

Dr. Linda Yin:

Hello. And welcome back to another episode of ENT In A Nutshell. My name is Linda Yin, and I am joined today by Dr. Eric Moore, who is a head and neck surgeon. Thanks for being here, Dr. Moore.

Dr. Eric Moore:

It's a pleasure to be here, Dr. Yin.

Dr. Linda Yin:

All right. Today, we're going to talk about some primary parapharyngeal neoplasms. Tumors of the parapharyngeal space can take on a variety of pathologies, but they share a common anatomic region that can pose some unique diagnostic and treatment challenges. So we're going to go through some of that today. Let's start with presentations. So what is the typical presentation of a patient with a parapharyngeal tumor?

Dr. Eric Moore:

Well, because parapharyngeal tumors are both slow growing in most instances, and also deep within the neck, most patients don't realize they have a parapharyngeal tumor. In fact, the most common presentation is a patient comes in where they've had an imaging study for some other reason or a physical examination for some other reason, and it's been pointed out to them that they have a parapharyngeal tumor. As the tumors get larger sometimes the patient will notice it. So they may feel it by putting their fingers just below the angle of their jaw, and just in front of their ear lobe, they may feel the tip of the mass or the tip of the iceberg. Or they may notice a lump in their throat as it medially pushes the parapharynx into the oropharyngeal area. But most of the time, these have been found in patients incidentally.

Dr. Linda Yin:

When you do find a parapharyngeal mass, what sort of physical exam maneuvers can you perform to examine it?

Dr. Eric Moore:

So visual examination, both of the patient from afar and visual examination up close of the neck, can sometimes give you a hint that there's displacement of the parotid laterally or the neck structures laterally that will hint at a parapharyngeal mass. Oropharyngeal examination will give you a hint. So if the patient has a bulging tonsil appearance, that may be a pair of pharyngeal mass displacing the pharyngeal constrictor and the tonsil medially. Bimanual palpation is successful. So you can put your fingers in the floor of the mouth and also just in front of the tonsil and then palpate the neck with the other hand and gain an appreciation that there's fullness in this area.

Dr. Linda Yin:

What about laryngoscopy, do you perform that?

Dr. Eric Moore:

I routinely do perform laryngoscopy, and it's helpful for patients with parapharyngeal masses to gain an appreciation for the extent and size of the mass. So you may see the pharynx bulging medially even further down below the tonsil. And also doing a laryngeal examination is important because the

parapharyngeal tumors can affect the cranial nerves, and so evaluating the airway and evaluating the larynx and looking for vocal cord paralysis is important.

Dr. Linda Yin:

What is the typical epidemiology of parapharyngeal neoplasms?

Dr. Eric Moore:

Well Linda, parapharyngeal neoplasms are uncommon tumors. They have a lot of similarities to parotid tumors, and in fact, many of them are salivary gland tumors, but they have very low incidents. Less than half percent of the population walking around would have a parapharyngeal tumor. And the vast majority of these tumors are benign rare neoplasms.

Dr. Linda Yin:

What is typically on your differential diagnosis when you see a parapharyngeal mass?

Dr. Eric Moore:

The parapharynx is largely filled with fat and lymph nodes and either a portion of the parotid gland or even ectopic salivary rests, which give rise to parapharyngeal tumors. And then finally it's occupied by the carotid artery and the jugular vein and the vagus nerve. And so these structures give rise to the tumors and the vast majority of parapharyngeal tumors are arising from the deep portion of the parotid gland or the ectopic rests of salivary tumors.

Other things that you can have in this area are tumors of fat. So you can have lipoma and liposarcomas. There are lymph nodes in the parapharyngeal space. So you can have all the pathologies that would go along with the lymph node, including lymphadenopathy of an infectious or inflammatory nature, or you could have primary lymphoma or metastatic tumors to the parapharynx.

And then finally those tumors that arise from the cranial nerves and the great vessels are paragangliomas or schwannomas or neurofibromas, which make up the vast majority of the parapharyngeal tumors.

Dr. Linda Yin:

In terms of incidents, what would you say is the most common type of neoplasm that's found in this area?

