Dr. Jason Barnes:

Hey there. Welcome to ENT in a Nutshell. My name's Jason Barnes, and today we will be talking about jugular paragangliomas. We are joined by Dr. Matt Carlson. Dr. Carlson, thanks for being here.

Dr. Matthew Carlson:

Here. Thanks so much for having me.

Dr. Jason Barnes:

I'll note that when we talk about jugular paragangliomas sometimes they're also called glomus jugulare. We might call them that during the interview today, but we'll start with presentation. Dr. Carlson, when folks present with this type of tumor, what do they often present with?

Dr. Matthew Carlson:

So most commonly patients who are presenting with jugular paraganglioma present with pulsatile tinnitus and conductive hearing loss. Less commonly, they may also have symptoms of lower cranial neuropathies, so dysphonia or dysphasia, and sometimes hyper nasal speech from palatal incompetence. Those are the most common presentations.

It's interesting that because of the position and where these tumors arise in the skull base, there's often a delay in diagnosis. So patients may present with slowly progressive symptoms for many years before diagnosis is actually established.

Dr. Jason Barnes:

And when we talk about the epidemiology of this tumor, what type of folks are, are walking into your clinic with this tumor?

Dr. Matthew Carlson:

So most patients who present with jugular paraganglioma are usually in their fifties, forties, fifties, or sixties, that could be in contrast to patients with familial paraganglioma who might present at a younger age. And jugular paraganglioma distinctly presents more commonly in women. The quoted rate is about a six to one ratio of women to men at presentation.

Dr. Jason Barnes:

When you first meet these folks, I'm sure they have a lot of questions, but what do you counsel them on in terms of the likelihood of this becoming malignant?

Dr. Matthew Carlson:

So there are some unique features or some features of jugular paraganglioma that we always think about in the back of our mind. One of those is the risk for malignancy. So the risk for malignancy for a head and neck paraganglioma is lower than having a malignancy associated with tumor paraganglioma outside the head and neck region. And the overall risk is about 1% or 3%.

What's interesting about the histopathology of malignancy is in contrast to other tumors, other types of cancers, the diagnosis of malignancy is not based on histopathological features. Meaning you can't biopsy the tumor itself and determine if it's malignant or not, but rather you have to find tumor within the lymph nodes to demonstrate metastasis.

Dr. Jason Barnes:

I understand there is a genetic component to this, which we'll talk about as well here in a few minutes. But when you see these folks in clinic and you perform a physical exam, what are some things you might look for that might tip you off that this is a jugular paraganglioma?

Dr. Matthew Carlson:

After presentation, they'll often they'll often come in with a CT scan plus or minus MRI. And so frequently the diagnosis is more or less obtained based on involvement of the jugular foramen. But on examination, almost always you'll see a middle ear component and that middle ear component might be just on the inferior aspect of the tympanic membrane or it might extended to fill the whole middle ear space on otoscopy. And you'll see a characteristic retro tympanic mass, and it has a very red or violation color, which is in contrast to many other tumors that can involve the middle ear space.

So when you talk about the differential diagnosis on otoscopy for a jugular paraganglioma, you have to include other tumors of the middle ear, such as facial nerve schwannoma, and endolymphatic SAC tumor, and encephalocele, that's herniating low from the tegmen tympani, a middle ear adenomas. And less commonly they could be mistaken for a inflammatory lesion, such as chronic otitis media with a polyp, with inflammatory polyps. I've seen that before where they've been referred for a jugular paraganglioma, but in fact, it was a really significant granulation tissue.

Much less commonly, they may be mistaken for abnormal location of a blood vessel going through the ear. So most commonly it would be a high jugular bulb, which has a more purple hue and it's usually posterior inferior.

And much less commonly, you can have an aberrant petrous carotid artery. And so instead of going inferior and anterior to the cochlea at that first genu, it can take a more lateral course and can actually abut the tympanic membrane and present in a similar way, a pulsatile mass in the middle of your space.

Dr. Jason Barnes:

In terms of physical exam, there are two signs that are kind of talked about in this topic, Brown's sign and Aquino's sign. Can you tell us about that?

