

Dr. Linda Yin:

Hi there and welcome to another episode of ENT in a nutshell. My name is Linda Yin and I will be your host. I am joined today by Dr. Paul Gidley, who is a neurotologist. Dr. Gidley, thank you so much for being here.

Dr. Paul Gidley:

Oh, it's a pleasure to be here.

Dr. Linda Yin:

Before we start the episode, I want to clarify for the listener that we'll be speaking exclusively here about temporal bone squamous cell carcinoma. So we'll be focusing on adults and specifically on this pathology. Let's start with the presentation. What is the typical presentation of a patient that comes into your clinic with a temporal bone squamous cell cancer?

Dr. Paul Gidley:

Typical presentation is somebody who has pain or drainage from their ear. And oftentimes they complain of pain and drainage for a long time, perhaps even up to a year before they finally get diagnosed as having a cancer involving the ear canal. Other signs can be hearing loss due to blockage of the ear canal from tumor. And then far advanced tumors may present with facial paralysis or sometimes lower cranial nerve deficits. Occasionally, these patients will also come in with lymphadenopathy either in their parotid gland or in their upper neck.

Dr. Linda Yin:

When you're evaluating patients that may have a temporal bone cancer, what kind of physical exam maneuvers would you perform?

Dr. Paul Gidley:

We do a very careful head and neck evaluation. Of course, we spend a lot of time looking at the ear. Oftentimes the ear examination can be very limited though. If you've got a cancer that completely fills the ear canal, there's not much to be seen other than tumor filling the ear canal. On the other hand, with smaller limited tumors, it's good to look at the tumor and decide if this is a tumor involving just the membranous ear canal, or if it involves both the membranous and bony canal. Then beyond the ear, we want to look at cranial nerve examination very carefully. Obviously, if somebody presents with facial paralysis, this is an important prognostic finding, but if they have lower cranial nerve deficits, this is also a very important finding. And then we spend time looking to see if there's any lymphadenopathy either in the parotid gland or in the neck.

Dr. Linda Yin:

Can you tell me a little bit about the epidemiology of a temporal bone squamous cell carcinoma? How common is it?

Dr. Paul Gidley:

Temporal bone squamous cell carcinoma is rare. The incidence is estimated to be about one case per million, maybe up to six cases per million, but very uncommon for primary tumors of the ear canal or temporal bone. In my practice, the most common cause of a tumor involving the ear canal and temporal

bone is actually from a tumor outside of these locations, usually from a parotid tumor or a preauricular skin cancer.

Dr. Linda Yin:

And what does that typical patient that presents with a temporal bone squamous cell look like? Are they elderly or are they a certain sex or gender?

Dr. Paul Gidley:

Roughly 75% of the patients did have temporal bone cancer are men. It usually affects people in their 60s, 70s, 80s.

Dr. Linda Yin:

Interesting. We'll jump into pathophysiology a little bit later too. Let's wrap up the presentation. So when a patient comes into your office and has some of these signs and symptoms, you might be thinking of squamous cell carcinoma, but is there anything else that should be on your differential diagnosis to rule out?

Dr. Paul Gidley:

Well, we talked about benign diseases. So of course, otitis externa, chronic otitis media, skull base osteomyelitis, you might think also of cholesteatoma.

Dr. Linda Yin:

We talked about the differential for general signs and symptoms that may be similar to a temporal bone malignancy, but how about a pathology differential? In addition to a squamous cell cancer, what kind of other pathologies can you see that are malignant in this area?

Dr. Paul Gidley:

Squamous cell cancer is definitely the most common that is seen, and depending on the literature, it probably accounts for 60 to 80% of ear canal cancers.

Second most common is basal cell carcinoma, at least in our series. And of course this seems linked to sun exposure.

Third most common is adenoid cystic carcinomas. And adenoid cystic carcinomas are very unusual. They usually present with pain, ear pain and patients will have ear pain for up to two years before these cancers get diagnosed. And this is because adenoid cystic cancers tend to be subcutaneous and very small, but very difficult to diagnose.

