

Dr. Alyssa Smith:

Hello, everyone. Welcome to another episode of ENT in a Nutshell. My name is Alyssa Smith and today, we're joined by pediatric otolaryngologist Dr. Raj Petersson. In this episode, we'll be discussing thyroglossal duct cysts. Thanks for being here, Dr. Petersson.

Dr. Raj Petersson:

Thanks for having me. Happy to be here.

Dr. Alyssa Smith:

I think the first thing that we should focus on is presentation for these patients. What signs or symptoms does a patient with a thyroglossal duct typically present with?

Dr. Raj Petersson:

Usually, they show up with a midline neck mass that could be asymptomatic or may have gotten infected at some point as the first presentation. We usually don't see them right at birth or during infancy, but it might be some years after and they might notice a lump there that didn't go away or sometimes, they may present to the ER or their pediatrician with a red swelling that's infected that raises the suspicion of a thyroglossal duct cyst.

Dr. Alyssa Smith:

You mentioned that patients can present young. What's kind of the age range that patients can present with?

Dr. Raj Petersson:

I usually see kids who are like a few years old. I mean, sometimes we'll have two-year-olds. I think it's rare to see them below that age. Lot of times, they might be like three, four, five, but they can present in adulthood as well, although that's more rare. Most of these are going to present some time in young childhood.

Dr. Alyssa Smith:

How common are thyroglossal duct cysts?

Dr. Raj Petersson:

They're the most common congenital neck mass in children and the prevalence is estimated at 7%. It's something we see pretty commonly in our clinic. They account for about a third of all congenital neck masses in kids.

Dr. Alyssa Smith:

And then focusing on the pathophysiology behind this, how exactly does it form?

Dr. Raj Petersson:

Sure. The thyroglossal duct cyst happens when you have a persistent tract left from the descent of the thyroid gland. The thyroid gland develops at the base of the tongue in a area called the foramen cecum at about three weeks of gestation, and then it'll descend down the midline before the hyoid cartilage

being formed in an area that would be anterior to the location of the expected hyoid bone, and the thyroid gland then goes down to its normal position below the thyroid cartilage.

If any part of this tract remnant does not involute, which is usually at about 10 weeks of gestation, you may get a thyroglossal duct cyst. These can present anywhere along that tract. Generally they're in front of the hyoid bone, but they can be below it, just above the thyroid gland. They can be above the hyoid bone, more towards the tongue base.

Rarely, we may see ones that aren't palpable at first because they're deeper in towards the tongue base. That foramen cecum is generally at the anterior two-thirds and posterior one-third junction of the tongue. It's behind the circumvallate papillae there. If this normal involution doesn't occur, we can get a thyroglossal duct cyst.

One reason that we think this may not appear right away at birth or early on is that foramen cecum might be open into the mouth and if that's the case, the fluid that may develop in that cyst would just kind of drain out through the mouth. If that foramen cecum closes, then that fluid will consolidate in the cyst and then you'll see the cyst. That could happen if it got infected or inflamed, for some reason.

Dr. Alyssa Smith:

So thinking about the workup of these patients, if someone presents with a midline neck mass, what should be on our differential diagnosis?

Dr. Raj Petersson:

For me, the top three that I think of is a thyroglossal duct cyst, a dermoid cyst and a lymph node. Those are going to be by far the most common. That's what we're usually going to think about. But, we also need to consider other possibilities, which could be some sort of a vascular malformation or a hemangioma, although these would be rarer in that location. A hemangioma, you would likely see some skin changes with it as well. And we talk about a branchial cleft cyst potentially arising in this location, and that differential might be more of a possibility if the thyroglossal duct cyst is a little more lateral than midline. It may be a branchial cleft cyst. It could be an epidermoid cyst. Other rare things, a laryngocele or lymphoma or a metastatic lymph node, with probably papillary thyroid cancer being the most common in this location just because of where it is.

But most commonly, it's going to be thyroglossal duct cyst, dermoid cyst and a reactive lymph node. Other rare infectious possibilities could be atypical mycobacteria but most of the time, those are going to be much lower down the list but important to think about.

Dr. Alyssa Smith:

With that in mind, what are some questions that you ask either the parents or the patient when you're gathering a history to kind of figure out exactly the specific diagnosis?