Dr. Matthew Carlson:

Yeah, so Brown's sign is the finding that on pneumatic otoscopy the pressure that's created by pneumatic otoscopy creates a blanching of the tumor. So you'll lose some of that rosy color. It'll turn more white. And the tumor has to fill most of the middle ear space or all of the middle ear space for this to really be effective. And I would say that at least in my experience, it's certainly less than half the time you can get a very strong blanching of the tumor with a pneumatic otoscopy, if you perform it on your exam.

Your second examination finding that you talked about Aquino's sign is related to chronic compression and cessation of pulsation or at least objective pulsation in the patient's ear. I have to say that I've never performed this, but it is of historical relevance.

The last feature that people talk about on examination that I didn't mention earlier is the rising sun sign. And that's the idea that as the tumor comes from the inferior aspect of the jugular frame and into the middle ear space, it can recruit blood vessels of the ear canal and it can become much more hypervascular. And it creates this characteristic hue, a redness, with a prominent vascularity primarily inferiorly along the annulus and extending into the ear canal.

Dr. Jason Barnes:

You kind of mentioned this already, but we should be performing a scope exam when we see these patients?

Dr. Matthew Carlson:

Yeah. If you're suspecting a jugular paraganglioma, you want to perform nasal pharyngoscopy and a comprehensive lower cranial nerve examination in addition to the facial nerve examination. Just very briefly, facial nerve involvement prior to any treatment with the jugular paraganglioma is quite uncommon. It certainly occurs in less than 5% of cases overall.

When we talk about lower cranial nerve involvement with jugular paraganglioma, there's some things that are distinct from what we will more commonly see in our otolaryngology practice. And specifically for involvement of the jugular foramen tumors patients will commonly present with more than one lower cranial nerve and the presentation is that of a higher more proximal involvement of the nerve. So typically in clinic, in ENT clinic, we'll often see patients with a recurrent laryngeal nerve paralysis, either from a thoracic procedure or from a head and neck procedure, for example, but patients with juggler paraganglioma will have a high vagal injury.

And why is this important? They'll often present with palatal incompetence with nasal pharyngeal reflux, a rhinolalia which is associated with hyper nasal speech. And they'll often they'll also have ipsilateral vocal cord paralysis with loss of sensation. So when you perform nasopharyngoscopy you can touch their supraglottis with your scope if they have a paralysis and they'll often be insensate in that area.

Patients can also present with hypoglossal involvement. And hypoglossal involvement tends to be a little bit more under-recognized I think in these patients. Particularly a young or middle-aged person may compensate very well for a hypoglossal progressive weakness and it might not even be known to the patient.

Certainly an older patient may have dysarthria, et cetera, but a younger person you'll often see atrophy, hemi atrophy at the tongue, even without the patient reporting any supporting symptoms.

Shoulder involvement or accessory nerve involvement is also not super common. It may present with accessory nerve syndrome, where they might have a drop of the shoulder. They may have pain associated with it and a winged scapula.

When we talk about lower cranial involvement, at least on the boards, there's a couple of different jugular foramen syndromes. The first is Vernet. Vernet syndrome is associated with lower cranial nerve involvement of 9, 10, 11. And Villaret is associated with 9, 10, 11, 12, and also a Horner syndrome plus or minus the facial nerve.

And these aren't commonly used clinically, but the way to remember those, at least how I remember them for boards, for example, is Vernet is shorter and it involves less cranial nerves and Villaret is longer and it involves ... It's the Royal flush, 9 through 12 plus Horner's and plus or minus the facial nerve.

Dr. Jason Barnes:

And when we talk about pathophysiology, what is a jugular paraganglioma and how is it related to other tumors that we might see in the body?

Dr. Matthew Carlson:

So I think it's valuable to go back to original nomenclature. So historically these were called chemodectomatas because they are derived from chemo receptor cells. And then they more were described as glomus tumors. And now the correct term that we use is paraganglioma. So you can have jugular paraganglioma, tympanic paraganglioma, vagal paraganglioma, and then carotid body tumors. Those are your different, most common head and neck paraganglioma.

