Dr. Jason Barnes:

Hey there. Welcome to ENT in a nutshell. My name is Jason Barnes, and today we will be discussing glomus tympanicum and we are joined by Dr. Matt Carlson. Dr. Carlson, thanks again for being here.

Dr. Matthew Carlson:

Thanks so much for having me.

Dr. Jason Barnes:

We do have another episode on glomus jugulare, and while these two types of tumors do share some similarities, we felt it would be worthwhile to have a different episode to specifically discuss glomus tympanicum. So, Dr. Carlson, why don't you tell us how a patient presents to your clinic when they do have a glomus tympanicum.

Dr. Matthew Carlson:

Yeah, so the great majority of patients who present with glomus tympanicum tumors present with pulsatile tinnitus, and that occurs in over 80% of patients. Also, coinciding with that is conductive hearing loss and by far and away, these are the two most common symptoms. Less commonly patients may have, with a larger glomus tympanicum tumor, the tumor can erupt through the eardrum and can cause bloody otorrhea.

Dr. Jason Barnes:

And do these patients have cranial nerve deficits?

Dr. Matthew Carlson:

So that's important characteristic that defines glomus tympanicum versus glomus jugulare. So glomus tympanicum tumors typically do not result in any sort of cranial neuropathy. Even with more advanced disease, it's uncommon for them to cause facial nerve paralysis and lower cranial nerve paralysis. And essentially by definition, if at diagnosis you identify a patient with invasion of the jugular foramen and lower cranial nerve paralysis, that condition would be defined as a jugular paraganglioma rather than a tympanicum.

Dr. Jason Barnes:

And when you evaluate these patients in clinic, what will you see on the physical exam?

Dr. Matthew Carlson:

So on physical examination, on otoscopy, you'll see a retro-tympanic mass, and it will have a characteristic deep red hue. That'll be in contrast to other tumors or other diseases that can present in the middle ear space. So for example, a facial nerve schwannoma will typically have a more pink or light pink or white appearance and more commonly, it will be in the kind of the posterior quadrant. A large encephalocele from the tegmen tympani may also be pulsatile just like a glomus tympanicum, but it has a more deep purple hue and it tends to be pedicled superiorly. Aberrant carotid artery would be an antero-inferior if it did have a more lateral course. A high jugular bulb would be more posterior, posterior inferior compared to a glomus tympanicum. And then you can have a chronic otitis media with a polyp, for example, might present with bloody otorrhea and rarely can be mistaken for a glomus tympanicum tumor.

Dr. Jason Barnes:

And when we talk about the type of patient that you see who presents to your clinic, what's the general demographic of folks with this type of tumor?

Dr. Matthew Carlson:

So most commonly patients with glomus tympanicum will present in their forties, fifties, or sixties, but certainly outside that age range is possible. Paralleling patients with jugular paraganglioma, there is a predilection towards women, but it's not strong. Jugular paragangliomas have a association with about six to one female to male involvement. And with tympanicum it's not quite so strong.

Dr. Jason Barnes:

And is there a familial component to this?

Dr. Matthew Carlson:

That's a good question. Patients with glomus tympanicum can have isolated disease, or they might have a familial component or even sporadic, multiple head-neck paragangliomas. Tympanicum aren't as strongly associated with familial paraganglioma syndromes compared to say a jugular paraganglioma or a vagale or carotid body tumor, but it still can occur. As we discussed in the other podcast on jugular paraganglioma, there are some familial conditions that might predispose one to developing tympanicum or jugulare and those would include MEN 2A, 2B, NF1 or Von Hippel-Lindau. Or it may be associated with the condition of familial paraganglioma syndrome, which is not as normal dominantly inherited a condition that's associated with the succinate dehydrogenase mutation of one of four different mutations.

Dr. Jason Barnes:

And moving on to pathophysiology, what is a glomus tympanicum?

Dr. Matthew Carlson:

So historically, all of these temporal bone paragangliomas were given different names and [inaudible 00:04:16] what was originally ascribed simply because they arise from chemo receptor cells associated with parasympathetic nerves. More specifically are cells derived from the paraganglia, they contain non-chromaffin staining chief cells. And when those cells cluster or lie in nests, they have a characteristic histopathological appearance called zellballen or rests of cells. And that's a common board question that's asked.