Dr. Raj Petersson:

I ask them how long it's been there, does it get bigger and smaller, has it ever been infected with signs of redness, has it ever drained into the neck, how often it might get bigger or larger or get infected. We also want to know if there's any family history of head and neck cancers or masses, especially thyroid cancers or lymphomas, going back to our differential. But really, it's sort of the duration, any infection history, does it bother them, do they have trouble swallowing, any trouble with the airway or breathing.

Dr. Alyssa Smith:

And then the next step would be our physical exam. What are some key findings that we're looking for?

Dr. Raj Petersson:

Typically, it's going to be a mobile neck mass in the midline, generally right around the hyoid bone. But like I said, it could be higher or lower from there, but usually in the vicinity. It's generally not fixed. You may see it sometimes elevate if they swallow or stick out their tongue, but this isn't very specific so if you don't see that, it doesn't rule it out. There's generally not a sinus or a drainage or a [inaudible 00:06:11] tract, but this could happen if it's been repeatedly infected and it's opened up into the skin. Those are the key things. Generally, they're painless unless it's infected. They're usually not tender, again, unless it's infected.

Dr. Alyssa Smith:

And then is there any role for radiology for these patients?

Dr. Raj Petersson:

When I consider radiology for these, it's usually not to examine the cyst itself, but to see what's in the surrounding. Really, the main thing for suspecting a thyroglossal duct cyst is to make sure there's normal thyroid gland in the area. Once that's confirmed, then you can get some more information with an ultrasound, is what we usually start with, to talk about the characteristics of the cyst itself.

The key points with the ultrasound are, is there a normal thyroid gland and then some characteristics about the cyst itself. It can tell us if it's cystic or it might tell us if it looks more like a lymph node. And then it is also important when I order these to get an ultrasound of the lateral neck as well to make sure that there's not any other lymphadenopathy that we might be missing in that area.

In general, I don't get a CT scan for these or any other imaging unless there's something weird about the characteristics of the cyst. I might get a CT scan if it's in a strange location, the size might be unusual. Sometimes if they're much bigger, then you really want to know the extent of it and so a CT with contrast could be helpful. An MRI can also be helpful, but most young children would need anesthesia for an MRI so we generally start with the CT with contrast and generally get enough information from that. But once we have the ultrasound and we're still suspecting it's a thyroglossal duct cyst, we don't need to do further imaging in general.

Dr. Alyssa Smith:

So moving on to treatment, do all patients need to be treated or can you sometimes go along the route of more conservative management?

Dr. Raj Petersson:

We generally recommend surgical treatment for these. The main reason for that is to prevent it from getting infected because likely, they will get infected at some point and once they are infected, they're harder to remove and prevent recurrence. We usually recommend surgical excision, and the procedure for that is called the Sistrunk procedure, which was actually developed in 1920, so a hundred years ago, by Dr. Walter Sistrunk at the Mayo Clinic in Rochester. This procedure is what allows us to remove as much of this as possible and decrease any chance of recurrence.

Dr. Alyssa Smith:

Can you briefly talk about exactly what is involved in the Sistrunk procedure and what differentiates this versus just a simple excision?

Dr. Raj Petersson:

Sure. A Sistrunk procedure involves removing the cyst as well as the entire expected tract of the thyroglossal duct cyst. Without removing all of it, we know that these will recur. First thing we do, in general, is I make a small incision overlying the cyst. It might be just under it or below it. I try to hide it in neck crease so that it'll heal well. The incision is generally small, it's about two to three centimeters, and we go down through the skin and subcutaneous tissues until we identify the strap musculature and divide that in the midline if we encounter that along the way. Once we get to the cyst wall, we try to be really careful about not getting into the cyst so we don't spill the contents and we can still find and follow it. Sometimes, I'll take a cuff of normal tissue around it, especially if they're really sticky if they've been infected before.

Once we get down to the cyst, we kind of follow in the midline along the muscles up to the hyoid bone, and I start by working on dissecting off the musculature in the central portion of hyoid bone because we do want to take that middle portion of the hyoid bone. When Sistrunk first described this, he described taking about a quarter inch of bone. In reality, I probably take about a centimeter, and that's because the tract is usually not just a single tract. There could be fingers reaching out from it, so that kind of ensures that we're getting all of it. We dissect down to the hyoid bone and expose the inferior and superior surface in that middle centimeter or so, and strip all the muscle off of it so we can get to that bone and then we're usually able to snip that bone with either heavy scissors or maybe some small bone snippers.