They are derived from chief cells within the paraganglia cells associated with the carotid body and the jugular foramen. They're neuroendocrine in origin and histologically they're very similar. So jugular paraganglioma histologically look very similar to carotid body tumors, and also pheochromocytomas. They're non chromaffin staining cells that tend to cluster in nests or rests that are commonly referred to as zellballen, which is a common board question for this.

Dr. Jason Barnes:

What are some of the complications involved in this tumor?

Dr. Matthew Carlson:

So originally we alluded to the fact that about 20% to 30% of patients will at presentation have lower cranial neuropathy. The majority will have pulsatile tinnitus and conductive hearing loss. If these tumors go untreated over the first three to five years, an additional 30% or so of patients will develop new or worsening lower cranial neuropathy. Over time, the tumor will often erupt into the ear canal and cause intermittent bloody otorrhea, which can be pretty significant in some cases.

And only with advanced disease do you tend to see other cranial neuropathies with more intercranial extension or surrounding the facial nerve. So you can have facial nerve paralysis, brainstem compression, and other related symptoms. That's a primarily the natural history of untreated disease as it grows.

You can also have secretion or functional tumor, and this is less common in head and neck paraganglioma compared to paraganglioma of the abdomen or chest. And about two to 5% of head and neck paraganglioma will secrete catacholamines.

The common symptoms of a secreting tumor are diaphoresis, palpitations, sweating. Sometimes patients will present with an arrhythmia, uncontrolled hypertension, headaches, et cetera. Less commonly patients may develop a malignant paraganglioma. With a malignant paraganglioma, as discussed earlier, the diagnosis is based on a metastasis to a surrounding lymph node rather than a direct histopathology taken from the parent tumor itself.

Dr. Jason Barnes:

When you see these patients in clinic, the workup is pretty extensive. Can you start by talking about the imaging that you should obtain and what the imaging looks like in these patients?

Dr. Matthew Carlson:

The first imaging tests that's usually obtained is a high resolution thin sliced temporal bone CT scan, and what you're looking for to characterize a jugular foramen tumor is jugular foramen involvement. And so, typically, a jugular tympanicum versus jugulare, you'll have erosion of the cortical jugular spine, which is the bone that intervenes between the carotid artery as it comes up into the temporal bone and the jugular bulb.

These tumors tend to exhibit moth eaten appearance of the bone and they spread. They have very poor defined margins and they can spread surrounding the jugular foramen. They can extend into to the petrous apex and they can have intercranial extension.

The second test that's usually obtained to further confirm the diagnosis and separate it from other jugular foramen tumors such as meningioma or schwannoma is the MRI scan. The MRI scan for a jugular paraganglioma will distinctly demonstrate a heterogeneous salt and pepper appearance. It's more prominent on T2, but can be seen on T1. It Will avidly enhance on T1. And again, it often has ill-defined borders or boundaries.

Now, when we're distinguishing these on MRI and CT scan from meningioma and also schwannomas, I think there's some characteristic features to talk about. So beyond the flow voids, when we look at a meningioma, meningiomas can involve primarily involved the jugular foramen or they can also secondarily invade the jugular foramen.

Meningiomas will not as frequently dilate the juggler foramen. They may have bone formation within the tumor exhibit on CT scan. They may also have hyperkeratosis of the surrounding base. They'll commonly exhibit dural tales as with meningioma elsewhere.

When we look at schwannomas, a schwannoma compared to a jugular foramen paraganglioma will more commonly involve or exhibit kind of a dumbbell appearance. So it'll will be wide within the posterior fossa, becomes constricted at the juggler frame it, and once then once again becomes larger in the neck. That pretty well distinguishes it from a juggler paraganglioma.

If the diagnosis still remains uncertain, which I'll tell you that probably 95% of the time the diagnosis can be made just on CT and MRI alone, but if the diagnosis still remains in question, conventional angiography may be beneficial. And of course, you'll, you'll see a very prominent vascular blush. That's most prominent with jugular paragangliogliomas compared to meningioma and also schwannomas.

Dr. Jason Barnes:

And another Other imaging technique that you might need to consider is when you're looking for multiple tumors in these patients. Can you talk a bit about that?

