:04:16.730 --> 00:04:20.769 This 18 it says X fusion specific

NOTE Confidence: 0.8215359625

00:04:20.769 --> 00:04:23.170 antibody for synovial sarcomas

NOTE Confidence: 0.8215359625

00:04:23.170 --> 00:04:26.530 that he talked about and showed

NOTE Confidence: 0.8215359625

00:04:26.530 --> 00:04:28.786 in this morning's conference,

NOTE Confidence: 0.8215359625

00:04:28.786 --> 00:04:31.686 and many, many, many others.

NOTE Confidence: 0.8215359625

00:04:31.686 --> 00:04:34.078 The list is long,

NOTE Confidence: 0.8215359625

00:04:34.080 --> 00:04:36.906 he's he's the editor in chief

NOTE Confidence: 0.8215359625

00:04:36.906 --> 00:04:39.848 of the fifth series of if IPS,

NOTE Confidence: 0.8215359625

00:04:39.850 --> 00:04:44.638 atlases of tumor, and non tumor pathology.

NOTE Confidence: 0.8215359625

00:04:44.640 --> 00:04:46.840 He is an associate editor.

NOTE Confidence: 0.8215359625

00:04:46.840 --> 00:04:49.408 For Sternberg's diagnostic

NOTE Confidence: 0.8215359625

00:04:49.408 --> 00:04:51.120 surgical pathology,

NOTE Confidence: 0.8215359625

00:04:51.120 --> 00:04:54.090 he is on the editorial board

NOTE Confidence: 0.8215359625

00:04:54.090 --> 00:04:56.070 of a JSP and

NOTE Confidence: 0.638818059166666

00:04:56.196 --> 00:04:58.250 modern pathology.

NOTE Confidence: 0.638818059166666

00:04:58.250 --> 00:05:02.066 He is he has a senior leadership position

NOTE Confidence: 0.638818059166666

00:05:02.066 --> 00:05:06.400 on the editorial boards of histopathology

NOTE Confidence: 0.638818059166666

00:05:06.400 --> 00:05:09.680 advances in anatomic pathology,

NOTE Confidence: 0.638818059166666

00:05:09.680 --> 00:05:11.849 surgical pathology, clinics,

NOTE Confidence: 0.638818059166666

00:05:11.849 --> 00:05:18.180 archives of pathology and lab medicine, and.

NOTE Confidence: 0.638818059166666

00:05:18.180 --> 00:05:19.170 Not surprisingly,

NOTE Confidence: 0.638818059166666

00:05:19.170 --> 00:05:23.799 he has more than 400 original articles,

NOTE Confidence: 0.638818059166666

00:05:23.799 --> 00:05:26.448 reviews and chapters,

NOTE Confidence: 0.638818059166666

00:05:26.450 --> 00:05:29.922 and harking back to his first fellowship

NOTE Confidence: 0.638818059166666

00:05:29.922 --> 00:05:33.109 were his spent three months in

NOTE Confidence: 0.638818059166666

00:05:33.109 --> 00:05:35.774 various subspecialities his not only

NOTE Confidence: 0.638818059166666

00:05:35.774 --> 00:05:39.411 on the editorial board of these soft

NOTE Confidence: 0.638818059166666

00:05:39.411 --> 00:05:42.844 tissue and bone blue book by WHO,

NOTE Confidence: 0.638818059166666

00:05:42.844 --> 00:05:45.676 but also on the blue books.

NOTE Confidence: 0.638818059166666

00:05:45.680 --> 00:05:47.684 He's contributed to breast.

NOTE Confidence: 0.638818059166666

00:05:47.684 --> 00:05:50.690 Do you want him to rasik

NOTE Confidence: 0.638818059166666

00:05:50.787 --> 00:05:53.108 and pediatric Bluebirds?

NOTE Confidence: 0.638818059166666

00:05:53.108 --> 00:05:57.380 So in conclusion, Dr.

NOTE Confidence: 0.638818059166666

00:05:57.380 --> 00:06:01.450 Hornig still has a lot of time on his hands,

NOTE Confidence: 0.638818059166666

00:06:01.450 --> 00:06:03.844 and I'll show what else he does

NOTE Confidence: 0.638818059166666

00:06:03.850 --> 00:06:07.420 by sharing my screen here.

NOTE Confidence: 0.638818059166666

00:06:07.420 --> 00:06:08.911 Recently this year,

NOTE Confidence: 0.638818059166666

00:06:08.911 --> 00:06:12.390 Doctor Hornick with his here on the

NOTE Confidence: 0.638818059166666

00:06:12.486 --> 00:06:15.461 keyboard has cut an album for his

NOTE Confidence: 0.638818059166666

00:06:15.461 --> 00:06:18.795 band with his band up there teardowns.

NOTE Confidence: 0.638818059166666

00:06:18.795 --> 00:06:22.005 And this is available on Spotify.

NOTE Confidence: 0.638818059166666

00:06:22.010 --> 00:06:25.718 If anyone cares if anyone is

NOTE Confidence: 0.638818059166666

00:06:25.718 --> 00:06:28.190 tired of learning pathology,

NOTE Confidence: 0.638818059166666

00:06:28.190 --> 00:06:30.500 we can all go back and listen

NOTE Confidence: 0.638818059166666

00:06:30.500 --> 00:06:32.220 to his band instead.

NOTE Confidence: 0.638818059166666

00:06:32.220 --> 00:06:36.486 So with that short introduction I will.

NOTE Confidence: 0.638818059166666

00:06:36.486 --> 00:06:39.534 See the floor to Doctor Hornick.

NOTE Confidence: 0.87832498875

00:06:40.820 --> 00:06:43.120 Thank you very much Montreux

NOTE Confidence: 0.87832498875

00:06:43.120 --> 00:06:44.977 for that Nice introduction.

NOTE Confidence: 0.87832498875

00:06:44.977 --> 00:06:47.839 Let me make sure this works.

NOTE Confidence: 0.87832498875

00:06:47.840 --> 00:06:49.400 I always have a little bit of trouble.

NOTE Confidence: 0.933226253333333

00:06:51.760 --> 00:06:53.643 Can you let me know if you

NOTE Confidence: 0.933226253333333

00:06:53.643 --> 00:06:55.400 can see my full screen?

NOTE Confidence: 0.933226253333333

00:06:55.400 --> 00:07:01.454 Yep. We can. Is it in the

NOTE Confidence: 0.933226253333333

00:07:01.454 --> 00:07:02.734 presentation mode or full screen?

NOTE Confidence: 0.877260148333333

00:07:02.750 --> 00:07:08.370 Yes, it's in the presentation mode. No notes.

NOTE Confidence: 0.857040776

00:07:10.310 --> 00:07:12.310 Oops was that good?

NOTE Confidence: 0.857040776

00:07:12.310 --> 00:07:14.330 Yeah, that's the full screen mode.

NOTE Confidence: 0.857040776

00:07:14.330 --> 00:07:17.868 OK, perfect. So as you heard,

NOTE Confidence: 0.857040776

00:07:17.868 --> 00:07:19.653 I have very diverse interests.

NOTE Confidence: 0.857040776

00:07:19.660 --> 00:07:21.072 My main area of.

NOTE Confidence: 0.857040776

00:07:21.072 --> 00:07:24.172 Interest where I do most of my research

NOTE Confidence: 0.857040776

00:07:24.172 --> 00:07:26.878 is in soft tissue tumor pathology

NOTE Confidence: 0.857040776

00:07:26.880 --> 00:07:30.132 and Doctor Prasad has invited me

NOTE Confidence: 0.857040776

00:07:30.132 --> 00:07:32.312 today to give a lecture focusing on

NOTE Confidence: 0.857040776

00:07:32.312 --> 00:07:34.200 soft tissue tumors of the head neck.

NOTE Confidence: 0.857040776

00:07:34.200 --> 00:07:36.979 This is a a complicated area obviously

NOTE Confidence: 0.857040776

00:07:36.979 --> 00:07:39.729 not only for soft tissue tumors,

NOTE Confidence: 0.857040776

00:07:39.730 --> 00:07:41.906 but this is a an area with lots

NOTE Confidence: 0.857040776

00:07:41.906 --> 00:07:44.151 of vital structures and a diverse

NOTE Confidence: 0.857040776

00:07:44.151 --> 00:07:46.191 array of different tumor types

NOTE Confidence: 0.857040776

00:07:46.191 --> 00:07:48.555 that can arise in these structures.

NOTE Confidence: 0.857040776

00:07:48.555 --> 00:07:52.109 And I'm going to talk about some really

NOTE Confidence: 0.857040776

00:07:52.109 --> 00:07:55.049 dramatic changes that have come to

NOTE Confidence: 0.857040776

00:07:55.049 --> 00:07:57.467 head neck pathology for mesenchymal

NOTE Confidence: 0.857040776

00:07:57.467 --> 00:08:00.931 neoplasia in the last ten years or so.

NOTE Confidence: 0.857040776

00:08:00.940 --> 00:08:04.080 I have the following disclosures.

NOTE Confidence: 0.857040776

00:08:04.080 --> 00:08:06.612 I'm a consultant to Addie Biosciences

NOTE Confidence: 0.857040776

00:08:06.612 --> 00:08:08.300 and Trey Con Pharmaceuticals,

NOTE Confidence: 0.857040776

00:08:08.300 --> 00:08:09.975 which is not relevant to

NOTE Confidence: 0.857040776

00:08:09.975 --> 00:08:12.650 the discussion for today.

NOTE Confidence: 0.857040776

00:08:12.650 --> 00:08:15.086 I'm going to be framing my

NOTE Confidence: 0.857040776

00:08:15.086 --> 00:08:17.270 presentation in terms of cases.

NOTE Confidence: 0.857040776

00:08:17.270 --> 00:08:19.713 I think it's nice to bring them

NOTE Confidence: 0.857040776

00:08:19.713 --> 00:08:21.760 home to practical applications,

NOTE Confidence: 0.857040776

00:08:21.760 --> 00:08:24.175 and many of the findings that I'm

NOTE Confidence: 0.857040776

00:08:24.175 --> 00:08:26.791 going to be discussing are nicely

NOTE Confidence: 0.857040776

00:08:26.791 --> 00:08:29.286 illustrated by by these cases.

NOTE Confidence: 0.857040776

00:08:29.290 --> 00:08:30.816 I'm going to start off with a

NOTE Confidence: 0.857040776

00:08:30.816 --> 00:08:32.540 with a tumor type that we've known

NOTE Confidence: 0.857040776

00:08:32.540 --> 00:08:34.641 about for a long time, but it's a.

NOTE Confidence: 0.857040776

00:08:34.641 --> 00:08:36.303 It's a nice framing for the

NOTE Confidence: 0.857040776

00:08:36.303 --> 00:08:37.780 rest of the discussion,

NOTE Confidence: 0.857040776

00:08:37.780 --> 00:08:40.468 because this is a tumor that is often

NOTE Confidence: 0.857040776

00:08:40.468 --> 00:08:42.817 overlooked and is prone to misdiagnosis.

NOTE Confidence: 0.857040776

00:08:42.820 --> 00:08:44.344 At this anatomic site.

NOTE Confidence: 0.857040776

00:08:44.344 --> 00:08:47.352 So we're starting off with a 43 year

NOTE Confidence: 0.857040776

00:08:47.352 --> 00:08:49.613 old man with a left sinus mass.

NOTE Confidence: 0.857040776

00:08:49.620 --> 00:08:51.180 As is often the case,

NOTE Confidence: 0.857040776

00:08:51.180 --> 00:08:52.050 these specimens,

NOTE Confidence: 0.857040776

00:08:52.050 --> 00:08:55.133 these two readings often look kind of messy.

NOTE Confidence: 0.857040776

00:08:55.133 --> 00:08:56.638 There's a lot of hemorrhage,

NOTE Confidence: 0.857040776

00:08:56.640 --> 00:08:59.265 and lots of separate fragments of tissue,

NOTE Confidence: 0.857040776

00:08:59.270 --> 00:09:00.350 and on the low power,

NOTE Confidence: 0.857040776

00:09:00.350 --> 00:09:01.943 it's kind of hard to see what's going on,

NOTE Confidence: 0.857040776

00:09:01.950 --> 00:09:04.494 although there are some sailor foci

NOTE Confidence: 0.857040776

00:09:04.494 --> 00:09:06.766 in the scanning image and let's

NOTE Confidence: 0.857040776

00:09:06.766 --> 00:09:08.782 focus on a few of these areas.

NOTE Confidence: 0.857040776

00:09:08.790 --> 00:09:10.995 So now we see those one fragmente

NOTE Confidence: 0.857040776

00:09:10.995 --> 00:09:12.592 with some blue cells.

NOTE Confidence: 0.857040776

00:09:12.592 --> 00:09:15.478 Hard to know what's going on.

NOTE Confidence: 0.857040776

00:09:15.480 --> 00:09:16.964 Higher power another area.

NOTE Confidence: 0.857040776

00:09:16.964 --> 00:09:19.990 We also have these streams of blue cells,

NOTE Confidence: 0.857040776

00:09:19.990 --> 00:09:22.486 but certainly to make anything more

NOTE Confidence: 0.857040776

00:09:22.486 --> 00:09:25.249 of this will have to go closer.

NOTE Confidence: 0.857040776

00:09:25.250 --> 00:09:27.210 So now you begin to see that there

NOTE Confidence: 0.857040776

00:09:27.210 --> 00:09:29.029 are these very large, rounded,

NOTE Confidence: 0.857040776

00:09:29.029 --> 00:09:31.665 slightly angulated cells embedded

NOTE Confidence: 0.857040776

00:09:31.665 --> 00:09:34.960 in this dense fibrous stroma.

NOTE Confidence: 0.857040776

00:09:34.960 --> 00:09:37.016 Even a higher power we can see this

NOTE Confidence: 0.857040776

00:09:37.016 --> 00:09:39.120 as a somewhat nested proliferation,

NOTE Confidence: 0.857040776

00:09:39.120 --> 00:09:41.360 infiltrating through the submucosal.

NOTE Confidence: 0.857040776

00:09:41.360 --> 00:09:44.048 We have some normal,

NOTE Confidence: 0.857040776

00:09:44.048 --> 00:09:46.736 normal glandular structures here.

NOTE Confidence: 0.857040776

00:09:46.740 --> 00:09:47.416 Higher power,

NOTE Confidence: 0.857040776

00:09:47.416 --> 00:09:50.593 this is a tumor that has significant

NOTE Confidence: 0.857040776

00:09:50.593 --> 00:09:52.879 crush artifact.

NOTE Confidence: 0.857040776

00:09:52.880 --> 00:09:54.740 Just the last few hypower images

NOTE Confidence: 0.857040776

00:09:54.740 --> 00:09:57.046 and you can see this is a small

NOTE Confidence: 0.857040776

00:09:57.046 --> 00:09:58.138 round blue cell tumor.

NOTE Confidence: 0.857040776

00:09:58.140 --> 00:09:59.616 Although the cells aren't so small,

NOTE Confidence: 0.857040776

00:09:59.620 --> 00:10:00.924 they're actually quite large.

NOTE Confidence: 0.857040776

00:10:00.924 --> 00:10:03.602 Then we come to really high power and

NOTE Confidence: 0.857040776

00:10:03.602 --> 00:10:05.576 you can see the primitive chromatin.

NOTE Confidence: 0.857040776

00:10:05.580 --> 00:10:07.908 Very large nuclei,

NOTE Confidence: 0.857040776

00:10:07.908 --> 00:10:10.236 very limited cytoplasm.

NOTE Confidence: 0.857040776

00:10:10.240 --> 00:10:12.694 So I think on morphologic grounds

NOTE Confidence: 0.857040776

00:10:12.694 --> 00:10:13.921 the differential diagnosis

NOTE Confidence: 0.857040776

00:10:13.921 --> 00:10:15.579 would certainly be broad.

NOTE Confidence: 0.857040776

00:10:15.580 --> 00:10:17.312 This is a young,

NOTE Confidence: 0.857040776

00:10:17.312 --> 00:10:19.477 not quite middle aged adults,

NOTE Confidence: 0.857040776

00:10:19.480 --> 00:10:21.559 so we have to think of sarcomas,

NOTE Confidence: 0.857040776

00:10:21.560 --> 00:10:22.235 lymphoma,

NOTE Confidence: 0.857040776

00:10:22.235 --> 00:10:22.910 Melanoma,

NOTE Confidence: 0.857040776

00:10:22.910 --> 00:10:25.610 sometimes sinonasal melanomas have

NOTE Confidence: 0.857040776

00:10:25.610 --> 00:10:28.534 round some morphology or given

NOTE Confidence: 0.857040776

00:10:28.534 --> 00:10:30.322 the the high nucleocytoplasmic

NOTE Confidence: 0.857040776

00:10:30.322 --> 00:10:32.110 ratio and the print

NOTE Confidence: 0.860127200769231

00:10:32.190 --> 00:10:34.008 of primitive appearance.

NOTE Confidence: 0.860127200769231

00:10:34.010 --> 00:10:36.174 We certainly could also

NOTE Confidence: 0.860127200769231

00:10:36.174 --> 00:10:37.797 consider endocrine carcinoma,

NOTE Confidence: 0.860127200769231

00:10:37.800 --> 00:10:40.558 a very wide panel of screening markers.

NOTE Confidence: 0.860127200769231

00:10:40.560 --> 00:10:43.345 Was done in this case and the first

NOTE Confidence: 0.860127200769231

00:10:43.345 --> 00:10:45.595 pass only revealed a single finding

NOTE Confidence: 0.860127200769231

00:10:45.595 --> 00:10:48.340 and that is very strong intense

NOTE Confidence: 0.860127200769231

00:10:48.340 --> 00:10:51.940 standing for Desmond in every cell.

NOTE Confidence: 0.860127200769231

00:10:51.940 --> 00:10:54.556 You can see on high power the limited

NOTE Confidence: 0.860127200769231

00:10:54.556 --> 00:10:57.889 silo plasm is nicely highlighted by the

NOTE Confidence: 0.860127200769231

00:10:57.889 --> 00:10:59.977 Desmond Stainless intermediate filament.

NOTE Confidence: 0.860127200769231

00:10:59.980 --> 00:11:01.678 Then additional confirmatory

NOTE Confidence: 0.860127200769231

00:11:01.678 --> 00:11:03.376 marker was performed.

NOTE Confidence: 0.860127200769231

00:11:03.380 --> 00:11:06.020 Actually, 2 markers MYLDE 1,

NOTE Confidence: 0.860127200769231

00:11:06.020 --> 00:11:08.510 which is positive in every nucleus,

NOTE Confidence: 0.860127200769231

00:11:08.510 --> 00:11:11.758 as is myogenin in a very strong,

NOTE Confidence: 0.860127200769231

00:11:11.760 --> 00:11:12.406 intense fashion.

NOTE Confidence: 0.860127200769231

00:11:12.406 --> 00:11:15.503 And I'm sure by now many of you in

NOTE Confidence: 0.860127200769231

00:11:15.503 --> 00:11:18.155 surgical pathology have made the diagnosis.

NOTE Confidence: 0.860127200769231

00:11:18.160 --> 00:11:20.920 This summarizes the immunity chemistry,

NOTE Confidence: 0.860127200769231

00:11:20.920 --> 00:11:22.450 all the markers that were negative,

NOTE Confidence: 0.860127200769231

00:11:22.450 --> 00:11:24.978 excluding your endocrine carcinoma,

NOTE Confidence: 0.860127200769231

00:11:24.978 --> 00:11:26.916 Melanoma, and lymphoma.

NOTE Confidence: 0.860127200769231

00:11:26.916 --> 00:11:29.528 This is alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.953476476666667

00:11:31.640 --> 00:11:33.320 I don't expect you to

NOTE Confidence: 0.953476476666667

00:11:33.320 --> 00:11:34.664 follow this entire table.

NOTE Confidence: 0.953476476666667

00:11:34.670 --> 00:11:37.210 This table was included in a review

NOTE Confidence: 0.953476476666667

00:11:37.210 --> 00:11:39.800 article that I I recently wrote for

NOTE Confidence: 0.953476476666667

00:11:39.800 --> 00:11:42.620 seminars and diagnostic pathology with a

NOTE Confidence: 0.953476476666667

00:11:42.620 --> 00:11:44.652 junior colleague of mine, Michael Kaelin,

NOTE Confidence: 0.953476476666667

00:11:44.652 --> 00:11:47.144 who's a bone and soft tissue pathologist

NOTE Confidence: 0.953476476666667

00:11:47.144 --> 00:11:49.200 at the University of Maryland,

NOTE Confidence: 0.953476476666667

00:11:49.200 --> 00:11:52.119 where we focused on these new emerging

NOTE Confidence: 0.953476476666667

00:11:52.119 --> 00:11:54.459 categories of round soul sarcomas.

NOTE Confidence: 0.953476476666667

00:11:54.460 --> 00:11:57.660 But we also included this table that really

NOTE Confidence: 0.953476476666667

00:11:57.660 --> 00:11:59.750 highlights some of the conventional lineages,

NOTE Confidence: 0.953476476666667

00:11:59.750 --> 00:12:01.445 markers keratin Desmond.