Probably the next most common cancer we see in the ear canal is melanoma, but there are several other very rare tumors that can involve the ear canal or temporal bone. Diagnosis depends on biopsy.

Dr. Linda Yin:

Before we jump too much into disease pathophysiology, can you tell us a little bit about some basic anatomy of the temporal bone and things that we should understand and pay attention to when understanding how diseases in this area can spread?

Dr. Paul Gidley:

Sure. This is really an important topic. Cancers that occur in the ear canal can spread through natural fissures in the cartilage of the ear canal. These are the fissures of Santorini. There are other immunologic openings such as the foramen of Huschke. And then you think about natural pathways of spread. So once a cancer is through the eardrum, it spreads through the middle ear and mastoid or eustachian tube. If it passes through the ear canal superiorly, obviously it will come into the middle fossa. If it passes through the bony canal anteriorly, it'll be in the temporomandibular joint. If it passes through the ear canal posteriorly, it may be into the facial nerve and mastoid. And then if it passes through the ear canal inferiorly, it will reach the jugular foramen and carotid artery.

Dr. Linda Yin:

What is the typical site of origin for squamous cell carcinomas that involves the temporal bone?

Dr. Paul Gidley:

Squamous cell cancers can arise inside the ear canal, either the membranous canal or the bony canal. Squamous cell cancer has been described on the eardrum and in the middle ear or mastoid. And so these are primary sites that involve the ear canal and temporal bone. In my practice, I actually see more tumors that come from external ear or preauricular skin that involve the ear canal. So these are usually neglected cancers that have grown to be very, very large. And then I also see a number of patients that have parotid malignancies that grow either into the ear canal or they grow along the stylomastoid foramen and into the temporal bone through that route.

Dr. Linda Yin:

Does it matter where the squamous cell cancer is coming from in terms of prognosis?

Dr. Paul Gidley:

Squamous cell cancer from any one of these primaries is dangerous. However, it seems that the tumors that are confined to the bony canal tend to have a much better outcome than tumors that come from preauricular skin. And this is probably because the tumors that are confined to the bony canal can easily be removed with en bloc resection. Patients that come in with preauricular skin cancers, often we see these as recurrent skin cancers, and they're very difficult to treat.

Dr. Linda Yin:

Are there any risk factors that would predispose someone to getting a temporal bone squamous cell cancer?

Dr. Paul Gidley:

Risk factors really haven't been fully elucidated. Of course, squamous cell cancer if it involves the outer ear or periarticular skin is usually related to sun exposure. Historically, if you look back at 19th century and early 20th century articles on temporal bone cancer, there's mention made about chronic otitis media and chronic mastoiditis as being precipitating events. More recently, there has been a question raised at least in the Japanese literature about ear cleaning, especially aggressive ear cleaning, maybe with Q-tips that might produce chronic inflammation, chronic irritation in the ear canal. And then lastly is radiation. So radiation such as for nasopharynx cancer can cause and been associated with squamous cell cancer in the ear canal.

Dr. Linda Yin:

Let's move on to talk about some workup now for this disease. After evaluation and clinic, and you're suspecting a cancer in the ear canal, what is the best imaging study to get?

Dr. Paul Gidley:

I like to start off with CT scan. The CT scan will show not only soft tissue, but bony anatomy. It also gives very good picture of parotid and upper neck anatomy. So I usually start off with CT scan. If I'm worried about perineural invasion or dural involvement, then I would get a contrast-enhanced MRI.

Dr. Linda Yin:

When you're looking at an image, can you walk us a little bit through your thought process and kind of areas to pay attention to?

Dr. Paul Gidley:

Sure. I try to be very systematic about looking at the ear canal and think about the ear canal and four quadrants: superior, anterior, inferior, and posterior. I look at middle ear and mastoid. I want to look at facial canal. I look at the temporomandibular joint. I look at middle fossa. I look at the jugular foramen and carotid canal.

Dr. Linda Yin:

So you mentioned biopsy as well. Are you typically biopsying these cancers in clinic if you see a tumor in the ear canal, or do you wait at the time of surgical resection and just biopsy to confirm before proceeding?