Dr. Matthew Carlson:

Yeah, that's a great point and something that I think is becoming more and more recognized. 10% of patients with sporadic head and neck paraganglioma, meaning they don't have a familial contributing component, will still have multicentric or multiple head and neck paraganglioma. And that number increases significantly to 10% to 50% of patients with familial syndromes will have multiple head and neck paraganglioma.

You also have to think about paraganglioma developing outside of the head and neck region. This is particularly important for a patient who presents with symptoms of catecholamine secretion. If the patient has symptoms of it, and more importantly if the patient demonstrates positive testing for catecholamine secretion, you should also be looking for a pheochromocytoma or another secreting tumor in the abdomen or chest, which we'll talk about in a little bit.

But when we're talking about primary head and neck involvement particularly for a jugular paraganglioma, I'll commonly still obtain a head and neck MRI to look for multicentric disease because that can significantly influence how you're treating these patients. If you know that a patient has bilateral involvement, you might treat the tumors differently because bilateral lower cranial nerve

paralysis in parts much more morbidity to the patient than say a unilateral involvement of a single tumor.

So when a patient has symptoms of catecholamine secretion, or if they have a familial component or genetic component, it's beneficial to look for disease outside the head and neck as well. And particularly for a patient who has elevated catecholamines it's commonly recommended to obtain an abdomen and chest CT scan to look for a pheochromocytoma.

If you're looking for systemic evaluation or systemic imaging radionuclide testing, the most common test used right now is Dotatate PET. And Dotatate PET has over 90% sensitivity and specificity for paraganglioma. Up until about 5 or 10 years ago, we were using FDG PET CT, which also has relatively good sensitivity and specificity, but it is now surpassed by Dotatate PET.

Dr. Jason Barnes:

What's the laboratory workup that you get for these patients and do you get it for every patient?

Dr. Matthew Carlson:

So that's a good question. Historically, we would only get laboratory testing in patients where we were concerned that they had symptoms of catecholamine secretion, so the patients with uncontrolled hypertension, headaches, pallor, flushing, et cetera, but more and more for patients with jugular paraganglioma we are getting routine testing. That's in contrast to tympanic tumors, which very rarely secrete, and we generally don't obtain testing.

But again, overall, the risk of a tumor secreting for a jugular paraganglioma is overall less than 5% for a head and neck tumor. So the laboratory workup most commonly is fractionated 24 hour urine metanephrines. You're looking for metabolites of norepinephrine, including metanephrine VMA, dopamine, et cetera. And when you're talking about elevation, it's common that you'll see just a mild elevation in the numbers, but for it to be real, you want at least a twofold or threefold increase over baseline in your metanephrine testing for you to consider it a positive test.

Dr. Jason Barnes:

There is a genetic component to this tumor. What's your genetic testing and what are some of the common mutations we see?

Dr. Matthew Carlson:

So molecular genetic testing is becoming more common for these tumors. Historically, we would only get genetic testing in patients who seemed at high risk. So the very young patient, particularly a young male, because they're less common in men, and also secreting tumors, malignant tumors, and patients with multicentric disease. But more and more, we are obtaining genetic testing.

The benefit of genetic testing is it might provide a prognosis for the disease and it might influence surveillance and even your approach to care. So jugular paragangliomas can associated with MEN type 2A and 2B, Von Hippel-Lindau, NF1, but more frequently they're discussed in the context of familial paraganglioma syndrome.

Familial paraganglioma syndrome is an autosomal dominantly inherited syndrome. It's typically associated with the succinate dehydrogenase mutation. There's multiple succinate dehydrogenase mutations you can have. They're typically classified as type one through type four.

Type one through three generally result in a more benign disease course, less likely to have secretion, less likely to have malignancy associated with them. And the type four, which is associated with succinate dehydrogenase B mutation, has a much worse prognosis overall. These patients are more likely to have a secreting, catecholamine secreting tumor, and also are more likely to have malignant potential.

Dr. Jason Barnes:

So once we've done the full workup, we've talked about physical exam genetics lab, work-up imaging, all of that. We diagnose a patient with a jugular paraganglioma. There are a couple of staging classifications when we talk about this tumor. Can you tell us about that?