Dr. Eric Moore:

Overwhelmingly, the most common neoplasm is salivary gland etiology tumors. So benign salivary tumors, such as pleomorphic adenoma, or oncocytoma, low grade salivary malignancies, acinic cell carcinoma, mucoepidermoid carcinoma, or even high grade salivary malignancies, make up the vast majority of the tumors. Most of the tumors are benign arising from the salivary gland.

Dr. Linda Yin:

Okay. I think before we get into any more details, we should probably define the anatomy of the parapharynx for the listeners. So what are the boundaries of the parapharynx?

Dr. Eric Moore:

The classic image of the parapharynx is described as an inverted pyramid. The flat broad surface at the top would be the base of the skull, consisting of the temporal bone and portion of the sphenoid bone. The lateral border consists of the medial pterygoid muscle, posterior belly of the digastric muscle, and the mandible. And the medial border consists of the pharynx, so immediately medial to the parapharynx is the buccopharyngeal fascia and the pharyngeal constrictor muscle. And then the inferior apex of the parapharynx is the hyoid bone. So it's a pyramid turned upside down bounded by the skull-base superiorly, the mandible laterally, and the buccopharyngeal fascia, pharyngeal constrictor, medially.

Dr. Linda Yin:

And from my understanding, the space itself can be subdivided into two different compartments. So what are these compartments called?

Dr. Eric Moore:

So this is the most important thing to understand about the parapharynx, because this really guides you towards diagnosis of the masses that occurs in that area, and deciding what to do and how to approach those masses. So classically, this is described as a prestyloid and poststyloid component. The actual division is fascia that arises from the styloid process going to the velopharynx. But on imaging, if you just have in your mind's eye, those masses that are anterior to the styloid process and posterior to the styloid process, then that can really guide you as to what you're dealing with, and how you would approach it.

Dr. Linda Yin:

So what is in the prestyloid compartment?

Dr. Eric Moore:

The prestyloid compartment is mostly that deep lobe of the parotid gland, ectopic salivary rests, lymph nodes, and fat. So the vast majority of the tumors occur in the prestyloid portion of the parapharynx, and then consist of those salivary origin tumors or lymph node etiologies.

And the poststyloid component then is housing the vagus nerve, the carotid artery, and the jugular vein. So the paragangliomas and the schwannomas and the neurofibromas, occur in the poststyloid component. And then those primary tumors of the salivary gland and the lymph node etiologies occur in the prestyloid component for the most part.

Dr. Linda Yin:

All right. And I know we already talked about benign salivary cancers in a different podcast, but of the other etiologies in the parapharyngeal space, what normal structures can these tumors arise from?

Dr. Eric Moore:

So this largely consists of poststyloid parapharyngeal tumor masses, and these would arise from the internal carotid artery in the form of paragangliomas or the jugular vein in the form of paragangliomas, or from the vagus nerve or the sympathetic trunk arising as paragangliomas or schwannomas, from the schwann cell nerve sheath covering, or neurofibromas themselves.

Dr. Linda Yin:

These neurogenic tumors, which cranial nerves are they typically originating from?

Dr. Eric Moore:

In the parapharynx, the neurogenic tumors largely arise from the vagus nerve, but they could also arise from branches of the trigeminal nerve. They could also arrive from the sympathetic trunk, or even lesser named unknown nerves, to muscles of mastication or muscles around the parapharynx.

Dr. Linda Yin:

Okay, let's move on to work up. So in addition to a physical exam and to laryngoscopy, are you getting any imaging studies on these patients?

Dr. Eric Moore:

It's very important to get imaging studies on patients with parapharyngeal tumors, as often your physical examination under represents the extent of the tumor and the imaging studies really help you hone down the diagnosis. So I think a CT scan is readily available to most patients and can really help you out with that prestyloid, poststyloid component. A CT scan can help you gauge the size of the tumor, its lobularity, and its consistency, which can sometimes help you in diagnosis, and its involvement of the surrounding bone, which may be important in both diagnosis and treatment planning.