Once that's done, I kind of work from inferior to superior. We know that tract is expected to occur along the midline going through the mylohyoid into the genioglossus, so we'll take a cuff of muscle and tissue around the expected location of the tract as we follow it towards where the foramen cecum would be. At this point, I find it's helpful to have an assistant put a finger in the mouth and push down at the base of the tongue in the location of the foramen cecum to push it down into the field of surgery. And once we follow it up into there, we have to take a cuff of the tongue base musculature and then what I'll do is I'll ligate it there with a silk suture and then we'll cauterize it off and try to deliver that all in one specimen.

Dr. Alyssa Smith:

You mentioned previously that patients can present kind of in the setting of an acute infection or inflammation of the cyst. If they do present with this acute inflammation, is that the time to take them to surgery or do you kind of treat with antibiotics and let it die down a little bit?

Dr. Raj Petersson:

No, we definitely don't want to go right to surgery when it's infected. We do try to treat it with antibiotics and ideally, you would calm it down with just antibiotics and then maybe two to three weeks after the antibiotics are done, go back to the operating room. Now, we want to get there before it potentially gets reinfected so I usually don't wait too long, but we give it a few weeks.

Now sometimes, they may get so enlarged that they might need a drainage. If that occurs, my preference is to not to do a formal incision and drainage because that just causes more scarring in the area. This is a case where we would try to control it with needle aspiration and antibiotics, if we can.

Dr. Alyssa Smith:

And then thinking about our preoperative workup, we discussed using the ultrasound to determine if there's any functional thyroid tissue present in its normal location. If there isn't any functional thyroid tissue present, what does that mean as far as treatment goes, and is surgery still indicated?

Dr. Raj Petersson:

It definitely makes us consider what we're doing in terms of surgery. It's a rare finding so I've not ever found that in any of my patients, but surgery is still indicated but it may give you some pause. If the patient has never had an infection, you may watch it for a while. You would probably want to do a thyroid hormone level workup and make sure the numbers are okay there, but what we don't ever want to do is remove that without ever knowing about the thyroid status.

However, if it's a cyst that is getting infected or repeatedly infected, I think we need to move ahead and remove it, but then we have the knowledge that there is no thyroid tissue identified and they may need to go on thyroid replacement hormone. At that point, I would get my endocrinology colleagues involved as well to weigh in and help with management. One reason to remove these is that there is a potential chance of developing carcinoma within the thyroglossal duct cyst, so that is one of the reasons that we would go ahead and remove it anyways.

Dr. Alyssa Smith:

In thinking about the complications from surgery, what are some that residents should be aware of when managing these patients postoperatively, as well as ones that you discussed with parents preoperatively?

Dr. Raj Petersson:

The biggest risk of surgery is recurrence. This is the thing that's most commonly going to happen so I counsel them about that. I usually tell them that the risk of recurrence is about 5% or less with the Sistrunk procedure. We also talk about hypoglossal nerve injury which can occur as well, especially if we get too lateral on the hyoid bone. To prevent that risk, we definitely stay close to the central portion of the hyoid bone where the expected tract is, and we try not to dissect aggressively laterally. So I stay within that one centimeter central portion.

Dr. Alyssa Smith:

And then thinking about follow-up, what is the follow-up schedule typically look like?

Dr. Raj Petersson:

I usually follow them up about two to three weeks after surgery, and that is more of a wound check to make sure they healed up appropriately. I may see them three months later and then maybe six months after that, but if they haven't recurred, then I don't necessarily do regular follow-ups with them. But I do tell them to call the office back or come back in if they notice a new growth in the area or they feel that it's coming back.

Dr. Alyssa Smith:

If they feel like there's a new growth or it's coming back, would you repeat an ultrasound or maybe go straight to surgery? What does that management look like?

Dr. Raj Petersson:

If it's fairly obvious on physical exam, we may just go right to surgery. At times, I will repeat an ultrasound just to confirm or make sure that it's still looking like a cyst and it's not something else going on that might require further imaging. A lot of it's going to depend on the history and physical exam of what we do there.