NOTE Confidence: 0.953476476666667

00:12:01.445 --> 00:12:05.400 Myogenin Myo D1 that we've already mentioned,

NOTE Confidence: 0.953476476666667

00:12:05.400 --> 00:12:06.620 as well as some more,

NOTE Confidence: 0.953476476666667

00:12:06.620 --> 00:12:09.848 much more recently developed markers that

NOTE Confidence: 0.953476476666667

00:12:09.848 --> 00:12:12.950 correlate with molecular genetic alterations,

NOTE Confidence: 0.953476476666667

00:12:12.950 --> 00:12:15.530 which in some cases are really

NOTE Confidence: 0.953476476666667

00:12:15.530 --> 00:12:18.267 sufficient to make the diagnosis of

NOTE Confidence: 0.953476476666667

00:12:18.267 --> 00:12:21.543 some of these round cell sarcoma types.

NOTE Confidence: 0.953476476666667

00:12:21.550 --> 00:12:24.085 So let's talk about Rhabdomyosarcoma

NOTE Confidence: 0.953476476666667

00:12:24.085 --> 00:12:25.099 in general.

NOTE Confidence: 0.953476476666667

00:12:25.100 --> 00:12:27.020 There are four different classes of

NOTE Confidence: 0.953476476666667

00:12:27.020 --> 00:12:29.280 tumors that we consider Rhabdomyosarcoma,

NOTE Confidence: 0.953476476666667

00:12:29.280 --> 00:12:32.948 as these are all sarcomas that shows

NOTE Confidence: 0.953476476666667

00:12:32.948 --> 00:12:34.520 skeletal muscle differentiation.

NOTE Confidence: 0.953476476666667

00:12:34.520 --> 00:12:37.490 The classic embryonal and alveolar

NOTE Confidence: 0.953476476666667

00:12:37.490 --> 00:12:39.272 Rhabdomyosarcoma is typically

NOTE Confidence: 0.953476476666667

00:12:39.272 --> 00:12:40.923 affect children, adolescents,

NOTE Confidence: 0.953476476666667

00:12:40.923 --> 00:12:42.312 and young adults,

NOTE Confidence: 0.953476476666667

00:12:42.312 --> 00:12:45.090 and they do have particular anatomic

NOTE Confidence: 0.953476476666667

00:12:45.165 --> 00:12:47.310 sites where these tumors arise,

NOTE Confidence: 0.953476476666667

00:12:47.310 --> 00:12:49.464 and if you're familiar with the

NOTE Confidence: 0.953476476666667

00:12:49.464 --> 00:12:51.549 epidemiology of these classes of tumors,

NOTE Confidence: 0.953476476666667

00:12:51.550 --> 00:12:55.366 it is often possible to suggest the diagnosis

NOTE Confidence: 0.953476476666667

00:12:55.370 --> 00:12:57.477 even before you look down the microscope,

NOTE Confidence: 0.953476476666667

00:12:57.480 --> 00:13:00.145 and I always emphasize the

NOTE Confidence: 0.953476476666667

00:13:00.145 --> 00:13:02.277 demographics is critical and

NOTE Confidence: 0.953476476666667

00:13:02.277 --> 00:13:04.748 understanding some basic epidemiology.

NOTE Confidence: 0.953476476666667

00:13:04.750 --> 00:13:07.606 Of sarcomas can really help you

NOTE Confidence: 0.953476476666667

00:13:07.606 --> 00:13:10.360 dramatically toward a specific diagnosis.

NOTE Confidence: 0.953476476666667

00:13:10.360 --> 00:13:13.412 We're not going to talk about 3

NOTE Confidence: 0.953476476666667

00:13:13.412 --> 00:13:14.666 amorphic Rhabdomyosarcoma spindle

NOTE Confidence: 0.953476476666667

00:13:14.666 --> 00:13:15.794 cells sclerostin Rhabdomyosarcoma.

NOTE Confidence: 0.953476476666667

00:13:15.794 --> 00:13:19.370 We're going to come back to a little bit

NOTE Confidence: 0.953476476666667

00:13:19.370 --> 00:13:22.010 later in the session this afternoon.

NOTE Confidence: 0.953476476666667

00:13:22.010 --> 00:13:25.340 Another example of alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.953476476666667

00:13:25.340 --> 00:13:27.720 This is really one of the classic

NOTE Confidence: 0.953476476666667

00:13:27.720 --> 00:13:29.736 round cell sarcomas that has

NOTE Confidence: 0.953476476666667

00:13:29.736 --> 00:13:31.580 fairly uniform nuclear morphology.

NOTE Confidence: 0.953476476666667

00:13:31.580 --> 00:13:32.346 The nuclear,

NOTE Confidence: 0.953476476666667

00:13:32.346 --> 00:13:34.644 often much larger than the nuclei

NOTE Confidence: 0.953476476666667

00:13:34.644 --> 00:13:36.040 of Ewing sarcoma,

NOTE Confidence: 0.953476476666667

00:13:36.040 --> 00:13:38.427 and if you're lucky you'll find an

NOTE Confidence: 0.953476476666667

00:13:38.427 --> 00:13:40.224 occasional reflight giant cell as you

NOTE Confidence: 0.953476476666667

00:13:40.224 --> 00:13:42.240 see in the middle of the field here.

NOTE Confidence: 0.953476476666667

00:13:42.240 --> 00:13:44.120 That's a very correct characteristic.

NOTE Confidence: 0.953476476666667

00:13:44.120 --> 00:13:48.620 Giant cell type of alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.953476476666667

00:13:48.620 --> 00:13:52.170 In contrast to embryonal Rhabdomyosarcoma,

NOTE Confidence: 0.953476476666667

00:13:52.170 --> 00:13:53.372 alveolar Rhabdomyosarcoma

NOTE Confidence: 0.953476476666667

00:13:53.372 --> 00:13:55.776 typically shows very strong,

NOTE Confidence: 0.953476476666667

00:13:55.780 --> 00:13:59.520 intense diffuse staining for myogenin.

NOTE Confidence: 0.953476476666667

00:13:59.520 --> 00:14:02.450 The skeletal muscle transcription factor.

NOTE Confidence: 0.953476476666667

00:14:02.450 --> 00:14:05.516 And Brian or Rhabdomyosarcoma usually shows

NOTE Confidence: 0.953476476666667

00:14:05.516 --> 00:14:08.160 much more heterogeneity of the nuclei.

NOTE Confidence: 0.953476476666667

00:14:08.160 --> 00:14:09.780 Some small rounded cells,

NOTE Confidence: 0.953476476666667

00:14:09.780 --> 00:14:11.612 some short spindle cells,

NOTE Confidence: 0.953476476666667

00:14:11.612 --> 00:14:13.902 often embedded within a lucid

NOTE Confidence: 0.953476476666667

00:14:13.902 --> 00:14:15.708 demonised for myxoid stroma.

NOTE Confidence: 0.953476476666667

00:14:15.710 --> 00:14:18.279 And here you can see the striking

NOTE Confidence: 0.953476476666667

00:14:18.279 --> 00:14:20.560 difference in Myogenin immunoreactivity.

NOTE Confidence: 0.953476476666667

00:14:20.560 --> 00:14:22.428 And Brian or Rhabdomyosarcoma

NOTE Confidence: 0.953476476666667

00:14:22.428 --> 00:14:24.296 will show quite variable.

NOTE Confidence: 0.953476476666667

00:14:24.300 --> 00:14:25.956 My agenda and expression.

NOTE Confidence: 0.953476476666667

00:14:25.956 --> 00:14:28.026 It's often between maybe 20

NOTE Confidence: 0.953476476666667

00:14:28.026 --> 00:14:30.104 and 60 or 70% of nuclei,

NOTE Confidence: 0.953476476666667

00:14:30.104 --> 00:14:32.960 but quite different from the strong diffuse

NOTE Confidence: 0.953476476666667

00:14:33.033 --> 00:14:36.195 standing we see in alveolar Rhabdomyosarcoma,

NOTE Confidence: 0.953476476666667

00:14:36.200 --> 00:14:37.226 and in fact,

NOTE Confidence: 0.953476476666667

00:14:37.226 --> 00:14:38.936 in a limited biopsy sample,

NOTE Confidence: 0.953476476666667

00:14:38.940 --> 00:14:41.622 the extent of myogenin is very

NOTE Confidence: 0.953476476666667

00:14:41.622 --> 00:14:44.328 helpful to support the diagnosis

NOTE Confidence: 0.953476476666667

00:14:44.328 --> 00:14:46.800 of alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.953476476666667

00:14:46.800 --> 00:14:48.736 These classes of sarcoma

NOTE Confidence: 0.953476476666667

00:14:48.736 --> 00:14:50.188 have distinct genetics.

NOTE Confidence: 0.953476476666667

00:14:50.190 --> 00:14:50.485 Again,

NOTE Confidence: 0.953476476666667

00:14:50.485 --> 00:14:52.845 I'm going to come back to spindle cells

NOTE Confidence: 0.953476476666667

00:14:52.845 --> 00:14:54.957 sclerostin Rhabdomyosarcoma little bit later.

NOTE Confidence: 0.953476476666667

00:14:54.960 --> 00:14:58.116 The really important thing to remember

NOTE Confidence: 0.953476476666667

00:14:58.116 --> 00:14:59.942 is alveolar Rhabdomyosarcoma's Harbor

NOTE Confidence: 0.953476476666667

00:14:59.942 --> 00:15:03.030 FOXO 1 gene fusions and this is really

NOTE Confidence: 0.953476476666667

00:15:03.096 --> 00:15:05.648 different from Brian or Rhabdomyosarcoma

NOTE Confidence: 0.953476476666667

00:15:05.648 --> 00:15:08.158 which does harbor wrasse mutations,

NOTE Confidence: 0.953476476666667

00:15:08.160 --> 00:15:10.208 but only in a small subset of cases.

NOTE Confidence: 0.8557288575

00:15:12.430 --> 00:15:15.006 So now let's talk a little bit more

NOTE Confidence: 0.8557288575

00:15:15.006 --> 00:15:17.469 detail about alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.8557288575

00:15:17.470 --> 00:15:20.176 Another good clinical clue to the

NOTE Confidence: 0.8557288575

00:15:20.176 --> 00:15:23.680 diagnosis is the fact that alveolar

NOTE Confidence: 0.8557288575

00:15:23.680 --> 00:15:26.001 Rhabdomyosarcoma often presents with

NOTE Confidence: 0.8557288575

00:15:26.001 --> 00:15:28.456 metastasis to cervical lymph notes,

NOTE Confidence: 0.8557288575

00:15:28.460 --> 00:15:32.996 so the presentation can be quite similar to.

NOTE Confidence: 0.8557288575

00:15:33.000 --> 00:15:34.818 Oropharyngeal or nasopharyngeal

NOTE Confidence: 0.8557288575

00:15:34.818 --> 00:15:37.848 carcinomas that often present with

NOTE Confidence: 0.8557288575

00:15:37.848 --> 00:15:40.340 cervical lymph node metastases.

NOTE Confidence: 0.8557288575

00:15:40.340 --> 00:15:42.525 The same thing goes for

NOTE Confidence: 0.8557288575

00:15:42.525 --> 00:15:43.399 alveolar Rhabdomyosarcoma,

NOTE Confidence: 0.8557288575

00:15:43.400 --> 00:15:45.696 so this is always important to have in

NOTE Confidence: 0.8557288575

00:15:45.696 --> 00:15:48.083 mind in your differential diagnosis when

NOTE Confidence: 0.8557288575

00:15:48.083 --> 00:15:51.080 you're dealing with a primitive round cell,

NOTE Confidence: 0.8557288575

00:15:51.080 --> 00:15:52.174 malignant neoplasm.

NOTE Confidence: 0.8557288575

00:15:52.174 --> 00:15:54.909 Alveolar Rhabdomyosarcoma has the worst

NOTE Confidence: 0.8557288575

00:15:54.909 --> 00:15:58.186 prognosis of all of the Rhabdomyosarcoma

NOTE Confidence: 0.8557288575

00:15:58.186 --> 00:16:00.380 types and in terms of differential

NOTE Confidence: 0.8557288575

00:16:00.380 --> 00:16:03.259 diagnosis and one of the reasons that this.

NOTE Confidence: 0.8557288575

00:16:03.260 --> 00:16:05.786 Class of sarcoma is often misdiagnosed

NOTE Confidence: 0.8557288575

00:16:05.786 --> 00:16:08.300 when it arises in the sinus.

NOTE Confidence: 0.8557288575

00:16:08.300 --> 00:16:11.331 Is is the fact that it's quite

NOTE Confidence: 0.8557288575

00:16:11.331 --> 00:16:14.530 common for these tumors to express

NOTE Confidence: 0.8557288575

00:16:14.530 --> 00:16:16.165 keratins and or synaptophysin,

NOTE Confidence: 0.8557288575

00:16:16.165 --> 00:16:18.235 and there was a very nice

NOTE Confidence: 0.8557288575

00:16:18.235 --> 00:16:19.960 paper published by Andrew.

NOTE Confidence: 0.8557288575

00:16:19.960 --> 00:16:22.368 Folks will come back to in a minute

NOTE Confidence: 0.8557288575

00:16:22.368 --> 00:16:24.399 that indicated the really high rate

NOTE Confidence: 0.8557288575

00:16:24.399 --> 00:16:26.104 of standing for these markers.

NOTE Confidence: 0.8557288575

00:16:26.110 --> 00:16:29.036 This is an example of metastatic alveolar

NOTE Confidence: 0.8557288575

00:16:29.036 --> 00:16:31.649 Rhabdomyosarcoma in the cervical lymph node.

NOTE Confidence: 0.8557288575

00:16:31.650 --> 00:16:34.090 You can see this is a nested tumor.

NOTE Confidence: 0.8557288575

00:16:34.090 --> 00:16:36.535 Where the differential diagnosis would

NOTE Confidence: 0.8557288575

00:16:36.535 --> 00:16:40.229 certainly be with a Melanoma and a

NOTE Confidence: 0.8557288575

00:16:40.229 --> 00:16:42.565 primitive poorly differentiated carcinoma.

NOTE Confidence: 0.8557288575

00:16:42.570 --> 00:16:45.534 Obviously it's easy to confirm the

NOTE Confidence: 0.8557288575

00:16:45.534 --> 00:16:48.340 diagnosis with Desmond or myogenic.

NOTE Confidence: 0.8557288575

00:16:48.340 --> 00:16:49.996 And this is the paper I alluded to.

NOTE Confidence: 0.8557288575

00:16:50.000 --> 00:16:51.176 Andrew fopen.

NOTE Confidence: 0.8557288575

00:16:51.176 --> 00:16:53.528 Colleagues from some different

NOTE Confidence: 0.8557288575

00:16:53.528 --> 00:16:55.880 institutions many years ago

NOTE Confidence: 0.8557288575

00:16:55.880 --> 00:16:57.948 highlighted this potentially serious

NOTE Confidence: 0.8557288575

00:16:57.948 --> 00:17:00.533 diagnostic pitfall of expression of

NOTE Confidence: 0.8557288575

00:17:00.533 --> 00:17:03.199 keratins and your endocrine markers

NOTE Confidence: 0.8557288575

00:17:03.199 --> 00:17:04.759 and alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.8557288575

00:17:04.760 --> 00:17:05.840 And in fact,

NOTE Confidence: 0.8557288575

00:17:05.840 --> 00:17:08.000 I would estimate that maybe once

NOTE Confidence: 0.8557288575

00:17:08.000 --> 00:17:10.548 every three or four years coming to

NOTE Confidence: 0.8557288575

00:17:10.548 --> 00:17:13.236 our head and neck on koleji clinic,

NOTE Confidence: 0.8557288575

00:17:13.240 --> 00:17:15.046 there will be a patient who was

NOTE Confidence: 0.8557288575

00:17:15.046 --> 00:17:17.125 thought to have a poorly differentiated

NOTE Confidence: 0.8557288575

00:17:17.125 --> 00:17:18.430 and render carcinoma.

NOTE Confidence: 0.8557288575

00:17:18.430 --> 00:17:21.134 Of the sinonasal tract that turned out in,

NOTE Confidence: 0.8557288575

00:17:21.140 --> 00:17:22.222 in retrospect,

NOTE Confidence: 0.8557288575

00:17:22.222 --> 00:17:26.009 on RE review to be alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.776388805714286

00:17:28.100 --> 00:17:30.074 OK, so that's it for alveolar rabdo.

NOTE Confidence: 0.776388805714286

00:17:30.080 --> 00:17:32.519 Now we're going to go to the second case.

NOTE Confidence: 0.776388805714286

00:17:32.520 --> 00:17:35.130 This is from a 70 year old man who

NOTE Confidence: 0.776388805714286

00:17:35.130 --> 00:17:37.981 had a lesion in the sphenoid sinus

NOTE Confidence: 0.776388805714286

00:17:37.981 --> 00:17:40.779 were starting with some sinus lesions.

NOTE Confidence: 0.776388805714286

00:17:40.780 --> 00:17:43.909 Another curettage specimen,

NOTE Confidence: 0.776388805714286

00:17:43.910 --> 00:17:45.440 not a lot going on here.

NOTE Confidence: 0.776388805714286

00:17:45.440 --> 00:17:47.195 Most of these fragments of

NOTE Confidence: 0.776388805714286

00:17:47.195 --> 00:17:48.599 tissue look pretty normal,

NOTE Confidence: 0.776388805714286

00:17:48.600 --> 00:17:51.015 but I think you'll probably notice there's

NOTE Confidence: 0.776388805714286

00:17:51.015 --> 00:17:54.326 just a few fragments that look very cellular.

NOTE Confidence: 0.776388805714286

00:17:54.330 --> 00:17:56.466 Here is one of those fragments.

NOTE Confidence: 0.776388805714286

00:17:56.470 --> 00:17:59.938 This is a spindle cell neoplasm.

NOTE Confidence: 0.776388805714286

00:17:59.940 --> 00:18:02.970 It's a pretty cellular process.

NOTE Confidence: 0.776388805714286

00:18:02.970 --> 00:18:05.240 Fascicle sheets of spindle cells

NOTE Confidence: 0.776388805714286

00:18:05.240 --> 00:18:07.510 that look fairly uniformly spaced,

NOTE Confidence: 0.776388805714286

00:18:07.510 --> 00:18:10.710 even if this intermediate magnification.

NOTE Confidence: 0.776388805714286

00:18:10.710 --> 00:18:13.216 And other fragments involved by the tumor.

NOTE Confidence: 0.776388805714286

00:18:13.220 --> 00:18:15.236 And now you could appreciate those thin,

NOTE Confidence: 0.776388805714286

00:18:15.240 --> 00:18:17.810 walled, dilated and branching blood

NOTE Confidence: 0.776388805714286

00:18:17.810 --> 00:18:21.789 vessels which we refer to as staghorn

NOTE Confidence: 0.776388805714286

00:18:21.789 --> 00:18:25.099 vessels or hemangiopericytoma like vessels,

NOTE Confidence: 0.776388805714286

00:18:25.100 --> 00:18:29.465 alluding to the tumors that we used to call

NOTE Confidence: 0.776388805714286

00:18:29.465 --> 00:18:31.749 hemangiopericytoma that now do not exist.

NOTE Confidence: 0.776388805714286

00:18:31.750 --> 00:18:33.258 Except for this tumor.

NOTE Confidence: 0.776388805714286

00:18:33.258 --> 00:18:35.143 Little higher powerview and again

NOTE Confidence: 0.776388805714286

00:18:35.143 --> 00:18:37.975 we see those thin walled dilated.

NOTE Confidence: 0.776388805714286

00:18:37.975 --> 00:18:40.395 Somewhat branching blood vessels,

NOTE Confidence: 0.776388805714286

00:18:40.400 --> 00:18:42.638 and you can begin to appreciate

NOTE Confidence: 0.776388805714286

00:18:42.638 --> 00:18:43.757 better than cytology.

NOTE Confidence: 0.776388805714286

00:18:43.760 --> 00:18:46.040 This is a uniform tumor.

NOTE Confidence: 0.776388805714286

00:18:46.040 --> 00:18:49.632 Oval to short spindle cells that are fairly

NOTE Confidence: 0.776388805714286

00:18:49.632 --> 00:18:51.908 evenly distributed and evenly spaced.

NOTE Confidence: 0.776388805714286

00:18:51.910 --> 00:18:53.400 Another nice field of that,

NOTE Confidence: 0.776388805714286

00:18:53.400 --> 00:18:55.690 the higher power with those

NOTE Confidence: 0.776388805714286

00:18:55.690 --> 00:18:56.875 classic blood vessels.