Dr. Matthew Carlson:

Yeah. There have been multiple staging systems that have been devised over the years, but the two most commonly cited ones are the Ugo Fisch classification and also the Glasscock-Jackson classification. More commonly, we don't, even in clinic pragmatically or practically speaking, we typically don't apply a class to it when we're talking about the care of the patient. We more commonly just describe what's involved and the areas involved because these classifications are trying to group patients very specifically, but these tumors don't always follow a very characteristic or predictable pattern in their growth.

So for preparation there's primarily two different classification systems that are used. The first is the Glasscock-Jackson classification. Type one is a small tumor involving a jugular bulb, middle ear, and mastoid. A Type two extends under the internal auditory canal and it may have intercranial extension. The type three erodes into the petrous apex and it also may have intercranial extension. And a type four extends beyond the petrous apex to involve the clivus and infratemporal fossa. And it may also have intercranial extension.

When we talk about the Ugo Fisch classification, a type A involves the middle ear cleft, and that's what we commonly refer to as a glomus tympanicum. A B involves the middle ear and mastoid.

Type C1. So we're going to talk about C1 through C4 and that's different varying extents of carotid involvement. C for carotid. C1 involves the carotid foramen. C2 involves the vertical portion of the carotid. C3 involves the horizontal carotid artery involving the petrous apex. And C4 has more extension beyond the petrous apex to involve the paraclival area, foramen lacerum, and infratemporal fossa.

And then we talk about D, and D is related to having intercranial extension. So D1 has less than two centimeter intercranial extension, D2 has greater than two centimeters intercranial extension, and D3 indicates an unresectable intracranial tumor.

Dr. Jason Barnes:

So once you have this tumor classified and diagnosed, what are some treatment options and how do you counsel patients on which treatment options to pursue?

Dr. Matthew Carlson:

So historically, like all of their skull-based lesions, the mere diagnosis of a jugular paraganglioma necessitated surgical resection with a goal of a radical resection or gross total resection. And as you might imagine, this results in significant morbidity.

With use of MRI and a better understanding of the natural history of disease, we've tended to become more conservative over time. So currently there are primarily three treatment options, but there's multiple treatments underneath these individual main treatment options.

The first and most common or historically the most common treatment is surgical resection. And under surgical resection, there's multiple different strategies. You might perform limited resection, and that might be just the middle ear component to try to alleviate pulsatile tinnitus. You might perform more aggressive subtotal resection. And usually when we're talking about subtotal resection, we're talking about resecting the majority of the tumor, but leaving the most critical part in the jugular foramen with the intent of reducing cranial nerve morbidity.

So there are several series that demonstrate that you can resect over 90% of very large tumors and leave the portion that's migrating or invading medially through the jugular foramen and keep intact lower cranial nerves. That treatment paradigm might be followed with radiosurgery or just observed for a period of time.

Lastly, gross total resection again is the historical gold standard, but less commonly used today. Gross total resection is beneficial for the patient with a smaller tumor where the tumor can be completely removed with low morbidity and providing the patient with a cure. But once you have a larger tumor that's less feasible and less commonly performed the. Risk of developing new or worsening cranial neuropathy with gross total resection approaches an additional 20% to 40% on top of the baseline risk of lower cranial neuropathy that a person might present with of 20% to 30%.

And I also want to present one idea that I think is a common misconception. And that is the idea that if a patient presents with lower cranial neuropathy, it's probably reasonable to perform surgery because there they already have their nerves gone.

A lot of these patients, even though they demonstrate significant paralysis of the lower cranial nerves, it's often an incomplete paralysis. Removing those nerves or resecting those nerves during gross total resection may even worsen a person who seemingly had lower cranial nerve paralysis to begin with. So said another way, even when a person has a nerve out after surgery, that nerve might be even worse for dysphonia, dysphagia, et cetera.

The second treatment option is radiosurgery and more and more that's being used today. Radiosurgery is a single treatment that's outpatient. And most commonly it's used under the gamma knife platform, but there are other platforms with Linux, CyberKnife, et cetera. With gamma knife radiosurgery, there's a rigid head frame that's used to stereotactically isolate the tumor and treat it very specifically and reduce surrounding morbidity to tissue.