Dr. Jason Barnes:

And occasionally I've seen a mention of specific nerves that are involved in this tumor or from which this tumor emanates. Can you talk more about that?

Dr. Matthew Carlson:

So all the paragangliomas of the head and neck, they originate from different original structures. And so just to answer your question more broadly, jugular paragangliomas arise from the adventitia of the jugular bulb. Carotid body tumors arise from the chemoreceptor cells in the carotid body. And then specifically for glomus tympanicum, they're thought to arise from the tympanic plexus related to

Jacobson's nerve, which is the tympanic branch of the ninth nerve, and also Arnold's nerve, which is a branch of the vagus nerve that involves the tympanic plexus.

Dr. Jason Barnes:

And when you suspect this type of tumor, what's your initial workup for these patients?

Dr. Matthew Carlson:

So typically the examination is very diagnostic. Again, you see a retro-tympanic mass, it's typically pulsatile, patients will have conductive hearing loss and pulsatile tinnitus, and an absence of cranial neuropathy. And that alone should allow you to make a diagnosis with pretty high certainty, at least working clinical diagnosis. The general rule of thumb is if you can see a well circumscribed lesion in the middle ear space, and you can see all the way around it on otoscopy, technically that should exclude the possibility of this being actually a jugular paraganglioma arising from the jugular bulb and coming up into the middle ear space.

I'll say that most of the time, these patients will have a temporal bone CT scan and that's just to make sure it doesn't extend into other areas of the temporal bone. So, for example, again, if it went down into the jugular foramen you would call this a jugular paraganglioma. If it was involving other structures, you might change your differential diagnosis depending on where it's emanating from. You also want to see the extent of the tumor itself. Is it just confined to the middle of your space on the promontory? Is extending down into the hypotympanum? Does it involve the mastoid part? Is there a rotation of the tegmen, et cetera?

Dr. Jason Barnes:

And do you typically get an MRI on these patients if you feel like you see the well circumscribed mass, and you feel very confident in what it is, do you still get an MRI?

Dr. Matthew Carlson:

If their presentation is very typical and their otoscopic examination is what we'd expect and their suspicion is very high and the CT findings are straightforward, then we typically don't get additional imaging.

Dr. Jason Barnes:

But if you were to get an MRI, what would that show?

Dr. Matthew Carlson:

So if you get an MRI, if it's a large enough lesion, you can see flow voids. And flow voids it has a characteristic, salt and pepper appearance more commonly better seen on T2, but you can see it on T1, post-gadolinium contrast administration. Now these tumors also avidly enhance with gadolinium and they're relatively heterogeneous tumors.

Dr. Jason Barnes:

And for these patients, do you typically get any labs? The reason I ask is because other paragangliomas such as a pheochromocytoma or even a jugular paraganglioma, we talk about working up for metanephrines and that kind of thing.

Dr. Matthew Carlson:

So, as we discussed in the previous podcast related to jugular paraganglioma, historically, we would only get catecholamine testing if a person was symptomatic with hypertension, flushing, headaches, or other symptoms of secretion and more commonly now, we are routinely getting catecholamine testing for all our patients with jugular paragangliomas. In contrast, a person with an isolated glomus tympanicum, I still think most people are not getting additional testing unless there's evidence of multicentric disease or other symptoms to suggest it, but the likelihood that a glomus tympanicum I'm itself as a secreting tumors extremely low and it's case reportable. And most people won't think that it's a high risk situation.

Dr. Jason Barnes:

And what about genetic testing? We talked about that with jugular paragangliomas, do you routinely seek out genetic testing in these cases?

Dr. Matthew Carlson:

So similar to jugular paraganglioma, historically we would never get a genetic testing on any of these patients and more and more for jugular paragangliomas we are getting genetic testing, but still we really don't commonly do it for isolated glomus tympanicums. And the reason is oftentimes these are isolated tumors that are not associated with the familial paraganglioma syndrome or another condition. If a person wanted to get genetic testing for this, again, you'd probably be looking for the same thing, the succinate dehydrogenase mutation type one through four. If they had multicentric disease, you could also look for other conditions that are associated with it.