NOTE Confidence: 0.776388805714286

00:18:56.875 --> 00:18:59.245 Some areas are devoid of vessels

NOTE Confidence: 0.776388805714286

00:18:59.245 --> 00:19:01.836 and you could see some somewhat

NOTE Confidence: 0.776388805714286

00:19:01.836 --> 00:19:03.532 sharply defined cell borders,

NOTE Confidence: 0.776388805714286

00:19:03.540 --> 00:19:05.381 and then we come to very high

NOTE Confidence: 0.776388805714286

00:19:05.381 --> 00:19:07.707 power and we can see that the

NOTE Confidence: 0.776388805714286

00:19:07.707 --> 00:19:09.179 nuclear morphology is uniform.

NOTE Confidence: 0.776388805714286

00:19:09.180 --> 00:19:10.998 The chromatin is actually fairly even.

NOTE Confidence: 0.776388805714286

00:19:11.000 --> 00:19:13.736 It's not so course there's limited,

NOTE Confidence: 0.776388805714286

00:19:13.740 --> 00:19:16.236 if any mitotic activity,

NOTE Confidence: 0.776388805714286

00:19:16.236 --> 00:19:19.356 and really not any significant

NOTE Confidence: 0.776388805714286

00:19:19.356 --> 00:19:22.230 nuclear atypia.

NOTE Confidence: 0.776388805714286

00:19:22.230 --> 00:19:23.910 One more high power view again,

NOTE Confidence: 0.776388805714286

00:19:23.910 --> 00:19:26.400 you could see the even spacing

NOTE Confidence: 0.776388805714286

00:19:26.400 --> 00:19:28.060 of these uniform nuclei.

NOTE Confidence: 0.776388805714286

00:19:28.060 --> 00:19:29.924 So what's our differential

NOTE Confidence: 0.776388805714286

00:19:29.924 --> 00:19:31.788 diagnosis for this case?

NOTE Confidence: 0.776388805714286

00:19:31.790 --> 00:19:33.210 Given the anatomic sites

NOTE Confidence: 0.776388805714286

00:19:33.210 --> 00:19:34.985 and given the prior case,

NOTE Confidence: 0.776388805714286

00:19:34.990 --> 00:19:37.808 we could also think about Melanoma carcinoma,

NOTE Confidence: 0.776388805714286

00:19:37.808 --> 00:19:39.956 which would obviously be much more

NOTE Confidence: 0.776388805714286

00:19:39.956 --> 00:19:42.194 common than the tumors were talking

NOTE Confidence: 0.776388805714286

00:19:42.194 --> 00:19:44.390 about in the session this afternoon.

NOTE Confidence: 0.776388805714286

00:19:44.390 --> 00:19:46.082 Maybe monophasic synovial sarcoma?

NOTE Confidence: 0.776388805714286

00:19:46.082 --> 00:19:49.600 This is a very uniform spindle cell neoplasm.

NOTE Confidence: 0.776388805714286

00:19:49.600 --> 00:19:49.933 Although,

NOTE Confidence: 0.776388805714286

00:19:49.933 --> 00:19:51.598 as the residents discussed this

NOTE Confidence: 0.776388805714286

00:19:51.598 --> 00:19:53.420 morning in the slide seminar,

NOTE Confidence: 0.776388805714286

00:19:53.420 --> 00:19:55.980 so noville sarcoma usually has

NOTE Confidence: 0.776388805714286

00:19:55.980 --> 00:19:58.028 somewhat overlapping nuclei because

NOTE Confidence: 0.776388805714286

00:19:58.028 --> 00:20:00.638 they have such limited cytoplasm.

NOTE Confidence: 0.776388805714286

00:20:00.640 --> 00:20:02.540 In this case, the nuclear,

NOTE Confidence: 0.776388805714286

00:20:02.540 --> 00:20:04.650 the nuclear, fairly evenly spaced,

NOTE Confidence: 0.776388805714286

00:20:04.650 --> 00:20:08.286 which would argue against synovial sarcoma.

NOTE Confidence: 0.776388805714286

00:20:08.290 --> 00:20:11.517 And then finally. Possibly a gloom angio.

NOTE Confidence: 0.776388805714286

00:20:11.520 --> 00:20:13.740 Perry cytoma, whatever that is.

NOTE Confidence: 0.776388805714286

00:20:13.740 --> 00:20:16.687 This is a tumor that's also called

NOTE Confidence: 0.776388805714286

00:20:16.687 --> 00:20:20.530 sinonasal hemangiopericytoma.

NOTE Confidence: 0.776388805714286

00:20:20.530 --> 00:20:22.546 Smooth muscle actin and some areas.

NOTE Confidence: 0.776388805714286

00:20:22.550 --> 00:20:24.050 It's not so impressive,

NOTE Confidence: 0.776388805714286

00:20:24.050 --> 00:20:26.300 but on other areas there was

NOTE Confidence: 0.776388805714286

00:20:26.380 --> 00:20:28.620 quite strong staining for SMAD.

NOTE Confidence: 0.776388805714286

00:20:28.620 --> 00:20:30.060 One other stand I'm going to

NOTE Confidence: 0.776388805714286

00:20:30.060 --> 00:20:31.510 show you is beta catenin.

NOTE Confidence: 0.776388805714286

00:20:31.510 --> 00:20:32.846 This is normal tissue.

NOTE Confidence: 0.776388805714286

00:20:32.846 --> 00:20:35.496 You can see the the upper respiratory

NOTE Confidence: 0.776388805714286

00:20:35.496 --> 00:20:37.784 epithelium has beautiful membranous

NOTE Confidence: 0.776388805714286

00:20:37.784 --> 00:20:40.072 staining for beta catenin,

NOTE Confidence: 0.776388805714286

00:20:40.080 --> 00:20:42.726 whereas in the tumor we have

NOTE Confidence: 0.776388805714286

00:20:42.726 --> 00:20:44.490 this incredibly intense nuclear

NOTE Confidence: 0.776388805714286

00:20:44.573 --> 00:20:46.928 and cytoplasmic staining was in,

NOTE Confidence: 0.776388805714286

00:20:46.930 --> 00:20:49.591 which is an aberrant pattern for

NOTE Confidence: 0.776388805714286

00:20:49.591 --> 00:20:52.196 nuclear Vatican for forbidding catenin.

NOTE Confidence: 0.776388805714286

00:20:52.200 --> 00:20:55.201 The other markers that were investigated

NOTE Confidence: 0.776388805714286

00:20:55.201 --> 00:20:57.356 in this case were negative,

NOTE Confidence: 0.776388805714286

00:20:57.360 --> 00:21:00.000 and I'm sure you've already guessed

NOTE Confidence: 0.776388805714286

00:21:00.000 --> 00:21:02.849 this is William Angio Perry Cytoma.

NOTE Confidence: 0.776388805714286

00:21:02.850 --> 00:21:05.139 This is a tumor that for many

NOTE Confidence: 0.776388805714286

00:21:05.139 --> 00:21:06.120 years was called

NOTE Confidence: 0.921085925833333

00:21:06.196 --> 00:21:08.260 sinonasal hemangiopericytoma.

NOTE Confidence: 0.921085925833333

00:21:08.260 --> 00:21:10.000 This terminology has evolved a bit

NOTE Confidence: 0.921085925833333

00:21:10.000 --> 00:21:13.650 over the years, in part because the

NOTE Confidence: 0.921085925833333

00:21:13.650 --> 00:21:15.856 hemangiopericytoma term has sort of

NOTE Confidence: 0.921085925833333

00:21:15.856 --> 00:21:17.488 been abandoned and most organ systems.

NOTE Confidence: 0.921085925833333

00:21:17.490 --> 00:21:19.403 So in the head neck there's trying

NOTE Confidence: 0.921085925833333

00:21:19.403 --> 00:21:21.244 to get away from it as well.

NOTE Confidence: 0.921085925833333

00:21:21.250 --> 00:21:23.812 This is a lesion that arises most

NOTE Confidence: 0.921085925833333

00:21:23.812 --> 00:21:26.040 commonly in the ethmoid sinus.

NOTE Confidence: 0.921085925833333

00:21:26.040 --> 00:21:28.368 It's benign, although it can recur

NOTE Confidence: 0.921085925833333

00:21:28.368 --> 00:21:30.630 locally in about 1/3 of cases.

NOTE Confidence: 0.921085925833333

00:21:30.630 --> 00:21:33.876 Most patients present with a nasal

NOTE Confidence: 0.921085925833333

00:21:33.876 --> 00:21:36.956 or sinus polyp fairly small in size,

NOTE Confidence: 0.921085925833333

00:21:36.956 --> 00:21:39.140 although occasionally they can be large.

NOTE Confidence: 0.921085925833333

00:21:39.140 --> 00:21:41.580 They often have some expression

NOTE Confidence: 0.921085925833333

00:21:41.580 --> 00:21:43.044 of actin filaments.

NOTE Confidence: 0.921085925833333

00:21:43.050 --> 00:21:46.002 These are thought to be perivascular cells.

NOTE Confidence: 0.921085925833333

00:21:46.002 --> 00:21:49.398 The cells that have contractile properties,

NOTE Confidence: 0.921085925833333

00:21:49.400 --> 00:21:51.740 which is why they express

NOTE Confidence: 0.921085925833333

00:21:51.740 --> 00:21:53.144 the actin filaments.

NOTE Confidence: 0.921085925833333

00:21:53.150 --> 00:21:56.318 Are there negative for Desmond Keratins?

NOTE Confidence: 0.921085925833333

00:21:56.320 --> 00:21:59.720 But we learned in 2015 by these Nice

NOTE Confidence: 0.921085925833333

00:21:59.720 --> 00:22:02.578 papers published by two different groups,

NOTE Confidence: 0.921085925833333

00:22:02.580 --> 00:22:04.608 one of them by Florian Haller

NOTE Confidence: 0.921085925833333

00:22:04.608 --> 00:22:06.612 and Abasa Gammy, and colleagues,

NOTE Confidence: 0.921085925833333

00:22:06.612 --> 00:22:09.067 the other by Jersey Losada,

NOTE Confidence: 0.921085925833333

00:22:09.070 --> 00:22:10.057 Mark home yet,

NOTE Confidence: 0.921085925833333

00:22:10.057 --> 00:22:12.031 and and a few other colleagues

NOTE Confidence: 0.921085925833333

00:22:12.031 --> 00:22:14.341 in and head and neck pathology

NOTE Confidence: 0.921085925833333

00:22:14.341 --> 00:22:15.865 that Glow Manju Perry.

NOTE Confidence: 0.921085925833333

00:22:15.870 --> 00:22:19.209 Cytoma harbors consistent

NOTE Confidence: 0.921085925833333

00:22:19.209 --> 00:22:23.849 activating mutations in C, TNN B1,

NOTE Confidence: 0.921085925833333

00:22:23.849 --> 00:22:26.447 the genes that encodes beta catenin.

NOTE Confidence: 0.921085925833333

00:22:26.450 --> 00:22:29.160 Which leads to this incredibly

NOTE Confidence: 0.921085925833333

00:22:29.160 --> 00:22:31.328 intense nuclear and cytoplasmic

NOTE Confidence: 0.921085925833333

00:22:31.328 --> 00:22:33.250 staining for beta catenin,

NOTE Confidence: 0.921085925833333

00:22:33.250 --> 00:22:35.398 which has become a very easy

NOTE Confidence: 0.921085925833333

00:22:35.398 --> 00:22:37.008 to apply diagnostic marker.

NOTE Confidence: 0.921085925833333

00:22:37.008 --> 00:22:40.116 The only problem with beta catenin in

NOTE Confidence: 0.921085925833333

00:22:40.116 --> 00:22:42.951 head and neck spindle cell neoplasms is

NOTE Confidence: 0.921085925833333

00:22:42.951 --> 00:22:46.170 the fact that it's not entirely specific.

NOTE Confidence: 0.921085925833333

00:22:46.170 --> 00:22:47.106 My colleagues,

NOTE Confidence: 0.921085925833333

00:22:47.106 --> 00:22:49.446 Vicki Jo and Chris Fletcher,

NOTE Confidence: 0.921085925833333

00:22:49.450 --> 00:22:51.921 published this follow up paper and head

NOTE Confidence: 0.921085925833333

00:22:51.921 --> 00:22:55.650 neck pathology soon after investigating

NOTE Confidence: 0.921085925833333

00:22:55.650 --> 00:22:57.450 immunohistochemistry for beta catenin.

NOTE Confidence: 0.921085925833333

00:22:57.450 --> 00:22:59.700 In a range of different

NOTE Confidence: 0.921085925833333

00:22:59.700 --> 00:23:01.190 spindle cell neoplasms,

NOTE Confidence: 0.921085925833333

00:23:01.190 --> 00:23:03.350 and they confirmed the beautiful staining

NOTE Confidence: 0.921085925833333

00:23:03.350 --> 00:23:06.058 we find in Glen Angio Perry Cytoma.

NOTE Confidence: 0.921085925833333

00:23:06.060 --> 00:23:08.740 But at least you should be aware that

NOTE Confidence: 0.921085925833333

00:23:08.740 --> 00:23:10.866 the majority of solitary fibrous

NOTE Confidence: 0.921085925833333

00:23:10.866 --> 00:23:13.256 tumors and synovial sarcomas also

NOTE Confidence: 0.921085925833333

00:23:13.256 --> 00:23:15.986 shown aberrant nuclear beta catenin,

NOTE Confidence: 0.921085925833333

00:23:15.990 --> 00:23:17.650 but it really doesn't show

NOTE Confidence: 0.921085925833333

00:23:17.650 --> 00:23:18.646 that incredibly diffuse,

NOTE Confidence: 0.921085925833333

00:23:18.650 --> 00:23:20.485 intense staining that you see

NOTE Confidence: 0.921085925833333

00:23:20.485 --> 00:23:22.320 in Gorman Joe Perry cytoma.

NOTE Confidence: 0.8837533

00:23:24.740 --> 00:23:27.060 Now we go on to case three or

NOTE Confidence: 0.8837533

00:23:27.060 --> 00:23:29.319 still in the Sinonasal region.

NOTE Confidence: 0.8837533

00:23:29.320 --> 00:23:31.273 This is from a 36 year old

NOTE Confidence: 0.8837533

00:23:31.273 --> 00:23:33.478 man with a nasal cavity mass

NOTE Confidence: 0.8837533

00:23:33.480 --> 00:23:37.140 extending into the ethmoid sinus.

NOTE Confidence: 0.8837533

00:23:37.140 --> 00:23:40.108 This is a much larger tumor than the

NOTE Confidence: 0.8837533

00:23:40.108 --> 00:23:41.970 previous samples I've shown you.

NOTE Confidence: 0.8837533

00:23:41.970 --> 00:23:44.490 This is a really purple looking

NOTE Confidence: 0.8837533

00:23:44.490 --> 00:23:46.170 tumor from scanning magnification.

NOTE Confidence: 0.8837533

00:23:46.170 --> 00:23:47.390 Little bit higher power.

NOTE Confidence: 0.8837533

00:23:47.390 --> 00:23:49.220 You could also you can already

NOTE Confidence: 0.8837533

00:23:49.278 --> 00:23:51.750 begin to appreciate that this tumor

NOTE Confidence: 0.8837533

00:23:51.750 --> 00:23:54.030 has a fascicular architecture.

NOTE Confidence: 0.8837533

00:23:54.030 --> 00:23:56.080 Again, it has that somewhat

NOTE Confidence: 0.8837533

00:23:56.080 --> 00:23:56.900 purple appearance.

NOTE Confidence: 0.8837533

00:23:56.900 --> 00:23:59.378 You can see some finwall dilated

NOTE Confidence: 0.8837533

00:23:59.378 --> 00:24:01.430 blood vessels at the top.

NOTE Confidence: 0.8837533

00:24:01.430 --> 00:24:04.244 We have entrapment of these these glands.

NOTE Confidence: 0.8837533

00:24:04.250 --> 00:24:06.410 The glands of the sinus,

NOTE Confidence: 0.8837533

00:24:06.410 --> 00:24:08.265 which sometimes look a little

NOTE Confidence: 0.8837533

00:24:08.265 --> 00:24:09.007 bit hyperplastic.

NOTE Confidence: 0.8837533

00:24:09.010 --> 00:24:12.436 They look very prominent in this

NOTE Confidence: 0.8837533

00:24:12.436 --> 00:24:14.720 embedded within this tumor.

NOTE Confidence: 0.8837533

00:24:14.720 --> 00:24:17.142 Higher Power area and we can now

NOTE Confidence: 0.8837533

00:24:17.142 --> 00:24:18.940 appreciate the nuclear morphology.

NOTE Confidence: 0.8837533

00:24:18.940 --> 00:24:23.260 Very bland, fairly uniform nuclei,

NOTE Confidence: 0.8837533

00:24:23.260 --> 00:24:24.270 somewhat overlapping.

NOTE Confidence: 0.8837533

00:24:24.270 --> 00:24:27.300 Coming back to our previous comment.

NOTE Confidence: 0.8837533

00:24:27.300 --> 00:24:30.200 Areas are much more purple,

NOTE Confidence: 0.8837533

00:24:30.200 --> 00:24:33.000 uniformly fascicular we see those

NOTE Confidence: 0.8837533

00:24:33.000 --> 00:24:35.800 fascicles nicely at high power.

NOTE Confidence: 0.8837533

00:24:35.800 --> 00:24:38.552 This tumor comes all the way up to

NOTE Confidence: 0.8837533

00:24:38.552 --> 00:24:41.118 the surface of the sinonasal mucosa.

NOTE Confidence: 0.8837533

00:24:41.120 --> 00:24:43.728 And then we come again to high power

NOTE Confidence: 0.8837533

00:24:43.730 --> 00:24:46.103 and you can see the uniform nuclear

NOTE Confidence: 0.8837533

00:24:46.103 --> 00:24:48.363 morphology and we even can see some

NOTE Confidence: 0.8837533

00:24:48.363 --> 00:24:50.940 areas with these wiry collagen

NOTE Confidence: 0.8837533

00:24:50.940 --> 00:24:53.790 bundles between the tumor cells.

NOTE Confidence: 0.8837533

00:24:53.790 --> 00:24:55.974 So most of what I've described

NOTE Confidence: 0.8837533

00:24:55.974 --> 00:24:58.634 now would fit very well with

NOTE Confidence: 0.8837533

00:24:58.634 --> 00:25:00.326 monophasic synovial sarcoma.

NOTE Confidence: 0.8837533

00:25:00.330 --> 00:25:02.451 But I think given the rarity of

NOTE Confidence: 0.8837533

00:25:02.451 --> 00:25:04.708 that tumor in this anatomic site,

NOTE Confidence: 0.8837533

00:25:04.710 --> 00:25:06.798 we have to think of some of the other

NOTE Confidence: 0.8837533

00:25:06.798 --> 00:25:08.967 tumors we discussed this morning like

NOTE Confidence: 0.8837533

00:25:08.967 --> 00:25:10.842 malignant peripheral nerve sheath tumor.

NOTE Confidence: 0.8837533

00:25:10.850 --> 00:25:11.822 Perhaps leiomyosarcoma,

NOTE Confidence: 0.8837533

00:25:11.822 --> 00:25:15.224 although this tumor is not so eosinophilic,

NOTE Confidence: 0.8837533

00:25:15.230 --> 00:25:17.220 it's a bit more purple.

NOTE Confidence: 0.8837533

00:25:17.220 --> 00:25:21.360 And then biphenotypic sinonasal sarcoma.

NOTE Confidence: 0.8837533

00:25:21.360 --> 00:25:23.944 As I go through each of these examples,

NOTE Confidence: 0.8837533

00:25:23.950 --> 00:25:25.402 I'm sure even if you don't

NOTE Confidence: 0.8837533

00:25:25.402 --> 00:25:26.560 know what you're looking at,

NOTE Confidence: 0.8837533

00:25:26.560 --> 00:25:29.580 you could probably guess the diagnosis by

NOTE Confidence: 0.8837533

00:25:29.580 --> 00:25:33.120 how I'm framing the differential diagnosis.

NOTE Confidence: 0.8837533

00:25:33.120 --> 00:25:34.640 Let's look at administer chemistry.

NOTE Confidence: 0.8837533

00:25:34.640 --> 00:25:36.690 Smooth muscle actin was positive

NOTE Confidence: 0.8837533

00:25:36.690 --> 00:25:37.920 in this case.

NOTE Confidence: 0.8837533

00:25:37.920 --> 00:25:40.250 A fairly kind of variable.

NOTE Confidence: 0.8837533

00:25:40.250 --> 00:25:41.729 Not so impressive.

NOTE Confidence: 0.8837533

00:25:41.729 --> 00:25:44.194 Overall positive in these areas.

NOTE Confidence: 0.8837533

00:25:44.200 --> 00:25:47.056 There was some limited staining for Desmond,

NOTE Confidence: 0.8837533

00:25:47.060 --> 00:25:49.484 and there was also some limited

NOTE Confidence: 0.8837533

00:25:49.484 --> 00:25:51.100 staining for S100 protein,

NOTE Confidence: 0.8837533

00:25:51.100 --> 00:25:53.998 but this is the characteristic nuclear

NOTE Confidence: 0.8837533

00:25:53.998 --> 00:25:56.328 and cytoplasmic staining pattern you

NOTE Confidence: 0.8837533

00:25:56.328 --> 00:25:59.208 should look for to feel confident that

NOTE Confidence: 0.8837533

00:25:59.208 --> 00:26:02.629 you're dealing with true staining for S 100.