MRI scans are also commonly used for tumors of the parapharynx. Sometimes you get the same information from these two types of scans, and other times you get complimentary information which may really help you out. So the MRI scan can help you out distinguish paragangliomas, from nerve sheath tumors, or neurofibromas. It can help you distinguish tumors of the parapharyngeal space, as far as they're infiltrating nature, well circumscribed nature, which can help you with benign or malignant etiology.

So the CT scan helps a lot with the vessels and the prestyloid, poststyloid differentiation, and MRI scan can help you out a little bit with the tumors interaction with the surrounding structures.

Dr. Linda Yin:

Is there any role for angiography, like a CTA or an MRA?

Dr. Eric Moore:

So for the prestyloid component parapharyngeal tumors, which again are very much more common, there isn't much role for angiography. Most of these tumors displace the vessels around them, but do not invade or surround the vessels, and angiography and embolization doesn't really play a role in management of these tumors. But the poststyloid component tumors, angiography can be very helpful. So for the paragangliomas, understanding the origin of the tumor can be assisted by angiography. Embolization sometimes of the tumor to decrease the vascularity during treatment is employed particularly for the carotid paragangliomas. So this may be very important in the treatment planning for these tumors.

Dr. Linda Yin:

When you look at imaging for these tumors, can you tell based on its own location, where they're originating from, and how does imaging help you narrow that differential?

Dr. Eric Moore:

So if you have good imaging with contrast, then you can see well, the relationship of the carotid artery, the jugular vein, and the vagus nerve. And that triangle consists of the carotid artery, [inaudible 00:10:57], the jugular vein antero laterally, and the vagus nerve sitting posteriorly and in between those two structures.

So then if you have a mass arising from one of those two structures, how it displaces those structures can help you as far as the etiology of the mass. And you can differentiate sometimes between a vagal paraganglioma and a jugular paraganglioma, or a carotid paraganglioma. For instance, if you have a vagal paraganglioma, then you would expect it being posteriorly to displace the carotid artery immediately, and a jugular vein anteriorly and laterally, versus if you had a jugular paraganglioma you would expect it to displace both of those structures, posteriorly and slightly medially, and this can help with your differential diagnosis.

Dr. Linda Yin:

What are some classic signs that we can look for in imaging that may really point us towards a particular diagnosis?

Dr. Eric Moore:

So some of the signs that are really associated with parapharyngeal tumors and are really helpful when you're thinking of differential diagnosis, are the lyre sign. The lyre sign is indicative of a carotid paraganglioma where the internal carotid artery and the external carotid artery are separated and rounded from each other, from the tumor that sits in the notch of the carotid body, between these two structures. And then finally the vessels come back around the tumor and come closer together. The classic lyre musical instrument.

Another very typical parapharyngeal sign is the dumbbell tumor. The parotid gland deep portion sits up against the styloid mandibular ligament and can transmit a tumor that emanates medially from this deep portion into the parapharyngeal space, and then the tumor enlarges with a waist or impingement around its middle, caused by the styloid mandibular ligament in the mandible. And this will give a dumbbell shaped appearance to the tumor with the medial portion in the parapharyngeal space and the lateral portion outside the parapharyngeal space, in the parotid gland deep portion.

And finally classic salt and pepper appearance of the tumor consists of heterogeneity of light and dark areas on MRI scan, which is typical of neurofibromas within the parapharyngeal neural structures.

Dr. Linda Yin:

Can these lesions easily be biopsied? And if so, what do you think is the best way to obtain a biopsy for pathology?

Dr. Eric Moore:

Well, fine needle aspiration is overwhelmingly the best way to biopsy all lesions in the head and neck, and not to disrupt the tumor, give the pathologist a very good sample for diagnosis, and not to impede any tumor treatment plans. But fine needle aspiration can be difficult for tumors that reside completely within the parapharyngeal space. For one, palpation of the tumors, which aids in fine needle biopsy can be impossible if the tumor is completely within the parapharyngeal space and not very large and shielded by the mandible. And also ultrasound guided fine needle aspiration, which is the one imaging tool that's used most commonly for FNA, can be impeded by the mandible and its shadow, and make it difficult to fine needle aspirate the tumor.