The risk of acquiring new cranial neuropathy after gamma knife radiosurgery using about 15 grade as the margin, which is the most common treatment dose that's used today, the risk of new or worsening cranial neuropathy is anywhere from 1% to 10%, depending on the series you read. And tumor control, that is not having the tumor grow substantially afterwards, is over 90% at least out to 10 years. So it has a very good track record for keeping the tumor at bay and not having it grow. And it has a very good track record as far as lower cranial neuropathy.

Stereotactic radio surgery may be used primarily or upfront, or it may be used as an adjunct after subtotal resection, or for recurrent disease after what was thought to be gross total resection with subsequent recurrence. There are some limitations with gamma knife radiosurgery, and probably the most pertinent limitation with stereotactic radiosurgery with gamma knife is the inferior extent of the tumor. With gamma knife you can only extend, at least with the current series, you can only extend down to C2. So if your tumor goes lower than that, you might not be able to encompass the whole tumor.

The last option is observation, and that's typically not used except for older patients or patients who are less well and not fit for surgery. But even in those patients more commonly they may undergo radiosurgery.

The natural history suggests that about 30% or 40% of the tumors will grow over five years and that number increases over time. But probably just as importantly, with observing the tumor, there's at least a 20% to 30% chance that the patient will acquire new or worsening cranial neuropathy over the first three to five years of observation compared to less than 10% for radiosurgery.

Dr. Jason Barnes:

When you talk about surgery, can you speak a little bit to your approach? So what you do preoperatively to ensure that surgery works as well as it can and what some of the common surgical approaches are for these tumors.

Dr. Matthew Carlson:

Yeah. So the preoperative considerations are ... I think the one thing that's important to mention, although rare, is a secreting tumor. So prior to surgery for secreting tumor, you need catecholamine control. And so you'll typically start with an alpha blockade followed by a beta blockade. If you reverse that order and you first use a beta blockade, you can have unopposed adrenergic response, which can really result in cardiovascular collapse. So it's important to do it in that order.

Almost always for a jugular paraganglioma we'll perform preoperative embolization with angiography, and there's different materials that you can use. Overall the risk of causing cranial neuropathy with the embolic agent is less than 5%, but there is a small risk associated with that. And preoperative embolization will reduce the amount of bleeding and the risk for transfusion.

The vessels most commonly embolized with preoperative embolization are the ascending pharyngeal artery and the occipital artery or with larger tumors you can have multiple artery involvement that's primarily associated with the external carotid system. But with much larger tumors you can also have parrots parasitization of intercranial blood vessels and dural vessels, too. And so, the larger tumors are more difficult to embolize sometimes.

Dr. Jason Barnes:

What's the surgical approach? There's a name for it from my understanding.

Dr. Matthew Carlson:

Yeah, Ugo Fisch popularized a more systematic approach for gross total tumor resection right around 1980 and the infratemporal fossa type A approach is used in most patients. Classically, the infratemporal fossa type A approach involves a subtotal petrosectomy.

So you're exonerating essentially most of the pneumatized ear cells of the temporal bone and you're closing the ear canal. So it's in many ways a very large canal wall procedure. You'll then mobilize the facial nerve. And when you mobilize the facial nerve in a infratemporal fossa type you're mobilizing it anteriorly across the second, basically pedicled across a GSPN.

And just moving the nerve with the infratemporal fossa type A approach will result in at least temporary but sometimes some degree of permanent facial nerve paralysis. This has led to modifications to the infratemporal fossa type A approach. And more and more patients are not mobilizing the facial nerve, but leaving it in a fallopian canal and working on the front and the back side

of the nerve and preserving the ear canal in many cases. So the patient doesn't have a conductive hearing loss.

You can also perform a limited inferior mobilization of the facial nerve, and that's just by the stylomastoid foramen. And in those couple of millimeters can also be really beneficial when you're trying to remove the tumor in the region of the jugular foramen.

Dr. Jason Barnes:

You've talked a lot about prognosis and expectations for the different types of approaches and treatments, but how do you counsel patients on followup? What's your long-term followup with these patients?