NOTE Confidence: 0.8837533

00:26:02.630 --> 00:26:04.480 TLE One was also positive.

NOTE Confidence: 0.8837533

00:26:04.480 --> 00:26:07.008 We didn't talk about Tilly one this morning.

NOTE Confidence: 0.8837533

00:26:07.010 --> 00:26:09.344 Tillie one has been developed as

NOTE Confidence: 0.8837533

00:26:09.344 --> 00:26:11.550 a marker for synovial sarcoma.

NOTE Confidence: 0.8837533

00:26:11.550 --> 00:26:14.154 It was identified by gene expression

NOTE Confidence: 0.8837533

00:26:14.154 --> 00:26:15.754 profiling about 15 years ago,

NOTE Confidence: 0.8837533

00:26:15.754 --> 00:26:18.263 but we now know that T one is

NOTE Confidence: 0.8837533

00:26:18.263 --> 00:26:19.647 really not so specific.

NOTE Confidence: 0.8837533

00:26:19.650 --> 00:26:22.112 It's only has kind of moderate specificity,

NOTE Confidence: 0.8837533

00:26:22.112 --> 00:26:24.280 although the sensitivity for

NOTE Confidence: 0.8837533

00:26:24.280 --> 00:26:26.990 synovial sarcoma is very high.

NOTE Confidence: 0.8837533

00:26:26.990 --> 00:26:29.250 So to summarize the immuno.

NOTE Confidence: 0.8837533

00:26:29.250 --> 00:26:32.855 SM Afocal Desmond S 100 TL E1.

NOTE Confidence: 0.8837533

00:26:32.860 --> 00:26:34.540 Whereas epithelial markers

NOTE Confidence: 0.8837533

00:26:34.540 --> 00:26:37.340 and socks 10 were negative.

NOTE Confidence: 0.8837533

00:26:37.340 --> 00:26:40.264 This is in fact,

NOTE Confidence: 0.8837533

00:26:40.264 --> 00:26:42.457 Biphenotypic sinonasal sarcoma.

NOTE Confidence: 0.8837533

00:26:42.460 --> 00:26:44.812 So what is this tumor that goes

NOTE Confidence: 0.8837533

00:26:44.812 --> 00:26:47.110 by this unusual descriptive name?

NOTE Confidence: 0.8837533

00:26:47.110 --> 00:26:47.433 Well,

NOTE Confidence: 0.8837533

00:26:47.433 --> 00:26:48.402 lucky for us,

NOTE Confidence: 0.8837533

00:26:48.402 --> 00:26:51.492 the name that we now use is a little

NOTE Confidence: 0.8837533

00:26:51.492 --> 00:26:53.646 bit easier to remember than the

NOTE Confidence: 0.8837533

00:26:53.646 --> 00:26:56.287 first descriptor of this tumor type.

NOTE Confidence: 0.905911205454546

00:26:56.290 --> 00:26:58.420 Lewis and colleagues published this

NOTE Confidence: 0.905911205454546

00:26:58.420 --> 00:27:01.696 paper almost ten years ago now where they

NOTE Confidence: 0.905911205454546

00:27:01.696 --> 00:27:04.294 first to find this interesting tumor

NOTE Confidence: 0.905911205454546

00:27:04.300 --> 00:27:06.814 that they called low grade sinonasal

NOTE Confidence: 0.905911205454546

00:27:06.814 --> 00:27:09.820 sarcoma with neural and myogenic features,

NOTE Confidence: 0.905911205454546

00:27:09.820 --> 00:27:12.625 which is a perfect description

NOTE Confidence: 0.905911205454546

00:27:12.625 --> 00:27:14.869 for this tumor type.

NOTE Confidence: 0.905911205454546

00:27:14.870 --> 00:27:17.710 This is a low grade sarcoma that seems

NOTE Confidence: 0.905911205454546

00:27:17.710 --> 00:27:20.926 to be unique to the sinonasal tract.

NOTE Confidence: 0.905911205454546

00:27:20.930 --> 00:27:23.828 It occurs in adults over a wide age range,

NOTE Confidence: 0.905911205454546

00:27:23.830 --> 00:27:27.340 slightly more common in female patients.

NOTE Confidence: 0.905911205454546

00:27:27.340 --> 00:27:29.806 It's most common in the nasal

NOTE Confidence: 0.905911205454546

00:27:29.806 --> 00:27:31.450 cavity and ethmoid sinus.

NOTE Confidence: 0.905911205454546

00:27:31.450 --> 00:27:34.384 Despite the fact that we call this a sarcoma,

NOTE Confidence: 0.905911205454546

00:27:34.390 --> 00:27:36.550 it doesn't appear to have metastatic

NOTE Confidence: 0.905911205454546

00:27:36.550 --> 00:27:38.586 potential, at least thus far.

NOTE Confidence: 0.905911205454546

00:27:38.586 --> 00:27:40.681 No metastatic cases have been

NOTE Confidence: 0.905911205454546

00:27:40.681 --> 00:27:42.327 published with molecular confirmation

NOTE Confidence: 0.905911205454546

00:27:42.327 --> 00:27:45.590 as I'll come back to in a minute.

NOTE Confidence: 0.905911205454546

00:27:45.590 --> 00:27:48.887 There have been several tumor related deaths.

NOTE Confidence: 0.905911205454546

00:27:48.890 --> 00:27:51.506 This seems to be a very rare event,

NOTE Confidence: 0.905911205454546

00:27:51.510 --> 00:27:54.720 primarily due to intracranial extension.

NOTE Confidence: 0.905911205454546

00:27:54.720 --> 00:27:56.220 Given the anatomic site,

NOTE Confidence: 0.905911205454546

00:27:56.220 --> 00:27:59.290 so these tumors can be locally aggressive,

NOTE Confidence: 0.905911205454546

00:27:59.290 --> 00:28:01.672 they seem to recur in about

NOTE Confidence: 0.905911205454546

00:28:01.672 --> 00:28:04.542 30 to 40% of cases.

NOTE Confidence: 0.905911205454546

00:28:04.542 --> 00:28:07.050 Sometimes patients experience

NOTE Confidence: 0.905911205454546

00:28:07.050 --> 00:28:09.558 multiple local recurrences.

NOTE Confidence: 0.905911205454546

00:28:09.560 --> 00:28:11.438 As we saw in our case,

NOTE Confidence: 0.905911205454546

00:28:11.440 --> 00:28:13.832 these tumors look strikingly

NOTE Confidence: 0.905911205454546

00:28:13.832 --> 00:28:16.430 similar to monophasic.

NOTE Confidence: 0.905911205454546

00:28:16.430 --> 00:28:20.482 Synovial sarcoma, including cytologic,

NOTE Confidence: 0.905911205454546

00:28:20.482 --> 00:28:23.638 uniformity, fascicular growth.

NOTE Confidence: 0.905911205454546

00:28:23.640 --> 00:28:26.195 Often somewhat WAVY nuclei and

NOTE Confidence: 0.905911205454546

00:28:26.195 --> 00:28:28.750 indistinct cytoplasm that have delicate

NOTE Confidence: 0.905911205454546

00:28:28.830 --> 00:28:31.758 stromal collagen similar to synovial

NOTE Confidence: 0.905911205454546

00:28:31.758 --> 00:28:34.688 sarcoma and solitary fibrous tumor.

NOTE Confidence: 0.905911205454546

00:28:34.690 --> 00:28:38.290 It's common to see occasional

NOTE Confidence: 0.905911205454546

00:28:38.290 --> 00:28:40.301 staghorn vessels confusingly,

NOTE Confidence: 0.905911205454546

00:28:40.301 --> 00:28:43.727 these tumors sometimes have rare rhabdom.

NOTE Confidence: 0.905911205454546

00:28:43.730 --> 00:28:44.504 I'll blast,

NOTE Confidence: 0.905911205454546

00:28:44.504 --> 00:28:46.052 which can be highlighted

NOTE Confidence: 0.905911205454546

00:28:46.052 --> 00:28:48.015 by Desmin myogenin Myo D1,

NOTE Confidence: 0.905911205454546

00:28:48.015 --> 00:28:50.780 which might lead you to consider the

NOTE Confidence: 0.905911205454546

00:28:50.862 --> 00:28:53.697 possibility of a malignant peripheral.

NOTE Confidence: 0.905911205454546

00:28:53.700 --> 00:28:55.712 Nerve sheath tumor with

NOTE Confidence: 0.905911205454546

00:28:55.712 --> 00:28:56.718 heterologous elements.

NOTE Confidence: 0.905911205454546

00:28:56.720 --> 00:28:59.276 But those tumors are almost invariably

NOTE Confidence: 0.905911205454546

00:28:59.276 --> 00:29:02.449 hygrade with a very high mitotic rates.

NOTE Confidence: 0.905911205454546

00:29:02.450 --> 00:29:04.790 Striking nuclear atypia necrosis

NOTE Confidence: 0.905911205454546

00:29:04.790 --> 00:29:07.130 or as biphenotypic sinonasal

NOTE Confidence: 0.905911205454546

00:29:07.130 --> 00:29:09.929 sarcoma is remarkably uniform.

NOTE Confidence: 0.905911205454546

00:29:09.930 --> 00:29:11.774 Rarely shows necrosis and

NOTE Confidence: 0.905911205454546

00:29:11.774 --> 00:29:14.079 has a low mitotic rate.

NOTE Confidence: 0.905911205454546

00:29:14.080 --> 00:29:15.829 The defining immunophenotypic

NOTE Confidence: 0.905911205454546

00:29:15.829 --> 00:29:18.744 features of this tumor type.

NOTE Confidence: 0.905911205454546

00:29:18.750 --> 00:29:22.635 ARCO expression of S 100 and muscle

NOTE Confidence: 0.905911205454546

00:29:22.635 --> 00:29:24.854 markers most often smooth muscle

NOTE Confidence: 0.905911205454546

00:29:24.854 --> 00:29:26.934 actin and muscle specific actin,

NOTE Confidence: 0.905911205454546

00:29:26.940 --> 00:29:29.436 but you can sometimes see Desmond

NOTE Confidence: 0.905911205454546

00:29:29.436 --> 00:29:33.066 as well and as I mentioned the small

NOTE Confidence: 0.905911205454546

00:29:33.066 --> 00:29:35.814 subset of cases with rabdo myoblasts

NOTE Confidence: 0.905911205454546

00:29:35.814 --> 00:29:38.132 have myogenin Myo D1 expression

NOTE Confidence: 0.905911205454546

00:29:38.132 --> 00:29:40.778 only in the skeletal muscle cells.

NOTE Confidence: 0.905911205454546

00:29:40.780 --> 00:29:42.358 As we saw in our case,

NOTE Confidence: 0.905911205454546

00:29:42.360 --> 00:29:44.472 TLE one can be positive which

NOTE Confidence: 0.905911205454546

00:29:44.472 --> 00:29:46.496 could lead to confusion with

NOTE Confidence: 0.905911205454546

00:29:46.496 --> 00:29:48.140 monophasic synovial sarcoma,

NOTE Confidence: 0.905911205454546

00:29:48.140 --> 00:29:50.000 and finally, socks tennis negative.

NOTE Confidence: 0.905911205454546

00:29:50.000 --> 00:29:51.960 So these are not true.

NOTE Confidence: 0.905911205454546

00:29:51.960 --> 00:29:54.040 Peripheral nerve sheath tumors.

NOTE Confidence: 0.905911205454546

00:29:54.040 --> 00:29:56.144 These are not MPNST's.

NOTE Confidence: 0.905911205454546

00:29:56.144 --> 00:29:59.408 They do not show true nerve

NOTE Confidence: 0.905911205454546

00:29:59.408 --> 00:30:00.496 sheath differentiation.

NOTE Confidence: 0.905911205454546

00:30:00.500 --> 00:30:01.272 In fact,

NOTE Confidence: 0.905911205454546

00:30:01.272 --> 00:30:03.974 we don't really know what the lineages

NOTE Confidence: 0.905911205454546

00:30:03.974 --> 00:30:06.610 of these unusual tumors but coexpression

NOTE Confidence: 0.905911205454546

00:30:06.610 --> 00:30:10.411 of S 100 and actin filaments is a

NOTE Confidence: 0.905911205454546

00:30:10.411 --> 00:30:12.487 very helpful diagnostic feature.

NOTE Confidence: 0.905911205454546

00:30:12.490 --> 00:30:14.820 Andre Oliveira and colleagues published

NOTE Confidence: 0.905911205454546

00:30:14.820 --> 00:30:17.150 this beautiful paper in Nature

NOTE Confidence: 0.905911205454546

00:30:17.218 --> 00:30:20.690 Genetics in 2014 from the Mayo Clinic,

NOTE Confidence: 0.905911205454546

00:30:20.690 --> 00:30:24.488 identifying a recurrent pack 3 mammal

NOTE Confidence: 0.905911205454546

00:30:24.488 --> 00:30:27.582 3 gene fusion in biphenotypic,

NOTE Confidence: 0.905911205454546

00:30:27.582 --> 00:30:30.930 sidell nasal sarcoma and over the next

NOTE Confidence: 0.905911205454546

00:30:30.930 --> 00:30:32.880 few years several different groups

NOTE Confidence: 0.905911205454546

00:30:32.880 --> 00:30:35.465 of published cases of biphenotypic

NOTE Confidence: 0.905911205454546

00:30:35.465 --> 00:30:38.110 sinonasal sarcoma with other packs,

NOTE Confidence: 0.905911205454546

00:30:38.110 --> 00:30:42.809 3 Fusion partners including FOXO One and two.

NOTE Confidence: 0.905911205454546

00:30:42.809 --> 00:30:45.672 But it seems that PAX three is

NOTE Confidence: 0.905911205454546

00:30:45.672 --> 00:30:48.190 almost always involved in the

NOTE Confidence: 0.905911205454546

00:30:48.190 --> 00:30:50.338 pathogenesis of these tumors.

NOTE Confidence: 0.905911205454546

00:30:50.340 --> 00:30:53.189 A couple of nice large series published

NOTE Confidence: 0.905911205454546

00:30:53.189 --> 00:30:55.410 since the initial descriptions.

NOTE Confidence: 0.905911205454546

00:30:55.410 --> 00:30:57.762 This was a nice paper published

NOTE Confidence: 0.905911205454546

00:30:57.762 --> 00:30:59.330 by a diverse group

NOTE Confidence: 0.83574612875

00:30:59.409 --> 00:31:02.647 of colleagues in Virchows archives in 2018.

NOTE Confidence: 0.83574612875

00:31:02.647 --> 00:31:04.669 A fairly large series where you

NOTE Confidence: 0.83574612875

00:31:04.669 --> 00:31:07.270 can see the anatomic distribution,

NOTE Confidence: 0.83574612875

00:31:07.270 --> 00:31:09.250 ethmoid cavity and nasal sinus,

NOTE Confidence: 0.83574612875

00:31:09.250 --> 00:31:10.468 or most common,

NOTE Confidence: 0.83574612875

00:31:10.468 --> 00:31:13.310 as we saw in the initial description.

NOTE Confidence: 0.83574612875

00:31:13.310 --> 00:31:16.062 And in this paper, mammal three was was

NOTE Confidence: 0.83574612875

00:31:16.062 --> 00:31:19.216 still the most common fusion partner by far.

NOTE Confidence: 0.83574612875

00:31:19.220 --> 00:31:21.608 A small subset of cases had

NOTE Confidence: 0.83574612875

00:31:21.610 --> 00:31:25.096 foxo one and NCO A1 fusions.

NOTE Confidence: 0.83574612875

00:31:25.100 --> 00:31:26.711 We haven't identified

NOTE Confidence: 0.83574612875

00:31:26.711 --> 00:31:28.859 all the fusion partners.

NOTE Confidence: 0.83574612875

00:31:28.860 --> 00:31:33.468 A small subset of cases have as of

NOTE Confidence: 0.83574612875

00:31:33.468 --> 00:31:37.387 yet undefined PAX 3 fusion partners.

NOTE Confidence: 0.83574612875

00:31:37.390 --> 00:31:38.359 And then finally,

NOTE Confidence: 0.83574612875

00:31:38.359 --> 00:31:39.974 most recently another large series

NOTE Confidence: 0.83574612875

00:31:39.974 --> 00:31:42.120 by the French sarcoma group with a

NOTE Confidence: 0.83574612875

00:31:42.120 --> 00:31:44.672 few other collaborators from the US.

NOTE Confidence: 0.83574612875

00:31:44.672 --> 00:31:47.630 This was another large series where

NOTE Confidence: 0.83574612875

00:31:47.630 --> 00:31:50.675 packs 3 mammal free fusions were found

NOTE Confidence: 0.83574612875

00:31:50.675 --> 00:31:55.905 in 37 out of 41 molecularly confirmed cases.

NOTE Confidence: 0.83574612875

00:31:55.910 --> 00:31:57.454 They identified another fusion

NOTE Confidence: 0.83574612875

00:31:57.454 --> 00:32:00.862 partner in the form of WW T R1 and

NOTE Confidence: 0.83574612875

00:32:00.862 --> 00:32:03.070 we have those same fusion partners

NOTE Confidence: 0.83574612875

00:32:03.150 --> 00:32:06.930 we saw in the previous case.

NOTE Confidence: 0.83574612875

00:32:06.930 --> 00:32:09.390 And again, my colleague Vicki Jo,

NOTE Confidence: 0.83574612875

00:32:09.390 --> 00:32:10.830 with some other collaborators

NOTE Confidence: 0.83574612875

00:32:10.830 --> 00:32:11.910 in my department,

NOTE Confidence: 0.83574612875

00:32:11.910 --> 00:32:14.350 published a study with us a few years

NOTE Confidence: 0.83574612875

00:32:14.350 --> 00:32:17.185 ago where we we looked at using packs

NOTE Confidence: 0.83574612875

00:32:17.185 --> 00:32:19.935 3 administer chemistry as a surrogate

NOTE Confidence: 0.83574612875

00:32:19.935 --> 00:32:22.155 for biphenotypic sinonasal sarcoma.

NOTE Confidence: 0.83574612875

00:32:22.160 --> 00:32:23.944 This worked very well.

NOTE Confidence: 0.83574612875

00:32:23.944 --> 00:32:27.849 We had a uniform staining in all 15 cases.

NOTE Confidence: 0.83574612875

00:32:27.850 --> 00:32:30.346 We investigated many of these cases.

NOTE Confidence: 0.83574612875

00:32:30.350 --> 00:32:32.042 We confirm by fish as you

NOTE Confidence: 0.83574612875

00:32:32.042 --> 00:32:33.979 see in the lower left image,

NOTE Confidence: 0.83574612875

00:32:33.980 --> 00:32:36.818 we have the break apart using.

NOTE Confidence: 0.83574612875

00:32:36.820 --> 00:32:39.082 Pax three directed probes and the

NOTE Confidence: 0.83574612875

00:32:39.082 --> 00:32:41.492 tumors we might consider in the

NOTE Confidence: 0.83574612875

00:32:41.492 --> 00:32:43.156 differential diagnosis are almost

NOTE Confidence: 0.83574612875

00:32:43.156 --> 00:32:45.280 always negative for PAX three.

NOTE Confidence: 0.83574612875

00:32:45.280 --> 00:32:47.840 This is a very easy to apply marker.

NOTE Confidence: 0.83574612875

00:32:47.840 --> 00:32:51.158 We have a very nice nuclear staining in

NOTE Confidence: 0.83574612875

00:32:51.158 --> 00:32:53.748 a characteristic case of biphenotypic

NOTE Confidence: 0.83574612875

00:32:53.748 --> 00:32:55.765 sinonasal sarcoma and one other

NOTE Confidence: 0.83574612875

00:32:55.765 --> 00:32:57.715 example where you see it just

NOTE Confidence: 0.83574612875

00:32:57.715 --> 00:32:59.348 underneath the surface epithelium.

NOTE Confidence: 0.938280502727273

00:33:02.130 --> 00:33:03.198 All right, we have a few

NOTE Confidence: 0.938280502727273

00:33:03.198 --> 00:33:04.310 more cases to go through.

NOTE Confidence: 0.938280502727273

00:33:04.310 --> 00:33:06.390 A few more different topics.

NOTE Confidence: 0.938280502727273

00:33:06.390 --> 00:33:09.901 Case four is from a 56 year old

NOTE Confidence: 0.938280502727273

00:33:09.901 --> 00:33:12.386 woman who presented with dyspnea,

NOTE Confidence: 0.938280502727273

00:33:12.390 --> 00:33:14.214 and she was found to have a large,

NOTE Confidence: 0.938280502727273

00:33:14.220 --> 00:33:15.366 soft palate tumor.

NOTE Confidence: 0.938280502727273

00:33:15.366 --> 00:33:18.040 So we're now moving away from the

NOTE Confidence: 0.938280502727273

00:33:18.121 --> 00:33:20.150 sinus is down into the palace.