Dr. Paul Gidley:

So depends on the location of the tumor, but tumors that on the outer ear or in the ear canal can often be biopsied in clinic with cup forceps or punch biopsy. Tumors that are in the middle ear will require operative intervention. So those would be done in the OR, and typically as a separate procedure from surgical resection. And I do that because I want to have a discussion with the patient regarding what the biopsy is showing and what the treatment will be to fix it.

Dr. Linda Yin:

I understand that these tumors are staged in a special way that some listeners may not be familiar with. So is there a special staging system for temporal bone squamous cell cancers?

Dr. Paul Gidley:

There is not an approved AJCC staging system for temporal bone cancers. And so the staging system developed by Pittsburgh has become the standard in the literature for the last 20 or 30 years.

Dr. Linda Yin:

Can you walk us through a little bit of the Ts aging in the Pittsburgh system?

Dr. Paul Gidley:

T1 tumors in the Pittsburgh staging system are tumors that are limited to the ear canal without any bony erosion or soft tissue involvement. T2 tumors are tumors that are limited to the ear canal with bony involvement, but not full thickness and limited soft tissue involvement less than half a centimeter. T3 tumors erode through the ear canal with limited soft tissue involvement, less than half a centimeter, or involving the middle ear or mastoid. T4 tumors are large tumors that erode into the cochlea, the petrous apex, the medial wall of the middle ear, carotid canal, jugular foramen or dura and have extensive soft tissue involvement such as the temporomandibular joint or styloid process, or they have evidence of facial paralysis.

Dr. Linda Yin:

What is the risk of nodal spread for an ear canal squamous cell?

Dr. Paul Gidley:

The incidents of lymph node involvement is actually low. Depending on the study that you look at, it's maybe between 10 and 20% of patients, but when lymph node involvement is identified, this is a sign of an advanced tumor. And in fact, in the Pittsburgh staging system, in positive patient is considered a stage four tumor.

Dr. Linda Yin:

Can you talk a little bit about T stage and how it relates to nodal involvement? Is there any particular T stage that would make you more worried about having to look for nodal disease and getting a scan of the neck?

Dr. Paul Gidley:

Well, you can have nodal disease at any T stage. Of course, with farther advanced tumors, you would think that the lymph node involvement is going to be slightly higher, but we do see patients with large T4 tumors who have no lymph node involvement.

Dr. Linda Yin:

Let's talk a little bit about treatment and management of this disease. Typically for head and neck cancers, I know we have surgery, radiation, and chemotherapy. Which of these is the best treatment for a temporal bone cancer?

Dr. Paul Gidley:

I like to think about the treatment based on staging and patients that have T1 or T2 tumors generally have a tumor that's confined to their ear canal. And these patients I would treat with surgery, usually it's lateral temporal bone resection with parotidectomy and neck dissection. Larger tumors, such as T3 tumors, usually involve the middle ear and they will need to have subtotal temporal bone resection with parotidectomy neck dissection. And then larger tumors, T4 tumors generally will require total temporal bone resection. Maybe that would also be a combined with resection of temporomandibular joint or mandibular condyle, perhaps combined with craniotomy and then parotidectomy and neck dissection. That's surgery.

Many of these patients, especially if they have bony involvement, perineural spread, recurrent tumors or lymph node involvement will need radiation postoperatively. More recently for large tumors,

and I'm talking about T3 and T4 tumors, a role is evolving for upfront chemotherapy or treatment with chemotherapy and radiation.

Dr. Linda Yin:

Dr. Gidley, I want to follow up and make sure I understand. So you mentioned parotidectomy and neck dissections. Is this something that you're performing on all patients that undergo surgery, or are there specific stages or disease features that would make you more inclined to do that?