Dr. Jason Barnes:

So we've talked about presentation, pathophysiology, workup. So once you have an idea of what this tumor is, or you're ready to call this glomus tympanicum, what is the classification system? How do we define these?

Dr. Matthew Carlson:

So this classification system is not widely distributed or known, but it can be tested on boards and so I think it's worth at least briefly mentioning. And the Glasscock-Jackson staging system, there are two of them. There's one that's specific for glomus tympanicum and there's a separate one that's specifically specific for jugular paraganglioma.

And so we'll review the glomus tympanicum staging system. So stage one is defined by all tumor margins visible on otoscopy. Stage two, the tumor fills the middle ear and margins are not visible. Stage three, the tumor extends into the mastoid cells. In stage four, the tumor erodes through the tympanic membrane or bone of the external auditory canal. Now that's the Glasscock-Jackson staging system for glomus tympanicum. The Ugo Fisch classification for jugular paraganglioma and glomus tympanicum blend into one single classification system under jugulotympanicum. And so if you'll recall from the prior podcast, a Fisch A involves just the middle ear space, which would be a small tympanicum. B involves the middle ear and mastoid, which would be medium-sized tympanicum. And then after that, in their staging system, you get into what we would commonly call a glomus jugulare.

Dr. Jason Barnes:

Gotcha. So what's the treatment for these?

Dr. Matthew Carlson:

So the treatment for glomus tympanicum diverged from what we typically talk about for glomus jugulare, and the reason is this glomus jugulare tumors are much larger and there's more morbidity with resection, specifically risk of lower cranial neuropathy with resection of glomus jugulare. So for a glomus tympanicum, the treatment is generally surgery. We don't radiate. We don't typically recommend radiosurgery for these lesions, simply because, in general, removal has low morbidity, a very high cure rate and low recurrence. And so the typical treatment is surgical resection with the goal of gross total resection. It would be very uncommon that you'd have to leave a little adherent disease behind perhaps to the facial nerve or the dura or something like that. The approaches that you use for surgery for resection would be based on the extent of the tumor. So a small tumor, isolated to the middle ear space, could be performed through a transcanal tympanomeatal flap, much like a stapedectomy. If it's a little bit bigger, you might do a postauricular transcanal approach with a canalplasty and removing some of the medial bony ear canal for better visualization.

If it's more involved, you might do a formal tympanomastoidectomy approach with intact canal wall, and you might perform a facial recess or an extended facial recess, where the chorda tympani nerve is divided inferiorly and that can take you into the hypotympanum. You can also resect mastoid involvement at that point. For very extensive disease and particularly if a person has bad hearing, you might perform a subtotal petrosectomy with ear canal closure, but those would be basically the surgical approaches you would use for a glomus tympanicum small, medium, or large.

Dr. Jason Barnes:

Does radiation play any role in this treatment paradigm?

Dr. Matthew Carlson:

We don't typically recommend radiosurgery for these lesions and that primarily gets back to the fact that a surgical resection is curative and it has a low morbidity. It also provides immediate benefit or alleviates two of the primary symptoms patients have with glomus tympanicums, and that is the pulsatile tinnitus and the conductive hearing loss. These are both usually resolved immediately following surgery. In contrast, if you perform radiosurgery, it probably would never get rid of the conductive hearing loss. There is some data that shows that over time, radiosurgery does provide some efficacy for pulsatile tinnitus in about 50 or 60% of patients. But this is over time and not an immediate benefit.

Dr. Jason Barnes:

And when we talked about glomus jugulare, we talked about preoperative embolization. Is that something that you do in these tumors?

Dr. Matthew Carlson:

We don't typically perform angiography or embolization in these cases. If the tumor was very large and it extended into the mastoid and was a large, it wouldn't be crazy to do so, but specifically for a small middle ear, isolated tumor, we typically wouldn't perform embolization.

Dr. Jason Barnes:

And briefly, what are the typical outcomes you see for these procedures?

Dr. Matthew Carlson:

So microsurgical resection will provide a relief of pulsatile tinnitus in the great majority of patients, improvement in the conductive hearing loss and it does provide a durable cure. So the rate of recurrence is probably primarily related to the size the original lesion was, and whether or not you perform gross total resection, near-total resection or subtotal resection, but with gross total resection, the risk of long-term recurrence is certainly less than 5%.