NOTE Confidence: 0.938280502727273

00:33:20.150 --> 00:33:21.950 This was from the resection.

NOTE Confidence: 0.938280502727273

00:33:21.950 --> 00:33:24.566 It was actually kind of a debulking because

NOTE Confidence: 0.938280502727273

00:33:24.566 --> 00:33:27.044 this patient was having breathing difficulty.

NOTE Confidence: 0.938280502727273

00:33:27.044 --> 00:33:29.761 They recognized his tumor and they did

NOTE Confidence: 0.938280502727273

00:33:29.761 --> 00:33:31.543 surgery urgently so that she could.

NOTE Confidence: 0.938280502727273

00:33:31.550 --> 00:33:33.270 They could make sure they

NOTE Confidence: 0.938280502727273

00:33:33.270 --> 00:33:34.646 could maintain an airway.

NOTE Confidence: 0.938280502727273

00:33:34.650 --> 00:33:36.630 We have this very large,

NOTE Confidence: 0.938280502727273

00:33:36.630 --> 00:33:38.518 highly cellular purple tumor.

NOTE Confidence: 0.938280502727273

00:33:38.518 --> 00:33:40.406 Which had higher power.

NOTE Confidence: 0.938280502727273

00:33:40.410 --> 00:33:42.846 We can now begin to appreciate

NOTE Confidence: 0.938280502727273

00:33:42.850 --> 00:33:44.863 the Fascicular architecture.

NOTE Confidence: 0.938280502727273

00:33:44.863 --> 00:33:46.876 The primitive nuclear

NOTE Confidence: 0.938280502727273

00:33:46.876 --> 00:33:48.705 morphology and other areas.

NOTE Confidence: 0.938280502727273

00:33:48.705 --> 00:33:51.400 We have a slightly nested or cordlike

NOTE Confidence: 0.938280502727273

00:33:51.474 --> 00:33:54.274 architecture with this somewhat

NOTE Confidence: 0.938280502727273

00:33:54.274 --> 00:33:56.218 prominent collagenous stroma.

NOTE Confidence: 0.938280502727273

00:33:56.220 --> 00:33:59.510 In other areas we have much more

NOTE Confidence: 0.938280502727273

00:33:59.510 --> 00:34:00.920 uniformly fascicular architecture.

NOTE Confidence: 0.938280502727273

00:34:00.920 --> 00:34:03.242 Primitive nuclear morphology.

NOTE Confidence: 0.938280502727273

00:34:03.242 --> 00:34:06.338 This somewhat coarse chromatin.

NOTE Confidence: 0.938280502727273

00:34:06.340 --> 00:34:07.400 Higher power in the first

NOTE Confidence: 0.938280502727273

00:34:07.400 --> 00:34:08.460 area as I showed you,

NOTE Confidence: 0.938280502727273

00:34:08.460 --> 00:34:11.386 we have the nests and cords of

NOTE Confidence: 0.938280502727273

00:34:11.386 --> 00:34:13.989 primitive cells within this very dense.

NOTE Confidence: 0.938280502727273

00:34:13.990 --> 00:34:16.750 Collagen rich stroma.

NOTE Confidence: 0.938280502727273

00:34:16.750 --> 00:34:18.970 In some areas in the stroma,

NOTE Confidence: 0.938280502727273

00:34:18.970 --> 00:34:20.112 rich foci.

NOTE Confidence: 0.938280502727273

00:34:20.112 --> 00:34:23.538 These nests almost look like they're

NOTE Confidence: 0.938280502727273

00:34:23.538 --> 00:34:27.026 forming these cleft like or pseudo

NOTE Confidence: 0.938280502727273

00:34:27.026 --> 00:34:29.282 vascular spaces a little bit LV.

NOTE Confidence: 0.938280502727273

00:34:29.282 --> 00:34:29.848 Older in.

NOTE Confidence: 0.938280502727273

00:34:29.850 --> 00:34:33.177 In cytology this is another classic

NOTE Confidence: 0.938280502727273

00:34:33.177 --> 00:34:37.059 appearance to this type of sarcoma.

NOTE Confidence: 0.938280502727273

00:34:37.060 --> 00:34:38.894 And finally we look at high power

NOTE Confidence: 0.938280502727273

00:34:38.894 --> 00:34:41.178 and we see the primitive chromatin.

NOTE Confidence: 0.938280502727273

00:34:41.180 --> 00:34:44.912 Course very limited cytoplasm in these

NOTE Confidence: 0.938280502727273

00:34:44.912 --> 00:34:48.140 nests within this collagenous matrix.

NOTE Confidence: 0.938280502727273

00:34:48.140 --> 00:34:49.920 So what about differential diagnosis?

NOTE Confidence: 0.938280502727273

00:34:49.920 --> 00:34:52.372 If you notice the vascular appearance

NOTE Confidence: 0.938280502727273

00:34:52.372 --> 00:34:54.964 or you the pseudo vascular parents,

NOTE Confidence: 0.938280502727273

00:34:54.970 --> 00:34:57.800 perhaps you might consider angiosarcoma.

NOTE Confidence: 0.938280502727273

00:34:57.800 --> 00:34:59.408 If you thought the funny stroma

NOTE Confidence: 0.938280502727273

00:34:59.408 --> 00:35:01.640 looked a bit like bone matrix,

NOTE Confidence: 0.938280502727273

00:35:01.640 --> 00:35:04.540 osteoid that hadn't quite calcified.

NOTE Confidence: 0.938280502727273

00:35:04.540 --> 00:35:06.880 You might consider extra stone lost,

NOTE Confidence: 0.938280502727273

00:35:06.880 --> 00:35:08.050 you Sir coma.

NOTE Confidence: 0.938280502727273

00:35:08.050 --> 00:35:10.780 This morning we saw an example of

NOTE Confidence: 0.938280502727273

00:35:10.869 --> 00:35:12.658 sclerosing epithelioid fibrosarcoma,

NOTE Confidence: 0.938280502727273

00:35:12.658 --> 00:35:15.053 so you might consider that

NOTE Confidence: 0.938280502727273

00:35:15.053 --> 00:35:17.290 as a possible diagnosis,

NOTE Confidence: 0.938280502727273

00:35:17.290 --> 00:35:21.330 given the very dense expression.

NOTE Confidence: 0.938280502727273

00:35:21.330 --> 00:35:24.630 Excuse me at deposition of collagen.

NOTE Confidence: 0.938280502727273

00:35:24.630 --> 00:35:27.342 Sclerosing Rhabdomyosarcoma, perhaps?

NOTE Confidence: 0.938280502727273

00:35:27.342 --> 00:35:29.150 Synovial sarcoma,

NOTE Confidence: 0.938280502727273

00:35:29.150 --> 00:35:30.502 given the uniformity of

NOTE Confidence: 0.938280502727273

00:35:30.502 --> 00:35:31.516 the nucleo morphology?

NOTE Confidence: 0.900500069

00:35:33.560 --> 00:35:35.024 So let's look at a minister

NOTE Confidence: 0.900500069

00:35:35.024 --> 00:35:36.000 chemistry on this case.

NOTE Confidence: 0.900500069

00:35:36.000 --> 00:35:38.560 Desman is very strongly positive

NOTE Confidence: 0.900500069

00:35:38.560 --> 00:35:41.120 and pretty much every cell.

NOTE Confidence: 0.900500069

00:35:41.120 --> 00:35:42.938 And mild D1 is also strongly

NOTE Confidence: 0.900500069

00:35:42.938 --> 00:35:45.140 positive even in the scanning image.

NOTE Confidence: 0.900500069

00:35:45.140 --> 00:35:47.546 You can appreciate that every nucleus

NOTE Confidence: 0.900500069

00:35:47.546 --> 00:35:49.609 has strong expression of my OG.

NOTE Confidence: 0.900500069

00:35:49.610 --> 00:35:51.071 One high power.

NOTE Confidence: 0.900500069

00:35:51.071 --> 00:35:54.480 We confirm that impression from low power.

NOTE Confidence: 0.900500069

00:35:54.480 --> 00:35:57.000 In contrast, if you look at myogenin,

NOTE Confidence: 0.900500069

00:35:57.000 --> 00:35:59.814 it's only a very small subset

NOTE Confidence: 0.900500069

00:35:59.814 --> 00:36:02.670 of nuclei that are positive.

NOTE Confidence: 0.900500069

00:36:02.670 --> 00:36:03.708 So in summary,

NOTE Confidence: 0.900500069

00:36:03.708 --> 00:36:05.784 we have skeletal muscle marker expression.

NOTE Confidence: 0.900500069

00:36:05.790 --> 00:36:08.688 We have negative staining for EMA.

NOTE Confidence: 0.900500069

00:36:08.690 --> 00:36:12.980 The vascular transcription factor ERG

NOTE Confidence: 0.900500069

00:36:12.980 --> 00:36:15.300 and the sclerosing epithelioid fibrous

NOTE Confidence: 0.900500069

00:36:15.300 --> 00:36:18.330 sarcoma marker mark for all negative.

NOTE Confidence: 0.900500069

00:36:18.330 --> 00:36:21.094 And you've guessed it,

NOTE Confidence: 0.900500069

00:36:21.094 --> 00:36:23.858 this is sclerostin Rhabdomyosarcoma.

NOTE Confidence: 0.900500069

00:36:23.860 --> 00:36:25.852 So at the beginning of the session this

NOTE Confidence: 0.900500069

00:36:25.852 --> 00:36:27.572 afternoon I mentioned I was going to

NOTE Confidence: 0.900500069

00:36:27.572 --> 00:36:29.578 come back to this topic of spindle cell,

NOTE Confidence: 0.900500069

00:36:29.580 --> 00:36:31.696 sclerosing revenue mile sarcoma.

NOTE Confidence: 0.900500069

00:36:31.696 --> 00:36:34.870 This is the most recent addition

NOTE Confidence: 0.900500069

00:36:34.870 --> 00:36:37.775 to these sarcomas that shows

NOTE Confidence: 0.900500069

00:36:37.775 --> 00:36:39.518 skeletal muscle differentiation.

NOTE Confidence: 0.900500069

00:36:39.520 --> 00:36:41.256 Spindle cell sclerostin Rhabdomyosarcoma

NOTE Confidence: 0.900500069

00:36:41.256 --> 00:36:44.838 is occur most often in the head and neck,

NOTE Confidence: 0.900500069

00:36:44.840 --> 00:36:47.912 although you can see them in the extremities

NOTE Confidence: 0.900500069

00:36:47.912 --> 00:36:51.178 and trunk or in parrot testicular locations.

NOTE Confidence: 0.900500069

00:36:51.180 --> 00:36:53.980 Paratesticular tumors are almost

NOTE Confidence: 0.900500069

00:36:53.980 --> 00:36:56.620 only seen in infants,

NOTE Confidence: 0.900500069

00:36:56.620 --> 00:36:57.860 male infants,

NOTE Confidence: 0.900500069

00:36:57.860 --> 00:37:00.070 and young boys as they'll

NOTE Confidence: 0.900500069

00:37:00.070 --> 00:37:01.120 come back to in a minute.

NOTE Confidence: 0.900500069

00:37:01.120 --> 00:37:04.666 These tumors often have my old

NOTE Confidence: 0.900500069

00:37:04.666 --> 00:37:07.030 one trans activating mutations.

NOTE Confidence: 0.900500069

00:37:07.030 --> 00:37:08.615 There is a slight female

NOTE Confidence: 0.900500069

00:37:08.615 --> 00:37:09.566 predominant for that.

NOTE Confidence: 0.900500069

00:37:09.570 --> 00:37:11.628 Group of tumors.

NOTE Confidence: 0.900500069

00:37:11.630 --> 00:37:12.252 These tumors,

NOTE Confidence: 0.900500069

00:37:12.252 --> 00:37:14.429 as we saw in our clinical case,

NOTE Confidence: 0.900500069

00:37:14.430 --> 00:37:17.965 often present as a rapidly growing mass.

NOTE Confidence: 0.900500069

00:37:17.970 --> 00:37:20.736 And symptoms are related to local

NOTE Confidence: 0.900500069

00:37:20.736 --> 00:37:23.363 compression because of their very

NOTE Confidence: 0.900500069

00:37:23.363 --> 00:37:25.573 characteristic anatomic sites of

NOTE Confidence: 0.900500069

00:37:25.573 --> 00:37:28.078 presentation in the head neck.

NOTE Confidence: 0.900500069

00:37:28.080 --> 00:37:31.656 There are another group of spindle

NOTE Confidence: 0.900500069

00:37:31.656 --> 00:37:33.161 cells thrusting rhabdomyosarcoma's

NOTE Confidence: 0.900500069

00:37:33.161 --> 00:37:35.027 that do not have mild you.

NOTE Confidence: 0.900500069

00:37:35.030 --> 00:37:37.550 One mutations that instead have gene

NOTE Confidence: 0.900500069

00:37:37.550 --> 00:37:39.980 fusions as they'll come back to in a minute.

NOTE Confidence: 0.900500069

00:37:39.980 --> 00:37:42.374 Those tumors have a very good prognosis.

NOTE Confidence: 0.900500069

00:37:42.380 --> 00:37:44.340 Those are almost only seen

NOTE Confidence: 0.900500069

00:37:44.340 --> 00:37:45.908 in very young infants,

NOTE Confidence: 0.900500069

00:37:45.910 --> 00:37:48.730 sometimes with congenital presentation.

NOTE Confidence: 0.900500069

00:37:48.730 --> 00:37:50.140 In contrast,

NOTE Confidence: 0.900500069

00:37:50.140 --> 00:37:53.602 the miod one mutant spindle cell

NOTE Confidence: 0.900500069

00:37:53.602 --> 00:37:55.596 speros in rhabdomyosarcoma's really

NOTE Confidence: 0.900500069

00:37:55.596 --> 00:37:58.104 have a dismal prognosis with a

NOTE Confidence: 0.900500069

00:37:58.104 --> 00:38:02.588 less than 20% five year survival.

NOTE Confidence: 0.900500069

00:38:02.590 --> 00:38:04.288 At one end of the spectrum,

NOTE Confidence: 0.900500069

00:38:04.290 --> 00:38:08.106 these tumors look very similar to

NOTE Confidence: 0.900500069

00:38:08.110 --> 00:38:09.961 leiomyosarcoma's with intersecting

NOTE Confidence: 0.900500069

00:38:09.961 --> 00:38:13.046 fascicles of elongated spindle cells

NOTE Confidence: 0.900500069

00:38:13.046 --> 00:38:16.167 with abundant bright pink seidel plasm.

NOTE Confidence: 0.900500069

00:38:16.170 --> 00:38:18.810 Some cases you'll appreciate us

NOTE Confidence: 0.900500069

00:38:18.810 --> 00:38:20.922 true skeletal muscle differentiation

NOTE Confidence: 0.900500069

00:38:20.922 --> 00:38:22.498 at the HD level.

NOTE Confidence: 0.900500069

00:38:22.500 --> 00:38:24.875 We have these occasional polygonal

NOTE Confidence: 0.900500069

00:38:24.875 --> 00:38:28.333 or strap like cells that have really

NOTE Confidence: 0.900500069

00:38:28.333 --> 00:38:31.840 intense somewhat orange or red or cytoplasm.

NOTE Confidence: 0.900500069

00:38:31.840 --> 00:38:35.032 Other examples have a much more

NOTE Confidence: 0.900500069

00:38:35.032 --> 00:38:36.628 primitive fibrosarcoma like

NOTE Confidence: 0.900500069

00:38:36.628 --> 00:38:38.696 appearance with coarse chromatin,

NOTE Confidence: 0.900500069

00:38:38.696 --> 00:38:41.768 a high mitotic rate and limited

NOTE Confidence: 0.900500069

00:38:41.768 --> 00:38:44.887 set up plastic differentiation at

NOTE Confidence: 0.900500069

00:38:44.887 --> 00:38:46.189 the other end of the spectrum.

NOTE Confidence: 0.900500069

00:38:46.190 --> 00:38:49.195 We have these incredibly densely

NOTE Confidence: 0.900500069

00:38:49.195 --> 00:38:51.599 hyalinized sclerosing tumors where

NOTE Confidence: 0.900500069

00:38:51.599 --> 00:38:55.904 we have this osteoid matrix like

NOTE Confidence: 0.900500069

00:38:55.904 --> 00:38:59.696 deposition of hyalinized collagen.

NOTE Confidence: 0.900500069

00:38:59.700 --> 00:39:01.667 And other cases kind of sit somewhere

NOTE Confidence: 0.900500069

00:39:01.667 --> 00:39:03.779 in the middle as we saw in our case,

NOTE Confidence: 0.900500069

00:39:03.780 --> 00:39:06.685 in our clinical case for this discussion.

NOTE Confidence: 0.900500069

00:39:06.690 --> 00:39:09.258 I think this this example has

NOTE Confidence: 0.900500069

00:39:09.258 --> 00:39:10.542 some fascicular architecture,

NOTE Confidence: 0.900500069

00:39:10.550 --> 00:39:13.693 but you could still appreciate in the

NOTE Confidence: 0.900500069

00:39:13.693 --> 00:39:16.600 background the dense collagenous stroma.

NOTE Confidence: 0.900500069

00:39:16.600 --> 00:39:18.888 Desmond is diffusely positive,

NOTE Confidence: 0.900500069

00:39:18.888 --> 00:39:21.099 similar to many rhabdomyosarcoma's

NOTE Confidence: 0.900500069

00:39:21.099 --> 00:39:23.055 the best skeletal muscle

NOTE Confidence: 0.900500069

00:39:23.055 --> 00:39:25.500 transcription factor to used for

NOTE Confidence: 0.79827149

00:39:25.573 --> 00:39:27.118 this tumor is miod want.

NOTE Confidence: 0.79827149

00:39:27.120 --> 00:39:29.240 Some cases are entirely

NOTE Confidence: 0.79827149

00:39:29.240 --> 00:39:30.830 negative for myogenic,

NOTE Confidence: 0.79827149

00:39:30.830 --> 00:39:33.476 so if you only run myogenin,

NOTE Confidence: 0.79827149

00:39:33.480 --> 00:39:35.916 you may get confused and think

NOTE Confidence: 0.79827149

00:39:35.916 --> 00:39:38.251 you're not dealing with this

NOTE Confidence: 0.79827149

00:39:38.251 --> 00:39:40.779 distinctive form of Rhabdomyosarcoma.

NOTE Confidence: 0.79827149

00:39:40.780 --> 00:39:43.264 So as I mentioned, these tumors

NOTE Confidence: 0.79827149

00:39:43.264 --> 00:39:46.249 most often have my O D1 mutations.

NOTE Confidence: 0.79827149

00:39:46.250 --> 00:39:48.812 There were three proper papers published

NOTE Confidence: 0.79827149

00:39:48.812 --> 00:39:51.580 the same year by Marc Ladanyi,

NOTE Confidence: 0.79827149

00:39:51.580 --> 00:39:54.500 Pockrus Hogendoorn and Christina Senescu.

NOTE Confidence: 0.79827149

00:39:54.500 --> 00:39:57.846 These really nice papers in nature Genetics,

NOTE Confidence: 0.79827149

00:39:57.850 --> 00:40:00.300 Journal of pathology and genes

NOTE Confidence: 0.79827149

00:40:00.300 --> 00:40:02.750 chromosomes and cancer identifying this

NOTE Confidence: 0.79827149

00:40:02.828 --> 00:40:05.112 very common recurrent trans activating

NOTE Confidence: 0.79827149

00:40:05.112 --> 00:40:08.360 mutation in my O D1 which leads to

NOTE Confidence: 0.79827149

00:40:08.448 --> 00:40:11.479 very strong expression of the mild D1.

NOTE Confidence: 0.79827149

00:40:11.480 --> 00:40:13.562 Protein, as I showed you in

NOTE Confidence: 0.79827149

00:40:13.562 --> 00:40:14.950 the last few slides.

NOTE Confidence: 0.79827149

00:40:14.950 --> 00:40:18.647 But we've learned over time that these tumors

NOTE Confidence: 0.79827149

00:40:18.647 --> 00:40:22.300 do not always show my OG one mutations.

NOTE Confidence: 0.79827149

00:40:22.300 --> 00:40:24.939 Some of these tumors have gene fusions

NOTE Confidence: 0.79827149

00:40:24.939 --> 00:40:27.797 with a variety of fusion partners and

NOTE Confidence: 0.79827149

00:40:27.797 --> 00:40:30.275 this nice paper published two years

NOTE Confidence: 0.79827149

00:40:30.346 --> 00:40:32.816 ago by the Group of Memorial Narse.

NOTE Confidence: 0.79827149

00:40:32.820 --> 00:40:35.090 Agaram was the first author.

NOTE Confidence: 0.79827149

00:40:35.090 --> 00:40:37.463 He's one of the bone and soft

NOTE Confidence: 0.79827149

00:40:37.463 --> 00:40:39.389 tissue pathologists at memorial in

NOTE Confidence: 0.79827149

00:40:39.389 --> 00:40:41.484 New York with Cristina Antonescu.