Dr. Alyssa Smith:

And then thinking about the natural history of these lesions, what if no treatment is pursued?

Dr. Raj Petersson:

If no treatment is pursued, like we alluded to, there is a risk of cancer developing within these. It's not very common. Less than 1% of the time, they may develop into a carcinoma, but it is a risk that is described that we know about.

Dr. Alyssa Smith:

All right. In summary, thyroglossal duct cysts are the most common congenital neck mass in children. Patients typically present with a midline mobile neck mass near the level of the hyoid bone. Thyroglossal duct cyst develop when there is failure of the thyroid descent tract or a thyroglossal duct to involute during development. This tract runs from the foramen cecum of the tongue to the low neck, usually passing anterior and in close relation to the hyoid.

Some physical exam findings that may point you in the direction of a thyroglossal duct cyst is elevation with swallowing or tongue protrusion, although this is not always present. A preoperative ultrasound of the neck should be performed to confirm the presence of normal thyroid tissue in its usual location. Surgical treatment of thyroglossal duct cyst include a Sistrunk procedure in which the cyst and its associated tract are removed as well as a central portion of the hyoid bone and the foramen cecum. Recurrence can be as high as 5% even if a Sistrunk procedure is performed, and is even higher with just simple excision. Dr. Petersson, thank you so much for joining us. Is there anything else you would like to add?

Dr. Raj Petersson:

Yeah, thanks for having me. Just in terms of recurrence, what do we do in that situation, we can go back to surgery and there's some options and generally, you can either do a repeat Sistrunk or more of an unblock central neck dissection type of procedure where we just take more surrounding strap musculature and tissue or maybe a little more hyoid bone with it. Those things may still lead to a 20 to 30% recurrence rate after that because once it's recurred, it just tells us that there's tract in there and now there's scarring and it becomes harder. There's newer techniques that people developed that are in the works about how to reduce that recurrence. Luckily, they don't recur very often if we do the Sistrunk in the first place.

And then the other point I would make is just with the cancer diagnosis. In adults who have undergone Sistrunk procedures for thyroglossal duct cyst, the median age of those patients was about 40. If it is going to be a cancer in there, 90-some percent would be a papillary cancer and a few percent might be a squamous cell cancer. But a lot of times, these cancers were not suspected and they were not found until the specimen went to final pathology. If you're going to see this in adults, I would make sure that cancer is in your differential when you do the surgery.

Dr. Alyssa Smith:

Awesome. Again, thank you so much for joining.

Dr. Raj Petersson:

Thanks for having me. It's my pleasure.

Dr. Alyssa Smith:

I'll now move on to the question portion of this podcast. As a reminder, I will ask a question, pause for a few seconds and then give the answer. The first question is what should be on your differential diagnosis for a midline neck mass in a pediatric patient? For pediatric patients with a midline neck mass, the top three on your differential diagnosis should include a thyroglossal duct cyst, a lymph node and a dermoid cyst. You can also consider some vascular malformations, hemangiomas, branchial cleft cysts, lymphomas, metastatic lymph nodes and atypical mycobacteria, but remember, the three things for a midline neck mass that should be at the top are the thyroglossal duct cyst, a lymph node and a dermoid cyst.

The second question is, what is the pathogenesis of the development of a thyroglossal duct cyst? Thyroid development begins at the third week of gestation, and the thyroid forms at the foramen cecum of the tongue, which is the junction of the anterior two-thirds in posterior one-third of the tongue. During development, it then descends to the low neck, usually passing anterior and in close relation to the hyoid. It reaches its final position by the seventh week of gestation, and normal involution of this tract is usually accomplished by week 10 of gestation. If there is failure of this tract to involute during development, a thyroglossal duct cyst can form. It's important to note that the cyst can form anywhere along this route of migration.

The third question is, what are some complications associated with a Sistrunk procedure? One complication is damage to the hypoglossal nerves that are in close association with the hyoid bone. This would present with tongue weakness and deviation of the tongue towards the side of the lesion on protrusion. Patients can also present with recurrence, and this has been noted to be up to 5% even in patients who have undergone a complete Sistrunk procedure. Finally, as with any neck surgery, there's always a risk of postoperative hematoma and infection. Thanks for joining, and we'll see you next time.