Dr. Jason Barnes:

And how do you follow up with these patients after surgery?

Dr. Matthew Carlson:

Particularly for middle ear tumors or tumors that were isolated to the middle ear, I think a clinical examination with otoscopy sufficient for most patients. A recurrence will typically present with the original symptoms. Patients will say, "You know what? I've had a recurrence of my pulsatile tinnitus in my ear." And they might have a conductive hearing loss on examination. In most situations, you'll be able to see the tumor once again.

Dr. Jason Barnes:

I think we've had a pretty comprehensive discussion regarding this topic, but before we summarize what we've talked about, is there anything else you'd like to add?

Dr. Matthew Carlson:

There are a couple last things that are sometimes asked about on board questions. And one of those is the Brown sign. So Brown sign is commonly discussed in the context of jugular paraganglioma, but it also can apply to glomus tympanicum. And that's the idea that the tumor will blanch or lose some of its redness when you perform pneumatic otoscopy. That's simply compressing the tumor and flushing some of the blood out of it on appearance. The second thing that's sometimes asked, in addition to what the paraganglia cells of origin are related to Jacobson's nerve and Arnold's nerve and the tympanic plexus, another question related to that is what the blood supply of these tumors commonly arise from. And glomus tympanicum typically arise from a tympanic twig or artery off of the ascending pharyngeal. And this is often encountered during surgery. When you're removing these tumors, you'll be taking out the tumor and you'll see a larger vessel coming up inferiorly, which can be usually easily controlled with bipolar coagulation or applying bone wax into the artery opening.

Dr. Jason Barnes:

Great, so in summary glomus tympanicum, when patients present with this, they often present with pulsatile tinnitus, subjective hearing loss and possibly aural fullness, but they should not have lower cranial nerve deficits, unlike glomus jugulare. The physical exam will often show a red, pulsatile mass in the middle ear. Folks that present are often women and the mean age of presentation is in the fifth or sixth decades of life. And in terms of pathophysiology, we're talking about a paraganglioma, which is nests of non-chromaffin staining cells that are referred to as the zellballen pattern.

When we work these patients up, we can get a CT which would show moth-eaten bone and in the cases that we get an MRI, we'll see a quote, unquote, "salt and pepper appearance" on T2 and somewhat on T1 with contrast. We don't really have to get laboratory and genetic workup in these cases, which is different from a glomus jugulare. And when we talk about diagnostic staging, there's a specific Glasscock-Jackson classification for this. In terms of treatment, again, in distinction from glomus

jugulare, we almost only operate on these patients and get a great surgical outcomes, often with these symptoms being vastly improved. Dr. Carlson, is there anything else you'd like to add?

Dr. Matthew Carlson:

Well, I think that sums everything up very well. That's glomus tympanic in a nutshell,

Dr. Jason Barnes:

It's now time to bring episode to a close, but before we do, I did just want to ask a few closing questions. As we normally do, I will ask a question, wait a few seconds for you to pause or think of an answer and then give you the answer.

So the first question for today is, what are the common presenting symptoms of glomus tympanicum? The most common presenting symptoms of glomus tympanicum are conductive hearing loss, pulsatile tinnitus, and possible bloody otorrhea.

The next question is, what is the most common blood supply of glomus tympanicum tumors? The most common blood supply of these tumors is usually a branch off of the ascending pharyngeal artery.

For our next question, what are Jacobson's nerve and Arnold's nerve? Jacobson's nerve is the tympanic branch of the glossopharyngeal nerve and Arnold's nerve is a branch of the vagus nerve, most often the auricular branch.

Our next question is, what is the specific classification system that is used for glomus tympanicum? And if you can, try to remember what those stages are. The classification system that's specific for glomus tympanicum is the Glasscock-Jackson classification system. This is four stages where stage one, the whole tumor is visible on otoscopy. Stage two, is the tumor is in the middle ear, but the margins are not visible. Stage three, is that the tumor extends into the mastoid and stage four, is that the tumor has eroded through the tympanic membrane or the external auditory canal.

Finally, for our last question, what is Brown's sign? Brown's sign is a blanching of a tumor with pneumatic otoscopy. That's all for today. Thanks so much for listening and we'll see you next time.