NOTE Confidence: 0.79827149

00:40:41.490 --> 00:40:44.346 And they're suggesting that you really

NOTE Confidence: 0.79827149

00:40:44.346 --> 00:40:47.394 should think of these as different

NOTE Confidence: 0.79827149

00:40:47.394 --> 00:40:50.580 risk groups depending on what the

NOTE Confidence: 0.79827149

00:40:50.580 --> 00:40:53.000 underlying molecular pathogenesis is.

NOTE Confidence: 0.79827149

00:40:53.000 --> 00:40:55.088 The mild you want mutant tumors,

NOTE Confidence: 0.79827149

00:40:55.090 --> 00:40:56.382 which are pretty much

NOTE Confidence: 0.79827149

00:40:56.382 --> 00:40:57.997 all the tumors in adults,

NOTE Confidence: 0.79827149

00:40:58.000 --> 00:41:00.400 have mild you one mutations.

NOTE Confidence: 0.79827149

00:41:00.400 --> 00:41:03.070 I have a really poor prognosis,

NOTE Confidence: 0.79827149

00:41:03.070 --> 00:41:05.818 whereas the tumors that present in

NOTE Confidence: 0.79827149

00:41:05.818 --> 00:41:08.651 very young infants have various gene

NOTE Confidence: 0.79827149

00:41:08.651 --> 00:41:11.850 fusions and have a very good prognosis.

NOTE Confidence: 0.79827149

00:41:11.850 --> 00:41:14.436 If you don't find miodowa mutations,

NOTE Confidence: 0.79827149

00:41:14.440 --> 00:41:17.386 those tumors also have a fairly

NOTE Confidence: 0.79827149

00:41:17.386 --> 00:41:18.368 good prognosis.

NOTE Confidence: 0.79827149

00:41:18.370 --> 00:41:20.834 There are some second hits that have been

NOTE Confidence: 0.79827149

00:41:20.834 --> 00:41:22.846 identified as you see from this study,

NOTE Confidence: 0.79827149

00:41:22.850 --> 00:41:24.578 but really the thing to remember

NOTE Confidence: 0.79827149

00:41:24.578 --> 00:41:26.700 is the my OG one mutations.

NOTE Confidence: 0.896061406666667

00:41:29.010 --> 00:41:31.188 All right, we are back to the nasal cavity.

NOTE Confidence: 0.896061406666667

00:41:31.190 --> 00:41:33.584 The last case I'm going to discuss

NOTE Confidence: 0.896061406666667

00:41:33.584 --> 00:41:36.060 this afternoon is from a young woman

NOTE Confidence: 0.896061406666667

00:41:36.060 --> 00:41:38.676 who was found to have a right nasal

NOTE Confidence: 0.896061406666667

00:41:38.676 --> 00:41:40.866 cavity mass when she presented to

NOTE Confidence: 0.896061406666667

00:41:40.866 --> 00:41:42.786 her primary care physician with

NOTE Confidence: 0.896061406666667

00:41:42.790 --> 00:41:45.670 a unremitting nasal congestion.

NOTE Confidence: 0.896061406666667

00:41:45.670 --> 00:41:48.550 Another very cellular tumor.

NOTE Confidence: 0.896061406666667

00:41:48.550 --> 00:41:50.700 This is the excision specimen,

NOTE Confidence: 0.896061406666667

00:41:50.700 --> 00:41:52.725 another tumor with a purple

NOTE Confidence: 0.896061406666667

00:41:52.725 --> 00:41:54.345 appearance from low power.

NOTE Confidence: 0.896061406666667

00:41:54.350 --> 00:41:55.700 But when you begin to look

NOTE Confidence: 0.896061406666667

00:41:55.700 --> 00:41:57.359 at a little bit higher power,

NOTE Confidence: 0.896061406666667

00:41:57.360 --> 00:41:59.971 you begin to see that these tumor

NOTE Confidence: 0.896061406666667

00:41:59.971 --> 00:42:02.266 cells have abundant eosinophilic

NOTE Confidence: 0.896061406666667

00:42:02.266 --> 00:42:04.618 granular cytoplasm.

NOTE Confidence: 0.896061406666667

00:42:04.620 --> 00:42:07.119 And in the background you see this

NOTE Confidence: 0.896061406666667

00:42:07.119 --> 00:42:09.092 very delicate capillary vascular

NOTE Confidence: 0.896061406666667

00:42:09.092 --> 00:42:11.596 network that compartmentalizes the

NOTE Confidence: 0.896061406666667

00:42:11.596 --> 00:42:14.820 tumor into these delicate bundles,

NOTE Confidence: 0.896061406666667

00:42:14.820 --> 00:42:16.996 or nests higher power.

NOTE Confidence: 0.896061406666667

00:42:16.996 --> 00:42:20.643 You can see the central nucleoli quite

NOTE Confidence: 0.896061406666667

00:42:20.643 --> 00:42:22.558 uniform appearance to the nuclei,

NOTE Confidence: 0.896061406666667

00:42:22.560 --> 00:42:26.196 although there are some very slight

NOTE Confidence: 0.896061406666667

00:42:26.196 --> 00:42:28.014 nuclear contour irregularities.

NOTE Confidence: 0.896061406666667

00:42:28.020 --> 00:42:30.099 Some of the cells have a little

NOTE Confidence: 0.896061406666667

00:42:30.099 --> 00:42:32.000 bit more optically clear cytoplasm,

NOTE Confidence: 0.896061406666667

00:42:32.000 --> 00:42:34.608 but most of the cells have this granular.

NOTE Confidence: 0.896061406666667

00:42:34.610 --> 00:42:36.398 Eosinophilic appearance and now

NOTE Confidence: 0.896061406666667

00:42:36.398 --> 00:42:39.080 in this image these images you

NOTE Confidence: 0.896061406666667

00:42:39.157 --> 00:42:41.247 can nicely see that delicate.

NOTE Confidence: 0.896061406666667

00:42:41.250 --> 00:42:43.113 Capillary vascular network.

NOTE Confidence: 0.896061406666667

00:42:43.113 --> 00:42:46.672 Looking very similar to the vascular

NOTE Confidence: 0.896061406666667

00:42:46.672 --> 00:42:50.620 pattern of clear cell renal cell carcinoma.

NOTE Confidence: 0.896061406666667

00:42:50.620 --> 00:42:53.764 High power you can nicely see those nucleoli.

NOTE Confidence: 0.896061406666667

00:42:53.770 --> 00:42:56.082 Occasional cells are multinucleated

NOTE Confidence: 0.896061406666667

00:42:56.082 --> 00:43:00.240 with slightly more nuclear atypia.

NOTE Confidence: 0.896061406666667

00:43:00.240 --> 00:43:00.647 Finally,

NOTE Confidence: 0.896061406666667

00:43:00.647 --> 00:43:03.089 high power we now begin to

NOTE Confidence: 0.896061406666667

00:43:03.089 --> 00:43:05.020 appreciate the mitotic activity.

NOTE Confidence: 0.896061406666667

00:43:05.020 --> 00:43:07.318 We found some scattered mitotic figures.

NOTE Confidence: 0.896061406666667

00:43:07.320 --> 00:43:10.470 I think it was up to maybe two or three

NOTE Confidence: 0.896061406666667

00:43:10.556 --> 00:43:13.628 and 10 high power fields in this case.

NOTE Confidence: 0.896061406666667

00:43:13.630 --> 00:43:15.526 So what is our differential diagnosis?

NOTE Confidence: 0.896061406666667

00:43:15.530 --> 00:43:18.758 I mentioned metastatic renal cell carcinoma,

NOTE Confidence: 0.896061406666667

00:43:18.760 --> 00:43:21.217 but we have to think about the

NOTE Confidence: 0.896061406666667

00:43:21.217 --> 00:43:23.617 other tumor types that also present

NOTE Confidence: 0.896061406666667

00:43:23.617 --> 00:43:25.737 with a similar nested architecture

NOTE Confidence: 0.896061406666667

00:43:25.737 --> 00:43:28.488 and a similar vascular pattern.

NOTE Confidence: 0.896061406666667

00:43:28.490 --> 00:43:30.062 So obviously today's session

NOTE Confidence: 0.896061406666667

00:43:30.062 --> 00:43:31.684 is on soft tissue tumors,

NOTE Confidence: 0.896061406666667

00:43:31.684 --> 00:43:33.630 so it can't be renal cell carcinoma.

NOTE Confidence: 0.896061406666667

00:43:33.630 --> 00:43:36.942 This is a this is a good testing strategy.

NOTE Confidence: 0.896061406666667

00:43:36.950 --> 00:43:39.920 Alveolar soft part sarcoma perhaps

NOTE Confidence: 0.896061406666667

00:43:39.920 --> 00:43:43.370 clear cell sarcoma malignant pecoma.

NOTE Confidence: 0.896061406666667

00:43:43.370 --> 00:43:45.130 Maybe we should consider a

NOTE Confidence: 0.896061406666667

00:43:45.130 --> 00:43:46.258 these tumors possibly.

NOTE Confidence: 0.896061406666667

00:43:46.258 --> 00:43:48.514 So let's look at a minister.

NOTE Confidence: 0.896061406666667

00:43:48.520 --> 00:43:51.982 Chemistry TFE 3 is very strongly

NOTE Confidence: 0.896061406666667

00:43:51.982 --> 00:43:53.713 and intensely positive,

NOTE Confidence: 0.896061406666667

00:43:53.720 --> 00:43:56.864 so that could suggest several of

NOTE Confidence: 0.896061406666667

00:43:56.864 --> 00:43:58.960 the possibilities I mentioned.

NOTE Confidence: 0.896061406666667

00:43:58.960 --> 00:44:00.770 The only other marker that

NOTE Confidence: 0.896061406666667

00:44:00.770 --> 00:44:02.758 was positive was HMB 45.

NOTE Confidence: 0.896061406666667

00:44:02.758 --> 00:44:05.003 That marker of melanocytic differentiation

NOTE Confidence: 0.896061406666667

00:44:05.003 --> 00:44:07.759 and it was not so impressive,

NOTE Confidence: 0.896061406666667

00:44:07.760 --> 00:44:10.208 but it did show that really

NOTE Confidence: 0.896061406666667

00:44:10.208 --> 00:44:11.840 distinctive granular subtle plasmic

NOTE Confidence: 0.896061406666667

00:44:11.908 --> 00:44:14.751 pattern of staining which we look

NOTE Confidence: 0.896061406666667

00:44:14.751 --> 00:44:17.973 for for tumors that have melanosomes.

NOTE Confidence: 0.896061406666667

00:44:17.980 --> 00:44:20.840 So to summarize, the immunophenotype.

NOTE Confidence: 0.896061406666667

00:44:20.840 --> 00:44:23.132 Muscle markers were negative.

NOTE Confidence: 0.896061406666667

00:44:23.132 --> 00:44:25.424 As well as Melanie.

NOTE Confidence: 0.896061406666667

00:44:25.430 --> 00:44:27.788 If you're thinking of Melanoma S

NOTE Confidence: 0.896061406666667

00:44:27.788 --> 00:44:29.360 100 stocks tender negative,

NOTE Confidence: 0.896061406666667

00:44:29.360 --> 00:44:31.910 those are also uniformly positive

NOTE Confidence: 0.896061406666667

00:44:31.910 --> 00:44:33.950 and clear cell sarcoma.

NOTE Confidence: 0.896061406666667

00:44:33.950 --> 00:44:34.838 And finally,

NOTE Confidence: 0.896061406666667

00:44:34.838 --> 00:44:37.058 thinking about renal cell carcinoma

NOTE Confidence: 0.896061406666667

00:44:37.058 --> 00:44:39.050 characins were also negative.

NOTE Confidence: 0.896061406666667

00:44:39.050 --> 00:44:42.560 This is a malignant pecoma.

NOTE Confidence: 0.896061406666667

00:44:42.560 --> 00:44:45.885 So Pecoma is this very mysterious tumor.

NOTE Confidence: 0.896061406666667

00:44:45.890 --> 00:44:49.370 That has a very unusual definition.

NOTE Confidence: 0.896061406666667

00:44:49.370 --> 00:44:53.920 Tacomas are said to be composed of

NOTE Confidence: 0.896061406666667

00:44:53.920 --> 00:44:56.504 perivascular epithelioid cells which

NOTE Confidence: 0.896061406666667

00:44:56.504 --> 00:44:59.040 are distinctive epithelioid cells

NOTE Confidence: 0.896061406666667

00:44:59.040 --> 00:45:01.410 that are often closely associated

NOTE Confidence: 0.896061406666667

00:45:01.410 --> 00:45:04.080 with blood vessel walls and that

NOTE Confidence: 0.896061406666667

00:45:04.080 --> 00:45:06.360 often Co Express Millen, acidic,

NOTE Confidence: 0.896061406666667

00:45:06.360 --> 00:45:09.320 and smooth muscle markers.

NOTE Confidence: 0.896061406666667

00:45:09.320 --> 00:45:11.324 We think of Petco misses being

NOTE Confidence: 0.896061406666667

00:45:11.324 --> 00:45:13.515 a family of tumors that include

NOTE Confidence: 0.896061406666667

00:45:13.515 --> 00:45:15.849 the much more common and better

NOTE Confidence: 0.896061406666667

00:45:15.849 --> 00:45:17.180 known family member,

NOTE Confidence: 0.8706597

00:45:17.180 --> 00:45:17.688 Angiomyolipoma,

NOTE Confidence: 0.8706597

00:45:17.688 --> 00:45:20.736 which usually presents in the kidney.

NOTE Confidence: 0.8706597

00:45:20.740 --> 00:45:23.427 But this morning we talked about tumors

NOTE Confidence: 0.8706597

00:45:23.427 --> 00:45:26.836 in the liver that are also often

NOTE Confidence: 0.8706597

00:45:26.836 --> 00:45:28.880 called angiomyolipomas that are in

NOTE Confidence: 0.8706597

00:45:28.880 --> 00:45:31.718 fact just pecoma's of the of the liver.

NOTE Confidence: 0.8706597

00:45:31.720 --> 00:45:35.656 We have a rare rare disorder called Lam

NOTE Confidence: 0.8706597

00:45:35.656 --> 00:45:37.102 Lymphangioleiomyomatosis that mostly

NOTE Confidence: 0.8706597

00:45:37.102 --> 00:45:39.783 occurs in the lungs and lymph nodes.

NOTE Confidence: 0.8706597

00:45:39.790 --> 00:45:42.548 And the tumor we're talking about today.

NOTE Confidence: 0.8706597

00:45:42.550 --> 00:45:45.259 A group of distinctive tumors that have

NOTE Confidence: 0.8706597

00:45:45.259 --> 00:45:48.090 been called pecoma not otherwise specified,

NOTE Confidence: 0.8706597

00:45:48.090 --> 00:45:51.611 or simply pecoma All these tumors share

NOTE Confidence: 0.8706597

00:45:51.611 --> 00:45:55.197 this distinctive magical cell type that we

NOTE Confidence: 0.8706597

00:45:55.197 --> 00:45:57.687 call the perivascular epithelioid cell,

NOTE Confidence: 0.8706597

00:45:57.690 --> 00:46:00.510 which has no known normal

NOTE Confidence: 0.8706597

00:46:00.510 --> 00:46:01.638 cellular counterpart,

NOTE Confidence: 0.8706597

00:46:01.640 --> 00:46:04.688 so this isn't really a known cell type,

NOTE Confidence: 0.8706597

00:46:04.690 --> 00:46:06.811 but it's a concept that was invented

NOTE Confidence: 0.8706597

00:46:06.811 --> 00:46:09.209 by a group from Italy from Verona

NOTE Confidence: 0.8706597

00:46:09.210 --> 00:46:11.346 more than 20 years ago now,

NOTE Confidence: 0.8706597

00:46:11.350 --> 00:46:14.032 and this concept has stuck and

NOTE Confidence: 0.8706597

00:46:14.032 --> 00:46:17.599 we use it to describe pecoma.

NOTE Confidence: 0.8706597

00:46:17.600 --> 00:46:19.749 The first large series of soft tissue,

NOTE Confidence: 0.8706597

00:46:19.750 --> 00:46:20.532 Petco Miss,

NOTE Confidence: 0.8706597

00:46:20.532 --> 00:46:22.987 was published by Andrew Phillip Thomas,

NOTE Confidence: 0.8706597

00:46:22.987 --> 00:46:25.969 Mensal Sharon Weiss and colleagues and

NOTE Confidence: 0.8706597

00:46:25.970 --> 00:46:29.108 2005 when they first proposed criteria

NOTE Confidence: 0.8706597

00:46:29.108 --> 00:46:32.730 for malignancy based on the small series.

NOTE Confidence: 0.8706597

00:46:32.730 --> 00:46:35.026 We now know that Petco massacre at a

NOTE Confidence: 0.8706597

00:46:35.026 --> 00:46:37.127 very broad range of anatomic sites.

NOTE Confidence: 0.8706597

00:46:37.130 --> 00:46:39.650 You can see them primarily presenting

NOTE Confidence: 0.8706597

00:46:39.650 --> 00:46:41.930 is very small skin tumors.

NOTE Confidence: 0.8706597

00:46:41.930 --> 00:46:44.165 They are relatively common in

NOTE Confidence: 0.8706597

00:46:44.165 --> 00:46:45.506 the gastrointestinal tract.

NOTE Confidence: 0.8706597

00:46:45.510 --> 00:46:48.240 I promised the GI pathology faculty earlier

NOTE Confidence: 0.8706597

00:46:48.240 --> 00:46:51.509 today that I would allude to the GI tract,

NOTE Confidence: 0.8706597

00:46:51.510 --> 00:46:53.718 even though today I'm talking about

NOTE Confidence: 0.8706597

00:46:53.718 --> 00:46:56.210 soft tissue tumors of the head neck.

NOTE Confidence: 0.8706597

00:46:56.210 --> 00:46:59.410 Because I am also a GI pathologist.

NOTE Confidence: 0.8706597

00:46:59.410 --> 00:47:02.110 So pack owners are much more

NOTE Confidence: 0.8706597

00:47:02.110 --> 00:47:04.370 common in women than men.

NOTE Confidence: 0.8706597

00:47:04.370 --> 00:47:09.018 They usually present in adults and unlike

NOTE Confidence: 0.8706597

00:47:09.018 --> 00:47:12.210 angiomyolipoma and lymphangioleiomyomatosis,

NOTE Confidence: 0.8706597

00:47:12.210 --> 00:47:14.002 pecoma unspecified the tumors

NOTE Confidence: 0.8706597

00:47:14.002 --> 00:47:15.794 in the soft tissue.

NOTE Confidence: 0.8706597

00:47:15.800 --> 00:47:19.304 GI Tracting uterus are rarely associated

NOTE Confidence: 0.8706597

00:47:19.304 --> 00:47:22.470 with the cancer predisposition syndrome,

NOTE Confidence: 0.8706597

00:47:22.470 --> 00:47:26.160 TSC, the tubers sclerosis complex.

NOTE Confidence: 0.8706597

00:47:26.160 --> 00:47:28.379 You can find them in the extremities,

NOTE Confidence: 0.8706597

00:47:28.380 --> 00:47:29.564 trunk wall and skin,

NOTE Confidence: 0.8706597

00:47:29.564 --> 00:47:31.340 but those are a small minority

NOTE Confidence: 0.8706597

00:47:31.340 --> 00:47:32.432 of all pecoma's.

NOTE Confidence: 0.8706597

00:47:32.432 --> 00:47:35.527 Most of the time you'll find them in

NOTE Confidence: 0.8706597

00:47:35.527 --> 00:47:38.335 central body cavity or visceral locations.

NOTE Confidence: 0.8706597

00:47:38.340 --> 00:47:42.078 This is a nice resection that was

NOTE Confidence: 0.8706597

00:47:42.080 --> 00:47:44.360 performed in our hospital of a

NOTE Confidence: 0.8706597

00:47:44.360 --> 00:47:46.620 primary pecoma of the pancreas.

NOTE Confidence: 0.8706597

00:47:46.620 --> 00:47:48.580 This was such a nice gross photograph

NOTE Confidence: 0.8706597

00:47:48.580 --> 00:47:50.991 that I included in The Who the last

NOTE Confidence: 0.8706597

00:47:50.991 --> 00:47:53.770 two times the 4th and 5th series.

NOTE Confidence: 0.8706597

00:47:53.770 --> 00:47:55.612 It looks kind of similar to

NOTE Confidence: 0.8706597

00:47:55.612 --> 00:47:56.533 the pancreatic parenchyma.

NOTE Confidence: 0.8706597

00:47:56.540 --> 00:47:58.332 It's very sharply demarcated

NOTE Confidence: 0.8706597

00:47:58.332 --> 00:47:59.676 from the pancreas,

NOTE Confidence: 0.8706597

00:47:59.680 --> 00:48:02.792 but this is a pecoma that looks really

NOTE Confidence: 0.8706597

00:48:02.792 --> 00:48:06.196 similar to the angiomyolipoma of the liver.