If you can palpate the tumor intraorally or externally, you could attempt fine needle aspiration without imaging studies, but most commonly I employ CT guided fine needle aspiration. The pathologists with CT guidance can approach the tumor below the angle of the mandible, sometimes transfacial, or even transorally and gain a good sample, and ensure from the imaging that their needle is going to give a representative piece of the tumor.

Dr. Linda Yin:

Let's talk about some treatment options now. So what treatment options are available to patients who have a parapharyngeal mass?

Dr. Eric Moore:

Well, here we have to go back to etiology and pathology. So the most common tumor being a benign salivary gland tumor, a pleomorphic adenoma. The most common treatment is surgical removal, just like it would be for tumors arising within the parotid gland.

The other tumors, such as lymph node etiology may just need biopsy, and then depending on whether they're lymphoma, non-surgical treatment. If they're metastatic tumor, maybe multimodality treatment combined with surgical treatment and radiation therapy.

And finally the poststyloid component tumors, paragangliomas, the tumor treatment becomes a little more heterogeneous and patient specific. So for paragangliomas you always want to keep in mind the individual patient situation, the age of the patient, the comorbidities of the patient, the site of origin of the tumor, whether it's carotid or vagal or jugular, may influence your treatment.

Most carotid body tumors, particularly in young patients without comorbidities are treated by surgical resection because this treatment yields high confidence of low recurrence rates and usually very good results. But paragangliomas of the vagus nerve, because of the deficit that you can get with high vagal injury, may be treated only with observation in some patients or with radiation therapy to try to arrest the growth, but not impair the function of the vagus nerve.

Dr. Linda Yin:

Let's talk a little bit about different surgical approaches. Our favorite topic. So what factors do you consider preoperatively in a patient when you're thinking about picking a surgical approach for the parapharynx?

Dr. Eric Moore:

So again, the most important factor to consider with surgical approach is the origin of the tumor and whether it's in that prestyloid or poststyloid area. The prestyloid tumors are usually very mobile and can be delivered out of that area well, without significant risk of injuring the great vessels or the vagus nerve. The poststyloid component tumors you need a little bit better access and proximal and distal control of those vessels during surgical procedures, and that's going to influence your surgical approach.

The common surgical approaches applied for parapharyngeal tumors are either just a transcervical approach, which is a typical incision just below the mandible, similar to a neck dissection incision, or a continuation of that incision up into the parotid area, so the so-called cervical parotid approach, to give you good access to the facial nerve if you think it's going to be in play with your tumor access. And these are the two most common approaches that we use overwhelmingly to access the parapharyngeal space.

Additional procedures may be necessary depending on the height of the tumor and its relationship to the skull base or its involvement with the great vessels. And so to gain additional access to this area, you can paralyze the patient and displace or sublux the mandible to gain additional exposure. You can divide the digastric muscle posteriorly to give yourself a larger window into the parapharyngeal space, or you can perform a partial temporal bone resection and amputate a portion of the mastoid process to gain better access, posteriorly and superiorly, into the parapharyngeal space.

The ultimate exposure to the parapharyngeal space consists of some kind of lateral or central mandibulotomy and mandibular swing, which gives you great access to the great vessels and a beautiful view of the skull base. But this is probably the most morbid approach to the parapharyngeal space, and quite honestly is rarely necessary to gain access to most of the tumors here.

And then a revisited approach, I say revisited because it was commonly used historically as the first access to the parapharynx, and then abandoned for fear of morbidity and poor exposure, but then reinvented with the advent of transoral surgery, is a transoral approach to the parapharyngeal space. And this can be a great access for those tumors in the prestyloid component that are sitting actually closer to the pharynx than they are to the neck skin and to the mandible.

Dr. Linda Yin:

Let's talk a little bit more about transoral approaches. I understand transoral robotic surgery can be used to remove some parapharyngeal space tumors? And what are some patients who are good candidates for that, and those who are not?