Dr. Paul Gidley:

That's a great question and a very controversial subject. Obviously if a patient has a parotid or a neck mass, that person needs to have parotidectomy and neck dissection. But as we mentioned previously, many of these patients don't have lymph node involvement. And as we discussed earlier, there can be direct extension into the parotid gland from your canal cancers. And so in my view, doing parotidectomy and neck dissection for all patients is important staging and I believe this to be true, especially for tumors which are being clinically staged T1 and T2. Even though the incidents of parotid disease and neck disease may be very small, for these patients, if the parotid and neck are without any disease, they may not need additional surgery specifically for T1 tumors. They may not need additional therapy such as radiation therapy, but if the parotid or lymph node dissection identifies metastatic disease, then this patient has more advanced disease than was clinically evident and they need to have adjuvant therapy. So for me, parotidectomy neck dissection are important for staging.

Dr. Linda Yin:

You spoke about several different types of temporal bone resections. So can we start with a lateral temporal bone resection? What does that mean and what are you doing in the surgery for that?

Dr. Paul Gidley:

Okay. The goal of lateral temporal bone resection is en bloc removal of the ear canal. To do that I start off with a complete mastoidectomy. I open up the attic to see the ossicles and then I drill away the root of the zygoma and follow between the dura and the ear canal until I've exposed all of the temporomandibular joint. And that really is the anterior, superior extent of the dissection. After that's completed, then I open a wide facial recess and I will follow the facial nerve out to the stylomastoid foramen. I usually remove the mastoid tip, and then I'm going to extend the facial recess along the facial nerve and drill away the inferior tympanic ring. The idea here is to stay between the facial nerve and the annulus, follow the annulus inferiorly until you reach the temporomandibular joint. And that's the interior inferior extent of the dissection.

Once that's completed, then I disarticulate the ossicular chain by removing the incus, divide the tensor tympani tendon, and then the ear canal is being attached by the bony annulus anteriorly in soft tissue. And so a little bit of thumb pressure on the ear canal allows it to fracture off anteriorly, and then the ear canal can be separated from parotid and the temporomandibular joint in the canal can be removed as one en bloc unit.

Dr. Linda Yin:

Going a little further now. How about a subtotal temporal bone resection? What further steps are required to accomplish that?

Dr. Paul Gidley:

Sure. Subtotal temporal bone resection could be done in two ways. It could be done en bloc or piecemeal, and there hasn't been any evidence that one is better than the other. I think the morbidity is slightly higher if you do it en bloc versus piecemeal. So I tend to do it as a piecemeal dissection. I begin that with a lateral temporal bone resection. And so the ear canal is removed. Once that's out of the way, then I will remove the labyrinth and the cochlea. And then a decision needs to be made about how to handle the facial nerve. If you've got cancer that's involved in the facial nerve and the patient presents with facial paralysis, the decision is fairly easy to go ahead and sacrifice the facial nerve and try to get a margin on the nerve. The more difficult intraoperative decision is if that nerve looks completely normal and the patient has normal facial function, how much resection would you do? And this is a difficult decision to make.

Dr. Linda Yin:

While we're on the topic of the facial nerve. So say you do have to sacrifice the facial nerve either due to gross disease or high suspicion of perineural invasion. If you're assessing the facial nerve, are you performing any sort of reconstruction of that nerve at the time of surgery?

Dr. Paul Gidley:

Sure. Patients that come in that have recently [inaudible 00:23:31] facial function, nerve grafts can be performed. We try to get a negative margin on the facial nerve. And if there's good segment inside the mastoid or middle ear and good segments are found extra temporarily, then a nerve graft can be performed. And we've found that even in patients that receive postoperative radiation therapy, they still benefit from a nerve graft and we'll still get nerve function.

For the patients that present with facial paralysis, these patients don't do well with nerve grafting. And so we would tend to rehabilitate that patient either with slayings or brow lift or eyelid surgery to handle their facial paralysis.

Now, a more difficult situation can be trying to get a negative margin on the facial nerve. And this topic is controversial. You could chase the facial nerve into the geniculate really through a mastoid type of approach, but to reach more proximal segments of the facial nerve, you would need to do labyrinthectomy. In my practice, I do not do labyrinthectomy to try to achieve a negative margin on the facial nerve. And that's because it's an added morbidity to the patient, but more to the point, those patients generally do not recur in their facial nerve or in the cerebellopontine angle. Those patients generally have far advanced disease and they usually recur either in their local site or in their neck or with distant disease.