NOTE Confidence: 0.8706597

00:48:06.200 --> 00:48:08.275 They were reviewed in the

NOTE Confidence: 0.8706597

00:48:08.275 --> 00:48:09.935 slide seminar this morning.

NOTE Confidence: 0.8706597

00:48:09.940 --> 00:48:12.460 And here you can see a very

NOTE Confidence: 0.8706597

00:48:12.460 --> 00:48:13.604 interesting accentuation around

NOTE Confidence: 0.8706597

00:48:13.604 --> 00:48:15.956 the blood vessel in the middle.

NOTE Confidence: 0.8706597

00:48:15.960 --> 00:48:18.372 We have these blood vessels that

NOTE Confidence: 0.8706597

00:48:18.372 --> 00:48:21.735 seem to have the tumor cells just

NOTE Confidence: 0.8706597

00:48:21.735 --> 00:48:24.930 underneath the endothelial lining.

NOTE Confidence: 0.8706597

00:48:24.930 --> 00:48:28.068 Sometimes these tumors have very striking,

NOTE Confidence: 0.8706597

00:48:28.070 --> 00:48:30.320 clear cell morphology and a

NOTE Confidence: 0.8706597

00:48:30.320 --> 00:48:31.670 beautiful nested appearance,

NOTE Confidence: 0.8706597

00:48:31.670 --> 00:48:33.870 making them almost indistinguishable

NOTE Confidence: 0.8706597

00:48:33.870 --> 00:48:37.170 from clear cell renal cell carcinoma.

NOTE Confidence: 0.8706597

00:48:37.170 --> 00:48:39.070 Some of them have trabeculae

NOTE Confidence: 0.8706597

00:48:39.070 --> 00:48:39.830 are architecture.

NOTE Confidence: 0.8706597

00:48:39.830 --> 00:48:44.030 This example also arose in the nasal cavity

NOTE Confidence: 0.8706597

00:48:44.030 --> 00:48:48.108 and then also had a TFE 3 gene fusion.

NOTE Confidence: 0.8706597

00:48:48.110 --> 00:48:51.335 Other examples look much more

NOTE Confidence: 0.8706597

00:48:51.335 --> 00:48:53.606 similar to leiomyosarcoma's with

NOTE Confidence: 0.8706597

00:48:53.606 --> 00:48:55.558 fascicular spindle cell morphology,

NOTE Confidence: 0.8706597

00:48:55.560 --> 00:48:58.800 but in contrast to true smooth muscle tumors,

NOTE Confidence: 0.873522042857143

00:48:58.800 --> 00:49:01.148 which typically show very

NOTE Confidence: 0.873522042857143

00:49:01.148 --> 00:49:02.909 dense core cytoplasm.

NOTE Confidence: 0.873522042857143

00:49:02.910 --> 00:49:05.260 The cytoplasmic quality of pecoma,

NOTE Confidence: 0.873522042857143

00:49:05.260 --> 00:49:08.386 as is almost always this delicate

NOTE Confidence: 0.873522042857143

00:49:08.390 --> 00:49:12.860 granular eosinophilic to clear cytoplasm.

NOTE Confidence: 0.873522042857143

00:49:12.860 --> 00:49:15.752 There is a distinctive sclerosing variant

NOTE Confidence: 0.873522042857143

00:49:15.752 --> 00:49:20.040 of pecoma that has a marked predilection

NOTE Confidence: 0.873522042857143

00:49:20.040 --> 00:49:22.696 for the pararenal retroperitoneum.

NOTE Confidence: 0.873522042857143

00:49:22.700 --> 00:49:26.600 As you see in these two images on this slide,

NOTE Confidence: 0.873522042857143

00:49:26.600 --> 00:49:29.102 we again have that orientation under

NOTE Confidence: 0.873522042857143

00:49:29.102 --> 00:49:31.759 the endothelium of the blood vessel,

NOTE Confidence: 0.873522042857143

00:49:31.760 --> 00:49:33.785 and these cords of somewhat

NOTE Confidence: 0.873522042857143

00:49:33.785 --> 00:49:36.576 clear cells within a very dense

NOTE Confidence: 0.873522042857143

00:49:36.576 --> 00:49:38.640 hyalinized collagenous stroma.

NOTE Confidence: 0.873522042857143

00:49:38.640 --> 00:49:41.094 As I mentioned, we define these

NOTE Confidence: 0.873522042857143

00:49:41.094 --> 00:49:43.828 tumors as showing a mixed Milan,

NOTE Confidence: 0.873522042857143

00:49:43.828 --> 00:49:46.260 acidic and myogenic phenotype.

NOTE Confidence: 0.873522042857143

00:49:46.260 --> 00:49:49.754 They're nearly always positive for HMB 45.

NOTE Confidence: 0.873522042857143

00:49:49.754 --> 00:49:52.196 That's the best marker for pecoma.

NOTE Confidence: 0.873522042857143

00:49:52.200 --> 00:49:54.324 Smooth muscle actin is the most

NOTE Confidence: 0.873522042857143

00:49:54.324 --> 00:49:56.320 sensitive of the muscle markers.

NOTE Confidence: 0.873522042857143

00:49:56.320 --> 00:49:56.760 However,

NOTE Confidence: 0.873522042857143

00:49:56.760 --> 00:49:59.840 it's important to be aware of the

NOTE Confidence: 0.873522042857143

00:49:59.840 --> 00:50:03.185 fact that in particular the clear cell

NOTE Confidence: 0.873522042857143

00:50:03.185 --> 00:50:06.459 examples that harbor TFE 3 fusions

NOTE Confidence: 0.873522042857143

00:50:06.459 --> 00:50:09.327 are sometimes entirely negative.

NOTE Confidence: 0.873522042857143

00:50:09.330 --> 00:50:12.528 For smooth muscle actin and desmin.

NOTE Confidence: 0.873522042857143

00:50:12.530 --> 00:50:14.833 You can find focal S 100 in

NOTE Confidence: 0.873522042857143

00:50:14.833 --> 00:50:16.410 a subset of cases,

NOTE Confidence: 0.873522042857143

00:50:16.410 --> 00:50:18.934 but it's usually unimpressive

NOTE Confidence: 0.873522042857143

00:50:18.934 --> 00:50:20.827 and predominantly cytoplasmic,

NOTE Confidence: 0.873522042857143

00:50:20.830 --> 00:50:23.014 which helps distinguish them

NOTE Confidence: 0.873522042857143

00:50:23.014 --> 00:50:25.744 in a phenotype from Melanoma.

NOTE Confidence: 0.873522042857143

00:50:25.750 --> 00:50:26.404 In addition,

NOTE Confidence: 0.873522042857143

00:50:26.404 --> 00:50:29.020 socks 10 that marker we think of as

NOTE Confidence: 0.873522042857143

00:50:29.093 --> 00:50:31.487 being very strongly expressed in close

NOTE Confidence: 0.873522042857143

00:50:31.487 --> 00:50:35.796 to 100% of melanomas is negative and pecoma.

NOTE Confidence: 0.873522042857143

00:50:35.800 --> 00:50:38.824 And in correlating with the presence

NOTE Confidence: 0.873522042857143

00:50:38.824 --> 00:50:42.280 of the gene fusions TFE 3 protein

NOTE Confidence: 0.873522042857143

00:50:42.280 --> 00:50:44.980 is expressed at high levels in

NOTE Confidence: 0.873522042857143

00:50:44.980 --> 00:50:46.969 about 10 to 15% of cases.

NOTE Confidence: 0.873522042857143

00:50:46.969 --> 00:50:49.152 And that's a good surrogate by

NOTE Confidence: 0.873522042857143

00:50:49.152 --> 00:50:51.200 administer chemistry for the

NOTE Confidence: 0.873522042857143

00:50:51.200 --> 00:50:53.760 presence of the gene rearrangement.

NOTE Confidence: 0.873522042857143

00:50:53.760 --> 00:50:54.852 This is a,

NOTE Confidence: 0.873522042857143

00:50:54.852 --> 00:50:55.216 uh,

NOTE Confidence: 0.873522042857143

00:50:55.216 --> 00:50:57.036 some examples of smooth muscle

NOTE Confidence: 0.873522042857143

00:50:57.036 --> 00:50:58.900 actin and desmin staining.

NOTE Confidence: 0.873522042857143

00:50:58.900 --> 00:51:00.406 This was actually from the case

NOTE Confidence: 0.873522042857143

00:51:00.406 --> 00:51:01.159 in the pancreas.

NOTE Confidence: 0.873522042857143

00:51:01.160 --> 00:51:03.337 I showed you a few minutes ago

NOTE Confidence: 0.873522042857143

00:51:03.340 --> 00:51:05.830 and these were are nice examples

NOTE Confidence: 0.873522042857143

00:51:05.830 --> 00:51:08.050 of the variability of staining

NOTE Confidence: 0.873522042857143

00:51:08.050 --> 00:51:10.475 for HMB 5:45 and Melanie.

NOTE Confidence: 0.873522042857143

00:51:10.480 --> 00:51:11.518 And in fact,

NOTE Confidence: 0.873522042857143

00:51:11.518 --> 00:51:13.594 in order to support the diagnosis,

NOTE Confidence: 0.873522042857143

00:51:13.600 --> 00:51:16.570 I usually run all four markers

NOTE Confidence: 0.873522042857143

00:51:16.570 --> 00:51:18.055 SMAD and Desmond.

NOTE Confidence: 0.873522042857143

00:51:18.060 --> 00:51:20.652 HMB 45 MLN A and if you do that,

NOTE Confidence: 0.873522042857143

00:51:20.660 --> 00:51:21.640 usually you'll get a hit,

NOTE Confidence: 0.873522042857143

00:51:21.640 --> 00:51:24.016 at least with one of the muscle markers,

NOTE Confidence: 0.873522042857143

00:51:24.020 --> 00:51:26.778 and one of them will anesthetic markers.

NOTE Confidence: 0.873522042857143

00:51:26.780 --> 00:51:29.186 We don't really have very good

NOTE Confidence: 0.873522042857143

00:51:29.186 --> 00:51:31.360 criteria for malignancy in pecoma.

NOTE Confidence: 0.873522042857143

00:51:31.360 --> 00:51:34.600 As I mentioned Andrew folks first

NOTE Confidence: 0.873522042857143

00:51:34.600 --> 00:51:36.328 suggested criteria but we don't

NOTE Confidence: 0.873522042857143

00:51:36.328 --> 00:51:38.070 have a lot of cases to go on.

NOTE Confidence: 0.873522042857143

00:51:38.070 --> 00:51:40.788 We know that features associated with

NOTE Confidence: 0.873522042857143

00:51:40.788 --> 00:51:43.380 malignant behavior include large tumor size,

NOTE Confidence: 0.873522042857143

00:51:43.380 --> 00:51:47.240 mitotic activity and atypia pleomorphism.

NOTE Confidence: 0.873522042857143

00:51:47.240 --> 00:51:49.662 So in my practice I really use

NOTE Confidence: 0.873522042857143

00:51:49.662 --> 00:51:51.257 the combination of essentially

NOTE Confidence: 0.873522042857143

00:51:51.257 --> 00:51:54.042 any mitotic activity with atypia

NOTE Confidence: 0.873522042857143

00:51:54.042 --> 00:51:57.110 pleomorphism the support the diagnosis.

NOTE Confidence: 0.873522042857143

00:51:57.110 --> 00:51:59.300 Of malignancy this is a malignant

NOTE Confidence: 0.873522042857143

00:51:59.300 --> 00:52:00.760 pecoma of the colon,

NOTE Confidence: 0.873522042857143

00:52:00.760 --> 00:52:04.780 a large fleshy sarcoma like lesion.

NOTE Confidence: 0.873522042857143

00:52:04.780 --> 00:52:07.624 These tumors can look very similar

NOTE Confidence: 0.873522042857143

00:52:07.624 --> 00:52:09.046 to metastatic Melanoma.

NOTE Confidence: 0.873522042857143

00:52:09.050 --> 00:52:12.560 With macro nucleoli striking pleomorphism

NOTE Confidence: 0.873522042857143

00:52:12.560 --> 00:52:16.070 and eosinophilic or amphiphilic cytoplasm,

NOTE Confidence: 0.873522042857143

00:52:16.070 --> 00:52:17.883 whereas in other cases they have much

NOTE Confidence: 0.873522042857143

00:52:17.883 --> 00:52:19.939 more of a clear cell appearance,

NOTE Confidence: 0.873522042857143

00:52:19.940 --> 00:52:22.710 looking very similar to metastatic

NOTE Confidence: 0.873522042857143

00:52:22.710 --> 00:52:25.030 clear cell renal cell carcinoma,

NOTE Confidence: 0.873522042857143

00:52:25.030 --> 00:52:29.206 this is obviously a metastasis to the liver.

NOTE Confidence: 0.873522042857143

00:52:29.210 --> 00:52:31.790 We again see those macronuclei,

NOTE Confidence: 0.873522042857143

00:52:31.790 --> 00:52:33.146 plia, morphic,

NOTE Confidence: 0.873522042857143

00:52:33.146 --> 00:52:35.858 and multinucleated tumor cells.

NOTE Confidence: 0.873522042857143

00:52:35.860 --> 00:52:38.370 Pete Argani from Hopkins with

NOTE Confidence: 0.873522042857143

00:52:38.370 --> 00:52:40.378 Sharon Weiss first presented

NOTE Confidence: 0.873522042857143

00:52:40.378 --> 00:52:43.299 this series of tumors with TFE 3

NOTE Confidence: 0.873522042857143

00:52:43.299 --> 00:52:45.739 gene fusions about 10 years ago.

NOTE Confidence: 0.873522042857143

00:52:45.740 --> 00:52:46.772 This was the case.

NOTE Confidence: 0.873522042857143

00:52:46.772 --> 00:52:48.320 I mentioned that I had just

NOTE Confidence: 0.94284840125

00:52:48.380 --> 00:52:51.418 a couple of years ago from the sinus that

NOTE Confidence: 0.94284840125

00:52:51.418 --> 00:52:54.574 trabeculae are tumor with fairly clear

NOTE Confidence: 0.94284840125

00:52:54.574 --> 00:52:57.609 cytoplasm that had a TFE 3 gene fusion.

NOTE Confidence: 0.94284840125

00:52:57.610 --> 00:53:01.154 A and RC Agram and Christina TSQ published

NOTE Confidence: 0.94284840125

00:53:01.154 --> 00:53:04.420 this beautiful paper six years ago in the

NOTE Confidence: 0.94284840125

00:53:04.420 --> 00:53:06.470 American Journal of Surgical Pathology,

NOTE Confidence: 0.94284840125

00:53:06.470 --> 00:53:08.680 highlighting what we understand about

NOTE Confidence: 0.94284840125

00:53:08.680 --> 00:53:11.347 the molecular genetics of Pecoma TSC.

NOTE Confidence: 0.94284840125

00:53:11.347 --> 00:53:15.003 2 mutations are found in the majority of

NOTE Confidence: 0.94284840125

00:53:15.003 --> 00:53:18.449 cases and we also have trends locations in

NOTE Confidence: 0.94284840125

00:53:18.449 --> 00:53:23.015 a range of of pecoma TFE 3 being the most

NOTE Confidence: 0.94284840125

00:53:23.015 --> 00:53:27.518 common gene involved in translocations.

NOTE Confidence: 0.94284840125

00:53:27.520 --> 00:53:31.516 Because of the alterations of TSC 2 in the

NOTE Confidence: 0.94284840125

00:53:31.516 --> 00:53:34.291 majority of Tacomas and visceral locations,

NOTE Confidence: 0.94284840125

00:53:34.291 --> 00:53:36.119 including in tumors that

NOTE Confidence: 0.94284840125

00:53:36.119 --> 00:53:38.020 pursue an aggressive course.

NOTE Confidence: 0.94284840125

00:53:38.020 --> 00:53:41.722 The malignant examples we can use

NOTE Confidence: 0.94284840125

00:53:41.722 --> 00:53:44.874 mtor inhibitors to effectively treat

NOTE Confidence: 0.94284840125

00:53:44.874 --> 00:53:47.769 patients with this tumor type.

NOTE Confidence: 0.94284840125

00:53:47.770 --> 00:53:49.947 These are some sort of case reports

NOTE Confidence: 0.94284840125

00:53:49.947 --> 00:53:51.622 and small series published almost

NOTE Confidence: 0.94284840125

00:53:51.622 --> 00:53:54.204 ten years ago now showing that M

NOTE Confidence: 0.94284840125

00:53:54.204 --> 00:53:56.244 Tor inhibitors such as Sorolla,

NOTE Confidence: 0.94284840125

00:53:56.250 --> 00:53:58.974 Miss or quite effective in treating

NOTE Confidence: 0.94284840125

00:53:58.974 --> 00:54:00.711 patients with malignant pecoma's.

NOTE Confidence: 0.94284840125

00:54:00.711 --> 00:54:03.679 This is an example of a patient treated

NOTE Confidence: 0.94284840125

00:54:03.679 --> 00:54:06.430 in our hospital at the Dana Farber Cancer

NOTE Confidence: 0.94284840125

00:54:06.430 --> 00:54:08.300 Institute by my colleague Andy Wagner,

NOTE Confidence: 0.94284840125

00:54:08.300 --> 00:54:10.841 who's he's really the world expert in

NOTE Confidence: 0.94284840125

00:54:10.841 --> 00:54:13.029 treating patients with malignant pecoma.

NOTE Confidence: 0.94284840125

00:54:13.030 --> 00:54:15.200 He's treated on the order of about

NOTE Confidence: 0.94284840125

00:54:15.200 --> 00:54:17.290 70 patients with this sarcoma type.

NOTE Confidence: 0.94284840125

00:54:17.290 --> 00:54:19.159 Over the last ten years or so,

NOTE Confidence: 0.94284840125

00:54:19.160 --> 00:54:21.855 and you can see this very dramatic

NOTE Confidence: 0.94284840125

00:54:21.855 --> 00:54:23.894 clinical benefit to serralles therapy

NOTE Confidence: 0.94284840125

00:54:23.894 --> 00:54:26.736 with a marked decrease in size of

NOTE Confidence: 0.94284840125

00:54:26.736 --> 00:54:28.715 these multiple metastatic lesions

NOTE Confidence: 0.94284840125

00:54:28.715 --> 00:54:31.709 in the abdominal cavity and pelvis.

NOTE Confidence: 0.94284840125

00:54:31.710 --> 00:54:32.326 Very recently,

NOTE Confidence: 0.94284840125

00:54:32.326 --> 00:54:34.482 this is a paper that was just

NOTE Confidence: 0.94284840125

00:54:34.482 --> 00:54:36.170 published online a few weeks ago.

NOTE Confidence: 0.94284840125

00:54:36.170 --> 00:54:39.158 There is now a modified albumin

NOTE Confidence: 0.94284840125

00:54:39.158 --> 00:54:41.150 conjugated version of sirolimus

NOTE Confidence: 0.94284840125

00:54:41.230 --> 00:54:44.266 that is also really effective in

NOTE Confidence: 0.94284840125

00:54:44.266 --> 00:54:46.290 patients with malignant pecoma,

NOTE Confidence: 0.94284840125

00:54:46.290 --> 00:54:49.590 especially patients that have TSC 2

NOTE Confidence: 0.94284840125

00:54:49.590 --> 00:54:52.090 mutations thus far in this series,

NOTE Confidence: 0.94284840125

00:54:52.090 --> 00:54:54.110 all the patients whose tumors

NOTE Confidence: 0.94284840125

00:54:54.110 --> 00:54:56.130 were confirmed to harbor TSC.

NOTE Confidence: 0.94284840125

00:54:56.130 --> 00:54:59.268 2 alterations were are alive either

NOTE Confidence: 0.94284840125

00:54:59.268 --> 00:55:02.160 without disease or with disease.

NOTE Confidence: 0.94284840125

00:55:02.160 --> 00:55:03.875 And you can see from that plot

NOTE Confidence: 0.94284840125

00:55:03.875 --> 00:55:04.960 in the lower left,

NOTE Confidence: 0.94284840125

00:55:04.960 --> 00:55:07.714 the striking clinical benefit in the

NOTE Confidence: 0.94284840125

00:55:07.714 --> 00:55:10.745 majority of patients with malignant pecoma

NOTE Confidence: 0.94284840125

00:55:10.745 --> 00:55:13.545 treated with this seromas conjugate.

NOTE Confidence: 0.915525006

00:55:15.650 --> 00:55:18.050 So for my final slide of

NOTE Confidence: 0.915525006

00:55:18.050 --> 00:55:19.650 summary of practice points,

NOTE Confidence: 0.915525006

00:55:19.650 --> 00:55:22.086 don't forget that head and neck location

NOTE Confidence: 0.915525006

00:55:22.086 --> 00:55:24.632 is very common for both alveolar

NOTE Confidence: 0.915525006

00:55:24.632 --> 00:55:26.548 Rhabdomyosarcoma and spindle cells.

NOTE Confidence: 0.915525006

00:55:26.550 --> 00:55:29.270 Grossing revenue miles sarcomas.

