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Skull Base Tumors

Guest Expert:

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Yale Cancer Center Answers

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Welcome to Yale Cancer Center Answers with doctors Francine Foss and Lynn Wilson. Dr. Foss is a Professor of Medical Oncology and Dermatology, specializing in the treatment of lymphomas. Dr. Wilson is a Professor of Therapeutic Radiology and an expert in the use of radiation to treat lung cancers and cutaneous lymphomas. If you would like to join the conversation, you can contact the doctors directly. The address is canceranswers@yale.edu and the phone number is 1-888-234-4YCC. This week, doctors Foss and Wilson welcome Dr. Elias Michaelides. Dr. Michaelides is Assistant Professor of Surgery and Pediatrics. He is also the Director of the Yale Hearing and Balance Program and Otolaryngology Program Director. Here is Francine Foss.

Foss Let us start off by having you tell us exactly what it is that you do, what is your position is, and what skull base tumors are?

Michaelides As you mentioned, I am an otolaryngologist, or an ear, nose, and throat surgeon, first. After my training in otolaryngology, I subspecialized in otology, neurotology, and skull base surgery, which is a subspecialty of ENT that focuses primarily on problems involving the ear and the skull base including tumors of the skull base. The skull base is a complicated region in the head and neck because it is solid bone. It houses the inner ear structures and also has major blood vessels that supply the brain and multiple nerves that supply the ear, the facial muscles, and other lower cranial nerves that help with swallowing and sensation in the head and neck. My specialty kind of evolved as an extension of ENT, focusing on the lateral skull base because of the expertise that we have as ear surgeons and removing bone around these structures. To access tumors in this area, you must remove bone while not injuring any of those vital structures. There are a variety of types of tumors that we find in the lateral skull base including acoustic neuromas, which are tumors of the nerves that supply the inner ear, and also other types of cancer that begin in the skin and extend into the ear structures in the lateral skull base.

Foss So, what you are doing specifically is one type of ENT cancer. There are other types as well?

Michaelides There are specialists in ENT that focus on head and neck cancers, and primarily neck cancers that involve the vocal cords and the tongue. I do not perform those types of surgeries, but I focus on tumors around the ear. Being in ear, nose, and throat, I do primarily ear things including all the tumors around that area.

Wilson How did you become interested in this field? Was it during medical school that you developed an interest in ENT, and then once you were in ENT, how did this specialization come about?

Michaelides After finding my interest in ear, nose, and throat, and all the variety of different problems in the head and neck region, I found myself being very interested in the intricate workings of the inner ear. There is a complicated system of small hearing bones that allow us to transform vibrations

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in the air into hearing and understanding, and also there are organs in the inner ear that allow us to pick up information about motion, which gives us our ability to balance. This is very transitional type of information, where we can get information about the world and transmit it into the brain to understand surroundings. I found it very interesting taking care of these types of problems, though we have to access them, and to access them we learn special techniques for safely removing bone of the skull to preserve those structures while allowing us to approach these diseases, and that is how I kind of got interested in taking care of these tumors of the lateral skull base.

- Wilson Can you tell our listeners a little bit about how that process actually works? Vibrations in the air and how our ear works in terms of the bones you mentioned and fluid, how do we get from there to our outer ear canal, where our parents told us not to stick a Q-tip in too far kind of thing? What happens from that place on inward?
- Michaelides The sound is really just vibrations in air. The vibrations in the air go through the ear canal and then strike the eardrum, which sets the eardrum vibrating. The vibrations in the eardrum then get transmitted to three small hearing bones called ossicles. These are the smallest bones in the body and they transmit the sound to the inner ear. The inner ear is the cochlea. The cochlea is that snail-shaped organ in the inner ear that takes the vibrations in the air and transforms them into nerve impulses. Those nerve impulses then pass through several nerves into the brain, where our brain then takes those nerve impulses and turns them into thoughts about the sounds we hear. So, if you hear a siren, you think of a fire truck, and if you hear different words, you turn that into thoughts. So, it is transformation from vibrations in the air into nerve impulses into thoughts.
- Wilson How does the fluid in the ear work? Why is that there?
- Michaelides Well, the fluid in the ear bathes these tiny little hair cells within the cochlea. The vibrations in the air get transferred to the fluid, sets the fluid vibrating, and those fluid vibrations move the tiny hairs cells back and forth. The mechanical energy of those vibrations then takes the cells and makes them fire, which starts the process of the nerves being fired that go to the brain.
- Foss Elias, you also mentioned that dizziness can happen from problems with the ear. Can you explain to the listeners how it is that the ear is responsible for dizziness?
- Michaelides The inner ear, in addition to providing our ability to hear, picks up the motion of your head. So, if you turn your head left and right or move it up and down, tiny little organs in the inner ear can pick up that motion and transmit signals to the brain telling you that you are moving. When you

7:13 into mp3 file [http://valecancercenter.org/podcasts/2011_0918_YCC_Answers -
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move, the brain knows that you are moving based on picking up this motion from those sensors. From that, we can make adjustments in the muscles in our back and our legs to keep ourselves upright. Any abnormalities in the way that the inner ear picks up motion or transmits those signals to the brain can lead to confusing signals, which then leads to dizziness.