Dr. Linda Yin:

And finally, let's talk about a total temporal bone resection. What surgical steps are required for that?

Dr. Paul Gidley:

Well, temporal bone resection, again, could either be performed en bloc or piecemeal. And again, I do that as a piecemeal resection, and it's very similar to what we talked about for subtotal temporal bone resection, remove the ear canal. Then the labyrinth and cochlea patients who have T4 disease, which is the setting for total temporal bone resection, they already have facial paralysis. So the facial nerve is going to be sacrificed. The internal auditory canal is removed and dissection continues into the anterior petrous apex. And that really marks the medial extent of the dissection. These patients who have T4

disease often have carotid artery involvement and that becomes a difficult situation in terms of managing carotid artery.

Dr. Linda Yin:

I imagine for some of these recessions, you're left with a pretty big surgical defect afterwards. How do you approach the defect when you think about reconstructing and rehabilitating the patient?

Dr. Paul Gidley:

It depends on the size of the defect. Patients who undergo lateral temporal bone resection, even with parotidectomy and neck dissection, many of those patients can be reconstructed with a temporalis flap. Patients that lose their outer ear generally will require a free flap. And then this would be true for patients with even larger resection, such as subtotal temporal bone resection, or total temporal bone resection. We frequently ask our plastic surgeons to help us with a microvascular free flap to reconstruct.

Dr. Linda Yin:

What is the risk of a CSF leak during these types of procedures and how do you manage that intraoperatively?

Dr. Paul Gidley:

CSF leak is actually fairly uncommon, especially if the surgery is limited to lateral temporal bone resection. Now, patients that have subtotal or total temporal bone resection, obviously the defect is much larger. If a dura is resected, we try to aim for primary closure of the dura using dural grafts. The most important factor for preventing CSF leak is to try to use soft tissue to hold CSF intracranially, and then use soft tissue to block off any areas of egress. So that could be not only through the station tube or through the ear canal or through the incision site.

Dr. Linda Yin:

How about hearing rehabilitation? What options do you have for these patients after surgery?

Dr. Paul Gidley:

Obviously with a lateral temporal bone resection, you're removing the sound conducting mechanism of the ear, and patients will have a maximum conductive hearing loss. Fortunately, doing this surgery, we usually do not create much sensory neural loss. So if these patients have good preoperative hearing, we can make use of osseointegrated implants to rehabilitate hearing.

Now, for patients that have subtotal or total temporal bone resection, they develop complete unilateral hearing loss. Some those patients can be rehabilitated with osseointegrated implants if they so choose. Many of them decide that unilateral hearing is enough for them to do their normal daily activities.

Dr. Linda Yin:

Finally, what is your typical follow-up and surveillance regimen for some of these patients after surgery?

Dr. Paul Gidley:



Patients will have surgery and then we see them postoperatively and discuss the pathology findings. If the pathology findings indicate signs of aggressiveness, such as bone or cartilage invasion, perineural invasion, lymph node involvement, then those patients will go on to get postoperative radiation therapy with or without concurrent chemotherapy. That lasts about six weeks and we see the patients after that completed that therapy. And we start them on every three month routine followup and routine followup would include a CT scan plus physical examination and history. We do that for the first two years. After that we see them for six months, until five years. Patients that make their five-year anniversary are then enrolled in survivorship.

Dr. Linda Yin:

Okay, well, those are all the questions that I had on temporal bone squamous cell cancers. Is there anything that we've missed or anything that you want to emphasize for the listener?

Dr. Paul Gidley:

No, I think you've covered it very comprehensively.

Dr. Linda Yin:

Well, thank you so much for being here, Dr. Gidley. It was a pleasure to speak to you.

Dr. Paul Gidley:

It's a pleasure to be with you. Thank you.

Dr. Linda Yin:

Okay. Let's start the summary section now.