Dr. Matthew Carlson:

Followup is largely dictated based on your treatments and the amount of remnant residual disease. So if you're observing a patient, you'll commonly get a scan six months later than yearly for several years, and you can go to biennial after that.

If you have performed microsurgical resection, the risk of recurrence is proportional to the volume of tumor left behind. So in the case of a very subtotal resection I'll image more frequently, but if I performed a very aggressive subtotal resection or near total resection I'll image less frequently. And with the gross total resection, I would usually obtain a scan in about one year and then at three to five years, and then more infrequently thereafter, but probably lifelong surveillance to some levels is beneficial.

When you consider radiosurgery, the important thing about radiosurgery is it doesn't get rid of the tumor. Sometimes it'll shrink, but it doesn't ever completely resolve the tumor. You'll always see it on the scan. And so we'll typically get an MRI about nine months to a year after treatment. And then, depending on the size of the tumor and the initial treatment response, we could obtain every one to three years for several years. But again, indefinite followup is needed for patients who undergo radiosurgery as well.

Dr. Jason Barnes:

Well, Dr. Carlson, thanks again so much for being here. Before we left, I wanted to give a final summary. Jugular paragangliomas are paragangliomas of the temporal bone. And they often present with pulsatile tinnitus, conductive hearing loss, and possible lower cranial nerve deficits.

The pathology is a pair of ganglioglioma of the adventitious of the jugular bulb and as is associated with the zellballen NS. Workup includes imaging, which demonstrates moth eaten bone on CT and a salt and pepper appearance on T1 with contrast, and its hyperintense on T2.

Additional workup includes a genetic workup, which could possibly identify mutations in the succinate dehydrogenase gene, or it could also be associated with Von Hippel-Lindau, NF1, or MEN 2A or 2B.

You should consider lab workup for 24 hour urine fractionated catecholamines or metanephrines or plasma metanephrines in the rare case that this is a hyper secreting tumor.

Glomus jugulare can be classified using the Fisch classification system or the Glasscock-Jackson classification system. Treatment options include radiation and surgery. Radiation is good at keeping the tumor from growing. And surgical approaches include the infratemporal fossa type A approach, which is

commonly cited for this tumor. And preoperative embolization and medical treatment should be considered.

Surgical resection does have a moderate chance of additional lower cranial nerve deficits. Followup includes imaging for confirmation of total resection. If surgery is done and to confirm that there are no additional tumors.

It's time to bring this show to a close, but before we do, I did just have some final questions to ask. Again, I will ask a question, give five or so seconds of pause so that you can think, or press pause yourself, and then I'll give the answer.

The first question is what is the classic histology of a pair of ganglia? The classic histology of a pair of ganglioglioma is the zellballen pattern, which is a nest of non chromaffin staining cells among vascular channels.

The Next question is what are the most common genetic mutations involved in temporal bone paragangliomas? The most common genetic mutations involved in juggler paragangliomas are succinate dehydrogenase mutations, Von Hippel-Lindau, NF1, and MEN 2A and 2B.

Next question, what are the two common classification schemes of glomus jugulare? And I'll even try to have you recite one of them. The two main ways of categorizing glomus jugulare are using the Fisch classification and the Glasscock-Jackson classification. And I'll run through them both right now.

The fish classification has type A, which is tumor limited to the middle ear, and type B, which is tumor limited to the tympanomastoid area with no intralabyrinthine compartment. Type C is tumor involving the intralabyrinthine compartment and extending into the petrous apex, and that has sub units. And type D is intracranial extension. There's types of C1 through three and types D1 and D2.

For the Glasscock-Jackson classification type one is a small tumor involving the jugular bulb, middle ear, and mastoid. Type two is tumor extending under the internal auditory canal. Type three is tumor extending into the petrous apex. And type four is tumor extending beyond the petrous apex into the clivus and infratemporal fossa.

Finally, what is the most commonly described surgical approach for jugular paraganglioma? A commonly cited surgical approaches is the infratemporal fossa type A approach, though there are several approaches to these tumors.

Thanks so much for listening and we'll see you next time.