Dr. Eric Moore:

Yeah, patient selection is going to be really key for the success of this procedure. So first off, you want to make sure that you have a fairly mobile tumor and ideally that it's in the prestyloid portion of the parapharyngeal space to give yourself good success with the transoral approach. So the classic transoral approach to parapharyngeal tumors would be either a benign tumor, like a pleomorphic adenoma, potentially lymphadenopathy of a benign nature that you still want to access, or maybe schwannomas in this area, but in the prestyloid portion of the parapharyngeal space. And these tumors can be easily accessed by an incision through the superior lateral pallet, going through the pharyngeal constrictor muscle, and then you have direct access to the tumor.

You have very poor exposure of the carotid artery, as it's lateral and posterior to the tumor at this point. So again, you want to make sure your tumor is mobile, and malignant tumors with an infiltrating border or tumors that have been very inflamed and are immobile would not be great candidates to try to deliver through a transoral approach.

And finally, the height of the tumor can influence a transoral approach. So those tumors that extend all the way to the skull base are relatively poor candidates. And finally, those tumors that extend laterally outside the parapharynx, so the classic dumbbell tumors, are poor candidates for a transoral approach. But a completely parapharyngeal pleomorphic adenoma can be very nicely excised transorally.

Dr. Linda Yin:

We talked about the fact that there's a lot of high value real estate in this area. So what are some surgical risks that we should counsel patients on, and complications from surgical resection?

Dr. Eric Moore:

Common things that occur after parapharyngeal space tumor removal relate to the structures of the surrounding area that are left behind. So trismus is fairly common after parapharyngeal space tumor removal, because of irritation of pterygoid muscle. Fortunately, this is relatively short lived and as the inflammation subsides, the trismus usually improves specifically with physiotherapy and non-steroidal anti-inflammatory drugs.

First bite pain is an interesting phenomena of parapharyngeal space tumor removal. This comes into play a lot with removal of tumors of salivary nature, but it's thought to be related to interruption of the sympathetics from the carotid plexus, going through the parapharynx. And once those sympathetics are interrupted, the unopposed parasympathetics to the salivary gland, will give patients a very strong first rush of saliva when they first start to chew or eat, described as first bite pain. Interestingly, first bite pain is more common with cervical parotid and cervical incisions than it is with transoral incisions. So it may have a lot to do with the approach and how those sympathetics are interrupted.

Those are common things that are fortunately relatively self limited and will subside with time. You can have more significant complications such as infections in the parapharyngeal space. It's a potential space. So when you remove large tumors and don't drain that potential space, if you have that area fill up with blood, it could get contaminated mostly from venous contamination from oral bacteria, and then you can wind up with a parapharyngeal space abscess. So adequate drainage is suggested to prevent that.

Fistulas from the salivary space, particularly if you expose the parapharyngeal tumors, both transorally and externally, as sort of a hybrid approach, which has become more popular. Transoral removal are not uncommon, and it's generally advised not to make a fistulous communication with the transoral and an external incision to prevent this problem.

And finally, you could have injury to those residents of the poststyloid portion of the parapharyngeal space. So you could have injury to the carotid artery that would result in bleeding or cerebrovascular accident or pseudo aneurysm. You could have injury to the vagus nerve, which would result in dysphasia or dysphonia for prolonged period of time. You could have injury to the sympathetic trunk, which would give you Horner's syndrome and ptosis and miosis. So there are a lot of important structures back there that have critical functions and it pays to tread lightly and treat those structures gently.

Dr. Linda Yin:

All right. Well, those are all the questions that I had on parapharyngeal neoplasms. Do you have anything else that you have that you think that we should know?

Dr. Eric Moore:

That was a great session, Linda, I really appreciate doing it. And the key point is that parapharyngeal tumors are uncommon. So when you come across them, spend a little extra time thinking about the anatomy, studying the imaging, because none of the approaches will be really familiar, particularly to young surgeons.

Dr. Linda Yin:

Great. Thanks for those tips, and thanks for being here.

Dr. Eric Moore:

I really appreciate it.