So temporal bone squamous cell carcinomas are rare tumors and they typically present with a triad of symptoms that are also common to other ear pathologies, including otorrhea, otalgia and hearing loss. Careful examination including otoscopy will be important in helping form the differential diagnosis and raises suspicion for malignancy. Other pathologies that should be on the differential diagnosis include benign ear pathologies like chronic otitis media or otitis externa, as well as other malignant tumors in this area, including parotid tumors, such as an [inaudible 00:32:18] carcinoma and other skin cancers, such as a basal cell or a melanoma.

The temporal bone has several innate weaknesses that can make potential routes of spread for malignancy. This includes the foramen of Huschke, as well as the fissures of Santorini. It's important to remember temporal bone squamous cell cancers can start in the temporal bone, namely the external auditory canal or the middle ear and the mastoid, or they can extend into the temporal bone from nearby structures, including the preauricular and the auricular skin.

The best imaging workup initially to obtain for a temporal bone squamous cell cancer is going to be a CT scan. This helps us visualize the bony anatomy as well as the soft tissue anatomy of the product and neck. MRI can also be important for dural involvement as well as perineural invasion.

Temporal bone squamous cell cancers are staged using a special system outside of the typical AJCC staging system, and this is called the Pittsburgh staging system. In it the T stage determines the local spread of the tumor. And this can range from tumors that are confined to the external auditory canal, which is a T1 tumor with no bony erosion, to tumors that are involving significant other

structures, including the cochlea, the petrous apex, the carotid canal, the jugular foramen, the dura, the facial nerve. And that will be a T4 tumor.

The best treatment for a primary temporal squamous cell cancer is going to be surgery. Surgery will involve a temporal bone resection, most commonly just a lateral temporal bone resection. Dr. Gidley also recommends performing a superficial parotidectomy as well as neck dissection at the time of tumor resection for staging and for treatment purposes.

Facial nerve management during a temporal bone resection can be tricky. Every attempt should be made to get clear margins on the facial nerve. If negative margins are attainable, even if the patient gets post-operative radiation, then a primary nerve graft can be attempted.

Surgical defects can be reconstructed after a temporal bone resection with a combination of local flaps, such as the temporalis flap, autologous grafts for any CSF leaks, and of course, microvascular free flaps for any larger defects.

Bone-anchored hearing aids, osseointegrated hearing aids can be used to rehabilitate hearing after a temporal bone resection. In the case of a lateral temporal bone resection, this will help with the maximum conductive hearing loss the patients experience after. And in the case of a subtotal or total temporal bone resection, there will be a complete sensory neural hearing loss as well, but sometimes the osseointegrated hearing aid can be used for unilateral hearing loss.

Let's move on to the question section now. I will be asking a few questions to summarize some key points from the talk. I will give a brief pause afterwards, and then the answer to the question.

First question. What are some risk factors for getting a temporal bone squamous cell cancer?

Historically it was thought of that perhaps there's a relationship between chronic ear disease and temporal bone malignancies, or recently studies have looked at the potential link between frequent picking and trashing of the ear canal and that chronic trauma as a risk factor for getting a temporal bone squamous cell cancer. Previous radiation therapy can also be a risk factor.

What is the most common malignancy that is found primarily arising out of the temporal bone?

The most common pathology found in primary temporal bone cancers is squamous cell carcinomas, which is the subject of this talk.

What are the weak points in the temporal bone that could be potential routes of spread of a temporal malignancy?

Potential weak points of spread in the temporal bone include the foramen of Huschke, which is an opening in the external auditory canal into the infratemporal fossa due to incomplete ossification of the tympanic ring, as well as the fissures of Santorini, which are naturally occurring defects in the cartilaginous portion of the external auditory canal.

What does a T3 tumor look like in the Pittsburgh staging system for temporal bone squamous cell cancers?

A T3 tumor is a tumor that has eroded through the osseous external auditory canal with full thickness bone of version, with limited soft tissue involvement or tumor that has gone past the tympanic membrane into the middle ear and mastoid.

Which structures are preserved in a lateral temporal bone resection?

In a lateral temporal bone resection, the stapes and the facial nerve are preserved. Removal of the labyrinth and cochlea does not occur unless you are doing a subtotal temporal bone resection. That's our show. Thank you for listening and tune in next time.