Foss Can you tell us how these base-of-skull tumors represent?

Michaelides Typically, if any of these tumors involve any part of the inner ear that has to do with hearing or balance, it can make those functions abnormal. So, if there is a tumor pushing against the nerves of the inner ear, you might have hearing loss, you might get ringing in your ear, or you might experience vertigo, which is a sensation of motion, which most people might call dizziness.

Foss These are fairly common symptoms in our patients that happen for a number of reasons. How do you actually sort this out when a patient may have a base of skull tumor and when there is another cause for these symptoms?

Michaelides There are many causes of hearing loss, starting from noise exposure, age, and from these tumors. One of the first steps we do is a hearing test to determine the pattern and the nature of the hearing loss and how much volume is lost and what pitches are lost. Looking at the pattern of hearing loss can give us some clues as to what might be causing it. Usually, if we see just a mild high-pitch hearing loss, which is a very common sign of the aging process, but if we see problems throughout the hearing spectrum and difficulty with the clarity of sound, that might indicate a tumor pushing against the hearing nerves.

Wilson Can you tell us a little bit about these different kinds of tumors? Do they spread? Do they tend to just grow locally? What is a typical patient like? Does it happen in younger people, middle age, older, can you give our listeners a sense of those details?

Michaelides There are two general categories of tumors of the skull base. There are the benign tumors, which are not cancerous, and there are malignant tumors, which are cancerous. The cancers that occur can eat away at the bones and all the different structures, and if they are extensive enough can spread to other areas in the body. The benign tumors, which luckily are most of the tumors that we find in the skull base, are benign and these tumors usually affect the inner ear by compressing the nerves or the other structures in the inner ear leading to the dizziness and the hearing loss or the ringing in the ears.

Foss At what age do we see these tumors?

10:20 into mp3 file [http://yalecancercenter.org/podcasts/2011_0918_YCC_Answers -
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- Michaelides Primarily we see benign tumors at almost any age, whereas cancers are more typically found in more elderly patients. Sometimes, the tumors that start in the skin around the ear might be from skin cancers that could be caused from too much sun exposure, but sometimes they can happen spontaneously with no real underlying reason. Most of the benign tumors that we see, like acoustic neuromas, just happen. We do not know why they occur, and they can occur in anybody at any age.
- Wilson You mentioned a little bit about symptoms. Can patients get pain with these compounds? Do they notice a bump near their ear, assuming that their hearing is okay?
- Michaelides For benign tumors, there is rarely any pain involved. All of the symptoms are usually, like I mentioned, dealing with the inner ear. With the cancers, there can be pain and some of the other nerves may be affected. A lot of the nerves that supply the vocal cords, the tongue, and the shoulders go through the temporal bone. So, if a cancer spreads to these areas, there may be weakness of the vocal cords leading to hoarseness, difficulty with speech because the tongue may not be as nimble as it was, because of its weakness, or there might be shoulder weakness also.
- Wilson The kind of specialty care that you provide, I do not think there are a lot of physicians that do exactly the kind of detailed work that you do; obviously, there are otolaryngologists who are generalists, but your type of subspecialization is fairly unusual. Is that true?
- Michaelides Yes, it is, because these tumors are not so common, we do not need a whole lot of people taking care of them, and the general otolaryngologists can care for the most common types of problems in the inner ear. Because of the complexity of the anatomy in this area, when there is a deeper tumor to get to, we do need that specialized care. Also, I do not work alone in most situations. When these tumors are deep within the inner ear, they often spread to the areas next to the brain. So, for many of the surgical procedures, I work together with a neurosurgeon. I might do the approach to the tumor, where I remove bone and other structures to get access, and a neurosurgeon might be involved to help remove the tumor from near the brain stem.
- Foss Elias, that brings up a good question, which is how you actually diagnose these tumors. Since many of them are deep, you do not just get a biopsy like we will get a biopsy of a lung lesion with a needle, for instance. How do you actually make the diagnosis?
- Michaelides The first step is doing the basic diagnostic tests, where we look at the hearing, the balance and so forth, and then if we suspect a tumor causing the symptoms, we will often turn to our radiology

13:30 into mp3 file [http://valecancercenter.org/podcasts/2011_0918_YCC_Answers -
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colleagues and do either an MRI scan or a CAT scan to help us look at those deeper areas.

Wilson What are the differences between those two scans, CT versus an MRI scan?

Michaelides The CT scan is very good at looking at the bony structures of the skull and the ear. On the other hand, the MRI scan is not very good at looking at bone but is very good at looking at muscles, brain, and other soft tissues around the area. So, we use both of these in conjunction often to get information about both of those structures.

Wilson: We are going to take a short break for a medical minute. Please stay tuned to learn more information about surgery for skull base tumors with Dr. Elias Michaelides.