Dr. Linda Yin:

Okay. Moving on to the summary section now. I'm going to sum up some key points from the talk. So parapharyngeal space tumors are uncommon. The anatomy is complex and expertise is needed for both diagnosis and treatment. The majority of parapharyngeal tumors are benign and only the minority are malignant in pathology. The most common tumors of the parapharynx are salivary gland tumors, and of these benign salivary gland tumors, so pleomorphic adenoma is the most common tumor of the space.

The parapharyngeal space is an inverted pyramid with the base at the skull base and the apex at the hyoid. Laterally it's bound by the medial pterygoid and the mandible. And immediately it's bound by the pharyngeal space, specifically the buccopharyngeal fascia and the constrictors, which are medial to that.

The parapharyngeal space can be divided into two compartments, the prestyloid, and the poststyloid compartments, and understanding of the contents of these compartments is very important to forming a differential diagnosis. The prestyloid space consists mostly of lymphatic tissue and fat, and the most common tumors of this area originate from the deep lobe of the parotid gland, so salivary gland tumors.

On the other hand, the poststyloid compartment consists of the sympathetic chain, the contents of the carotid sheath, including the internal carotid artery and the internal jugular vein, as well as the vagus nerve. And so this is a more common space for neurogenic tumors, including schwannomas or neurofibromas. This is also the common space for neuroendocrine tumors, namely a paraganglioma.

MRI is really the imaging modality of choice along with CT. And it can be helpful in understanding the relationship between the tumor and the surrounding important structures that we just named. The relationship of the tumor to the surrounding vasculature, specifically the internal carotid artery and the internal jugular vein, can really help distinguish the pathology and potentially the nerve of origin of neurogenic tumors.

Surgery is typically the preferred treatments for most of these pathologies, maybe outside of a paraganglioma, and several surgical approaches are possible. Prestyloid tumors can often be accessed with just a transcervical approach or a combined transcervical or parotid approach, in which the parotid gland is also removed, and the facial nerve is identified and preserved. For additional access the tip of the mastoid can also be removed, and that's a transmastoid approach. And the mandibulotomy can rarely be used to again, gain greater access.

Transoral approaches, including transoral robotic surgery are becoming more popular. And these are good for medially based tumors that are very well separated away from the neurovascular bundle, particularly the internal carotid artery.

Preoperatively patients should be counseled on the risks of operating in the parapharyngeal space, and this can include more common risks such as first bite syndrome, which comes from disruption of the sympathetic innervation to the parotid gland that travels along the carotid. Or less common risks, including injury to the contents of the carotid sheath, including injury to the internal carotid, the vagus nerve, which can end in dysphonia or dysphasia, as well as Horner's syndrome from injury to the sympathetic trunk.

Okay, well, we're going to do some questions now. So I'll be giving some questions that sum up some key points from the talk. And then I'll give you a brief second to think about the answer, and then I'll provide the answer.

So, first question is, what is the most common tumor of the parapharyngeal space?

So the most common tumors of the parapharyngeal space are benign tumors of salivary gland origin, and typically this is a pleomorphic adenoma.

Next question. What are the two compartments that make up the parapharyngeal space and what separates them?

The parapharyngeal space can be separated into the prestyloid and poststyloid compartments. And as the name suggests, this is separated by the styloid process, as well as the fascia that extends from the styloid into the [inaudible 00:27:49] pharynx.

What are some important contents of the poststyloid space?

The most important contents of the poststyloid space include the contents of the carotid sheath, namely the common carotid artery, the internal jugular vein, and the vagus nerve. Lower cranial nerves can also run through this area as well as the cervical sympathetic chain.

What is a dumbbell sign? And what does it mean on imaging?

The dumbbell sign is very classic for a deep lobe parotid tumor that is extended into the parapharyngeal space. The middle or the narrow part of the dumbbell is due to compression from the styloid mandibular ligament.

What is the most commonly used surgical approach to resect prestyloid tumors?

Prestyloid tumors, especially benign ones, that can easily be separated from the surrounding structures can usually be accessed with a transcervical approach.

All right, that's the show. Thank you for listening, and be sure to tune in to future episodes.