NOTE Confidence: 0.915525006

00:55:29.270 --> 00:55:31.701 Beware of keratin and

NOTE Confidence: 0.915525006

00:55:31.701 --> 00:55:32.874 neuroendocrine marker expression,

NOTE Confidence: 0.915525006

00:55:32.874 --> 00:55:37.155 so you don't go down the tubes and make a

NOTE Confidence: 0.915525006

00:55:37.155 --> 00:55:39.019 misdiagnosis of alveolar Rhabdomyosarcoma.

NOTE Confidence: 0.915525006

00:55:39.020 --> 00:55:41.960 I've now shown you lots of examples

NOTE Confidence: 0.915525006

00:55:41.960 --> 00:55:43.850 of biphenotypic sinonasal sarcoma.

NOTE Confidence: 0.915525006

00:55:43.850 --> 00:55:46.222 This distinctive pack three

NOTE Confidence: 0.915525006

00:55:46.222 --> 00:55:48.001 translocation associated low

NOTE Confidence: 0.915525006

00:55:48.001 --> 00:55:50.890 grade sarcoma that Co expresses.

NOTE Confidence: 0.915525006

00:55:50.890 --> 00:55:53.370 Muscle markers and neural markers

NOTE Confidence: 0.915525006

00:55:53.370 --> 00:55:57.060 and finally Pecoma Harbor either TSC,

NOTE Confidence: 0.915525006

00:55:57.060 --> 00:56:02.046 2 deletions, or TFE 3 fusions.

NOTE Confidence: 0.915525006

00:56:02.050 --> 00:56:03.914 So I'm going to close there and I

NOTE Confidence: 0.915525006

00:56:03.914 --> 00:56:06.309 want to thank you very much to the

NOTE Confidence: 0.915525006

00:56:06.309 --> 00:56:07.952 department into Manju Prasad for

NOTE Confidence: 0.915525006

00:56:07.952 --> 00:56:10.283 inviting me to visit your department today.

NOTE Confidence: 0.915525006

00:56:10.290 --> 00:56:12.460 I'm sorry that it had to be

NOTE Confidence: 0.915525006

00:56:12.460 --> 00:56:14.510 converted to a virtual visit,

NOTE Confidence: 0.915525006

00:56:14.510 --> 00:56:16.561 but I always have to now advertise

NOTE Confidence: 0.915525006

00:56:16.561 --> 00:56:18.720 for my rock band the teardowns.

NOTE Confidence: 0.915525006

00:56:18.720 --> 00:56:19.719 As you heard,

NOTE Confidence: 0.915525006

00:56:19.719 --> 00:56:21.717 we released our first album that's

NOTE Confidence: 0.915525006

00:56:21.717 --> 00:56:23.548 available on all streaming services

NOTE Confidence: 0.915525006

00:56:23.550 --> 00:56:25.468 and after a very long delay we're

NOTE Confidence: 0.915525006

00:56:25.468 --> 00:56:26.947 beginning to play some shows

NOTE Confidence: 0.915525006

00:56:26.947 --> 00:56:28.936 around town where we're playing in

NOTE Confidence: 0.915525006

00:56:28.936 --> 00:56:31.082 Summerville coming up next month and

NOTE Confidence: 0.915525006

00:56:31.082 --> 00:56:33.074 then in Jamaica Plain in Boston.

NOTE Confidence: 0.915525006

00:56:33.080 --> 00:56:33.650 In January,

NOTE Confidence: 0.915525006

00:56:33.650 --> 00:56:35.645 I doubt that we're ever going to

NOTE Confidence: 0.915525006

00:56:35.645 --> 00:56:37.499 travel outside of the Boston area.

NOTE Confidence: 0.915525006

00:56:37.500 --> 00:56:39.540 Since this is a hobby for all of

NOTE Confidence: 0.915525006

00:56:39.540 --> 00:56:41.619 us who have different careers,

NOTE Confidence: 0.915525006

00:56:41.620 --> 00:56:42.500 and honestly,

NOTE Confidence: 0.915525006

00:56:42.500 --> 00:56:45.140 we're not good enough to make

NOTE Confidence: 0.915525006

00:56:45.140 --> 00:56:47.967 this our our our full time job.

NOTE Confidence: 0.915525006

00:56:47.970 --> 00:56:49.608 And I'm happy to answer any questions,

NOTE Confidence: 0.915525006

00:56:49.610 --> 00:56:50.020 thank you.

NOTE Confidence: 0.92153966

00:56:57.380 --> 00:56:59.581 That was wonderful, Jason.

NOTE Confidence: 0.92153966

00:56:59.581 --> 00:57:03.223 I'm sure you're good enough to.

NOTE Confidence: 0.743524154444444

00:57:03.230 --> 00:57:06.674 Play for us at one of

NOTE Confidence: 0.743524154444444

00:57:06.674 --> 00:57:08.396 our departmental events.

NOTE Confidence: 0.743524154444444

00:57:08.400 --> 00:57:10.700 Uhm, there was a comment.

NOTE Confidence: 0.743524154444444

00:57:10.700 --> 00:57:15.368 My computer crashed.

NOTE Confidence: 0.743524154444444

00:57:15.370 --> 00:57:17.566 All the way through your presentation.

NOTE Confidence: 0.743524154444444

00:57:17.570 --> 00:57:21.907 So I lost all the comments in chat and

NOTE Confidence: 0.743524154444444

00:57:21.907 --> 00:57:24.683 I know that singing Pan had a comment,

NOTE Confidence: 0.743524154444444

00:57:24.690 --> 00:57:29.954 so singing can you unmute yourself and?

NOTE Confidence: 0.743524154444444

00:57:29.960 --> 00:57:34.680 And say your comments.

NOTE Confidence: 0.743524154444444

00:57:34.680 --> 00:57:38.772 I think it was about alveolar

NOTE Confidence: 0.743524154444444

00:57:38.772 --> 00:57:42.230 Rhabdomyosarcoma metastatic to lymph nodes.

NOTE Confidence: 0.743524154444444

00:57:42.230 --> 00:57:44.210 Can you hear me yes?

NOTE Confidence: 0.743524154444444

00:57:44.210 --> 00:57:45.770 Hi, so thank you for the

NOTE Confidence: 0.743524154444444

00:57:45.770 --> 00:57:47.390 wonderful presentation. So

NOTE Confidence: 0.85823402

00:57:47.400 --> 00:57:49.936 I just want to make a quick comment

NOTE Confidence: 0.85823402

00:57:49.940 --> 00:57:52.358 so I haven't seen. Like if you cases

NOTE Confidence: 0.60355163

00:57:52.370 --> 00:57:55.650 of metastatic tremors. Psycho Martin.

NOTE Confidence: 0.60355163

00:57:55.650 --> 00:57:59.238 If notes showing us purely sinusoidal

NOTE Confidence: 0.60355163

00:57:59.238 --> 00:58:01.910 pattern with polymer cells.

NOTE Confidence: 0.60355163

00:58:01.910 --> 00:58:03.198 And the tumor cells

NOTE Confidence: 0.717474528333333

00:58:04.240 --> 00:58:06.900 he feels in a positive for CD56 and

NOTE Confidence: 0.664428318

00:58:06.910 --> 00:58:10.300 AWK. So one case was misdiagnosed

NOTE Confidence: 0.664428318

00:58:10.300 --> 00:58:13.630 as a hog positive and approximately

NOTE Confidence: 0.664428318

00:58:13.630 --> 00:58:16.720 70 former by our third pathologists.

NOTE Confidence: 0.664428318

00:58:16.720 --> 00:58:19.348 So just beware.

NOTE Confidence: 0.664428318

00:58:19.350 --> 00:58:21.813 Both types of randomized sarcoma

NOTE Confidence: 0.664428318

00:58:21.813 --> 00:58:25.540 may have expression that thank you.

NOTE Confidence: 0.86485611875

00:58:25.930 --> 00:58:27.746 No thank you. That's that's a great point.

NOTE Confidence: 0.86485611875

00:58:27.750 --> 00:58:29.946 I think alveolar Rhabdomyosarcoma is a

NOTE Confidence: 0.86485611875

00:58:29.946 --> 00:58:32.225 very good mimicker of many different

NOTE Confidence: 0.86485611875

00:58:32.225 --> 00:58:34.498 tumor types because of these very

NOTE Confidence: 0.86485611875

00:58:34.498 --> 00:58:36.090 strange appearances of expression

NOTE Confidence: 0.86485611875

00:58:36.090 --> 00:58:38.710 and alk is quite commonly positive,

NOTE Confidence: 0.86485611875

00:58:38.710 --> 00:58:40.850 and that's a very interesting.

NOTE Confidence: 0.86485611875

00:58:40.850 --> 00:58:41.910 Interesting problem,

NOTE Confidence: 0.86485611875

00:58:41.910 --> 00:58:45.220 thank you and all 56 as well. So yes.

NOTE Confidence: 0.646748133333333

00:58:47.300 --> 00:58:51.580 Oh, I always wondered why we had both

NOTE Confidence: 0.646748133333333

00:58:51.580 --> 00:58:56.306 my D1 and myogenin so clearly it has.

NOTE Confidence: 0.646748133333333

00:58:56.306 --> 00:59:00.006 Prove to be useful both these stains

NOTE Confidence: 0.646748133333333

00:59:00.006 --> 00:59:03.914 and why both might should be or might

NOTE Confidence: 0.646748133333333

00:59:03.914 --> 00:59:06.690 be ordered and can be useful. Yeah,

NOTE Confidence: 0.927304191428571

00:59:06.700 --> 00:59:08.135 I think that's a very good point,

NOTE Confidence: 0.927304191428571

00:59:08.140 --> 00:59:10.816 and in fact, before we recognize

NOTE Confidence: 0.927304191428571

00:59:10.816 --> 00:59:13.120 spindle cell and sclerostin rabdo,

NOTE Confidence: 0.927304191428571

00:59:13.120 --> 00:59:14.392 I don't think we really needed

NOTE Confidence: 0.927304191428571

00:59:14.392 --> 00:59:17.304 my OD one and for many years the

NOTE Confidence: 0.927304191428571

00:59:17.304 --> 00:59:18.934 antibodies available were pretty bad.

NOTE Confidence: 0.927304191428571

00:59:18.940 --> 00:59:20.548 They often showed a lot of

NOTE Confidence: 0.927304191428571

00:59:20.548 --> 00:59:21.352 subtle plasmic staining.

NOTE Confidence: 0.927304191428571

00:59:21.360 --> 00:59:22.998 The new clones are much better.

NOTE Confidence: 0.927304191428571

00:59:23.000 --> 00:59:24.850 They actually worked beautifully and

NOTE Confidence: 0.927304191428571

00:59:24.850 --> 00:59:27.250 they're really helpful for this class of.

NOTE Confidence: 0.927304191428571

00:59:27.250 --> 00:59:28.960 Of Rhabdomyosarcoma.

NOTE Confidence: 0.663590046470588

00:59:29.710 --> 00:59:33.715 Yeah, and I brought my my journey into our

NOTE Confidence: 0.663590046470588

00:59:33.715 --> 00:59:37.836 lab and I stopped using my D1 altogether,

NOTE Confidence: 0.663590046470588

00:59:37.840 --> 00:59:42.943 but clearly we'll have to look at our clones.

NOTE Confidence: 0.663590046470588

00:59:42.950 --> 00:59:45.351 The other thing I wanted to ask

NOTE Confidence: 0.663590046470588

00:59:45.351 --> 00:59:47.990 Jason is many of these antibodies.

NOTE Confidence: 0.663590046470588

00:59:47.990 --> 00:59:51.450 This newer antibodies that affusion

NOTE Confidence: 0.663590046470588

00:59:51.450 --> 00:59:54.544 specific or gene aberration specific.

NOTE Confidence: 0.663590046470588

00:59:54.544 --> 00:59:58.520 Do you offer them as test only

NOTE Confidence: 0.663590046470588

00:59:58.624 --> 01:00:01.198 like like the other labs?

NOTE Confidence: 0.922619525625

01:00:01.290 --> 01:00:02.506 Yeah, unfortunately we don't

NOTE Confidence: 0.922619525625

01:00:02.506 --> 01:00:04.330 because we just don't have the

NOTE Confidence: 0.922619525625

01:00:04.389 --> 01:00:05.985 staffing to be a reference lab.

NOTE Confidence: 0.922619525625

01:00:05.990 --> 01:00:08.890 At this point we can barely keep

NOTE Confidence: 0.922619525625

01:00:08.890 --> 01:00:10.390 up with our volume in House.

NOTE Confidence: 0.922619525625

01:00:10.390 --> 01:00:13.310 We we have about.

NOTE Confidence: 0.922619525625

01:00:13.310 --> 01:00:16.982 650 to 700 a day not counting him path,

NOTE Confidence: 0.922619525625

01:00:16.990 --> 01:00:19.468 so it's just it's it's impossible,

NOTE Confidence: 0.922619525625

01:00:19.470 --> 01:00:21.734 so we for now we're only doing it

NOTE Confidence: 0.922619525625

01:00:21.734 --> 01:00:23.374 as part of diagnostic consults.

NOTE Confidence: 0.922619525625

01:00:23.374 --> 01:00:26.398 I think in a couple of years

NOTE Confidence: 0.922619525625

01:00:26.400 --> 01:00:28.689 some of you might have heard our

NOTE Confidence: 0.922619525625

01:00:28.689 --> 01:00:30.227 institutions are probably going to

NOTE Confidence: 0.922619525625

01:00:30.227 --> 01:00:32.211 be forming a central lab for all of

NOTE Confidence: 0.922619525625

01:00:32.271 --> 01:00:34.186 the mass General Brigham Hospital.

NOTE Confidence: 0.922619525625

01:00:34.190 --> 01:00:36.556 So at that point, maybe will do.

NOTE Confidence: 0.922619525625

01:00:36.560 --> 01:00:37.409 Reference lab work,

NOTE Confidence: 0.922619525625

01:00:37.409 --> 01:00:39.107 but at this point we can't.

NOTE Confidence: 0.94822818

01:00:40.510 --> 01:00:43.702 Thank you. Any other

NOTE Confidence: 0.94822818

01:00:43.702 --> 01:00:45.806 questions from anyone else?

NOTE Confidence: 0.867772310625

01:00:45.820 --> 01:00:47.764 I see that I see Don Pot had a

NOTE Confidence: 0.867772310625

01:00:47.764 --> 01:00:49.528 comment that I'm a very good cook.

NOTE Confidence: 0.867772310625

01:00:49.530 --> 01:00:50.874 If I want to thank you for that.

NOTE Confidence: 0.867772310625

01:00:50.880 --> 01:00:52.824 I I only really started cooking

NOTE Confidence: 0.867772310625

01:00:52.824 --> 01:00:55.218 full force after we had started the

NOTE Confidence: 0.867772310625

01:00:55.218 --> 01:00:57.163 lock down with the pandemic and

NOTE Confidence: 0.867772310625

01:00:57.163 --> 01:00:58.794 now I'm really obsessed with it and

NOTE Confidence: 0.867772310625

01:00:58.794 --> 01:01:00.176 I love taking photographs during

NOTE Confidence: 0.867772310625

01:01:00.176 --> 01:01:02.192 the process as dog pot as seen.

NOTE Confidence: 0.867772310625

01:01:02.200 --> 01:01:05.637 So it's a it's a new hobby and I I love

NOTE Confidence: 0.867772310625

01:01:05.637 --> 01:01:08.104 cooking very different ethnic groups.

NOTE Confidence: 0.867772310625

01:01:08.104 --> 01:01:11.254 I love like Thai and and and

NOTE Confidence: 0.867772310625

01:01:11.254 --> 01:01:12.989 various other types of food.

NOTE Confidence: 0.867772310625

01:01:12.990 --> 01:01:16.122 And dump, as other question had to do with.

NOTE Confidence: 0.867772310625

01:01:16.130 --> 01:01:18.066 Fusion detection we haven't.

NOTE Confidence: 0.867772310625

01:01:18.066 --> 01:01:22.139 We haven't brought on one of the the RNA

NOTE Confidence: 0.867772310625

01:01:22.139 --> 01:01:26.287 based NGS fusion panels yet in our lab.

NOTE Confidence: 0.867772310625

01:01:26.290 --> 01:01:29.566 We have really only been reserving

NOTE Confidence: 0.867772310625

01:01:29.570 --> 01:01:30.982 molecular confirmation for the

NOTE Confidence: 0.867772310625

01:01:30.982 --> 01:01:32.747 cases that we struggle with,

NOTE Confidence: 0.867772310625

01:01:32.750 --> 01:01:35.046 which because we have so many antibodies

NOTE Confidence: 0.867772310625

01:01:35.046 --> 01:01:37.708 that I bring on and I enjoy doing as my

NOTE Confidence: 0.867772310625

01:01:37.710 --> 01:01:39.830 kind of translational research effort,

NOTE Confidence: 0.867772310625

01:01:39.830 --> 01:01:41.230 we don't do a lot of genetics,

NOTE Confidence: 0.867772310625

01:01:41.230 --> 01:01:42.510 but we've mostly been using

NOTE Confidence: 0.867772310625

01:01:42.510 --> 01:01:43.534 fish at this point.

NOTE Confidence: 0.867772310625

01:01:43.540 --> 01:01:46.460 Really old school conventional fish.

NOTE Confidence: 0.867772310625

01:01:46.460 --> 01:01:49.340 On occasion we'll do Archer,

NOTE Confidence: 0.867772310625

01:01:49.340 --> 01:01:51.080 one of the fusion panels,

NOTE Confidence: 0.867772310625

01:01:51.080 --> 01:01:54.414 but it's at MGH or Children's Hospital.

NOTE Confidence: 0.867772310625

01:01:54.414 --> 01:01:54.968 We don't.

NOTE Confidence: 0.867772310625

01:01:54.968 --> 01:01:57.240 We don't do it right now in our lab.

NOTE Confidence: 0.867772310625

01:01:57.240 --> 01:01:59.268 I think that.

NOTE Confidence: 0.867772310625

01:01:59.270 --> 01:02:01.370 Especially for round cell sarcomas,

NOTE Confidence: 0.867772310625

01:02:01.370 --> 01:02:03.158 it's really valuable to

NOTE Confidence: 0.867772310625

01:02:03.158 --> 01:02:04.946 have that method because.

NOTE Confidence: 0.867772310625

01:02:04.950 --> 01:02:10.118 If it isn't Ewing sarcoma, it is very hard.

NOTE Confidence: 0.867772310625

01:02:10.118 --> 01:02:14.196 To confirm the diagnosis of the

NOTE Confidence: 0.867772310625

01:02:14.196 --> 01:02:16.485 other so-called undifferentiated

NOTE Confidence: 0.867772310625

01:02:16.485 --> 01:02:18.774 round soul circles.

NOTE Confidence: 0.867772310625

01:02:18.780 --> 01:02:19.650 Thank you.

NOTE Confidence: 0.867772310625

01:02:19.650 --> 01:02:20.280 Yeah,

NOTE Confidence: 0.615612145

01:02:20.410 --> 01:02:21.790 in the head and neck area,

NOTE Confidence: 0.615612145

01:02:21.790 --> 01:02:24.415 this sarcoma is extremely baffling

NOTE Confidence: 0.615612145

01:02:24.415 --> 01:02:27.040 because that's not the first

NOTE Confidence: 0.615612145

01:02:27.130 --> 01:02:29.548 thing that comes to our mind.

NOTE Confidence: 0.615612145

01:02:29.550 --> 01:02:32.651 Fortunately for me, I also sign out

NOTE Confidence: 0.615612145

01:02:32.651 --> 01:02:36.347 on the bone and soft tissue service,

NOTE Confidence: 0.615612145

01:02:36.350 --> 01:02:37.910 so sometimes it strikes me

NOTE Confidence: 0.615612145

01:02:37.910 --> 01:02:40.289 it's not a head and neck tumor,

NOTE Confidence: 0.615612145

01:02:40.290 --> 01:02:42.630 but a soft tissue tumor.

NOTE Confidence: 0.615612145

01:02:42.630 --> 01:02:45.830 But they can be very challenging and we

NOTE Confidence: 0.615612145

01:02:45.830 --> 01:02:49.190 are trying to validate the orchard fusion,

NOTE Confidence: 0.615612145

01:02:49.190 --> 01:02:50.195 Plex version three,

NOTE Confidence: 0.615612145

01:02:50.195 --> 01:02:53.190 and we can't wait to get it started.

NOTE Confidence: 0.82844081

01:02:57.720 --> 01:03:01.717 If there are no other questions then.

NOTE Confidence: 0.82844081

01:03:01.720 --> 01:03:06.207 I'd like to thank Doctor Hornick profusely.

NOTE Confidence: 0.82844081

01:03:06.210 --> 01:03:10.249 For his generous time and for these

NOTE Confidence: 0.82844081

01:03:10.249 --> 01:03:13.648 very lucid discussions. Thank you, thank

NOTE Confidence: 0.9396385525

01:03:13.660 --> 01:03:16.996 you again thanks everyone. Have a great day.