*Medical
Minute*

It is estimated that nearly 200,000 men in the US will be diagnosed with prostate cancer this year, and over 2,000 new cases will be diagnosed in Connecticut alone. One in six American men will develop prostate cancer in the course of his lifetime. Major advances in the detection and treatment of prostate cancer have dramatically decreased the number of men who die from this disease. Screening for prostate cancer can be performed quickly and easily in a physician's office using two simple tests, a physical exam and a blood test. Clinical trials are currently underway at federally designated comprehensive cancer centers like the one at Yale to test innovative new treatments for prostate cancer. The da Vinci Robotic Surgical System is an option available for patients at Yale that uses three-dimensional imaging to enable the surgeon to perform a prostatectomy without the need for a large incision. This has been a medical minute and more information is available at yalecancercenter.org. You are listening to the WNPR Health Forum on the Connecticut Public Broadcasting Network.

Wilson Welcome back to Yale Cancer Center Answers. This is Dr. Lynn Wilson and I am joined by my co-host, Dr. Francine Foss. Our guest today is Dr. Elias Michaelides, and we are discussing surgery for skull base tumors. Elias, can you give our listeners some more details about acoustic neuromas. I know you have talked about these a little bit already, but can you give us more detail about what they are like for patients and some of the procedures that are involved in solving these problems?

Michaelides First and foremost, acoustic neuromas are very uncommon tumors, and they are benign tumors, which means their growth does not spread and they are not cancerous. The most important part is to reassure patients when they are diagnosed with acoustic neuromas that they are not overly dangerous. They grow very slowly and rarely cause life-threatening problems. They typically

16:04 into mp3 file [http://yalecancercenter.org/podcasts/2011_0918_YCC_Answers -
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cause hearing loss because they are based on the nerves that supply the inner ear, either from the balance nerve or from the hearing nerve. As a tumor grows, they compress the nerve and cause the symptoms that we talked about, which are the hearing loss, dizziness, and tinnitus. In general, these tumors are based on the nerves and to take care of these problems we must either remove them surgically, or stop them from growing, and to slow them from growing we can use very focused radiation treatments or we can remove them completely with surgery. The way that we make the decision on the best way to treat these, depends on the size of the tumor, the extent of the changes and the hearing and the balance function, and the medical condition of the patient.

Foss Can you talk about these specialized surgical procedures? I imagine that they are very intricate, getting into this tough area.

Michaelides The tumors are based on the nerves of the hearing and balance, and they are situated within the solid bone of the skull, deep into the skull, in fact, around the inner ear structures. So, to get surgical access to these areas, we must remove some of this bone. Again, there are multiple large blood vessels and other nerves, and the cochlea and the balance organs to contend with. Our goal is to avoid injury to any of these structures that we can to provide appropriate surgical access to preserve as many structures as possible. There are several different surgical approaches depending on how much hearing is left. Unfortunately, for some patients, the hearing is damaged so much that we are not able to save it, and then we use a direct route where we go through the inner area. In other circumstances with tumors very small, we are able to remove the tumor while saving the hearing nerves, so that we can preserve hearing. With these approaches we go around the ear by either coming above it or below it, and going next to the brain, carefully moving the brain out of the way to access the areas to take the tumor out.

Foss So, if you remove bone say from the skull to do this kind of procedure, does that bone get put back in?

Michaelides Depending on the approach, we can make a window into the skull and replace the bone. In other circumstances, when we go directly through the ear, there is enough surrounding bone that we do not need to replace it and we are able to just close over the incision and it heals up nicely.

Foss How often do patients have complications from this procedure?

Michaelides It is a difficult approach, and it is surrounded by very important structures; the brain, blood vessels, and hearing organs. So, there is the risk of complications in any type of surgery around

19:12 into mp3 file [http://yalecancercenter.org/podcasts/2011_0918_YCC_Answers -
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this area. Sometimes, the facial nerve, which supplies the muscles of the face, runs right next to these tumors and can be compressed also. There can be some facial weakness, which can be temporary or long term in removing these tumors. Other risks are the same risks that go along with any type of neurosurgical procedure, which can include spinal fluid leaks or even bleeding around the brain, but these are very rare complications.

- Wilson As far as treatment options, you mentioned radiotherapy, have the options changed over the years or are these two primary choices something that have both been available for a long, long time?
- Michaelides The field of skull base surgery has evolved over the past 50 years, only. Before being able to do complicated bone removal, most of these tumors were not treated very well and often led to more complications. Now, with the fine techniques and high-powered microscopes and very delicate instruments, we are able to take these tumors out, and there is continual improvement in the surgical techniques. Over the past 25 years or so, we have added the ability to do very focused radiation treatments using a device called a stereotactic radiotherapy or a Gamma Knife, which is a type of radiation that focuses its treatment only on the tumor and not the surrounding tissues and can stop the growth of the tumor. This is often a very good option if someone is quite ill from other medical problems or is elderly and might not be able to withstand a major surgical procedure. Interestingly, for very small tumors in elderly patients, we often do not do anything. Since these tumors are so slow-growing, with a small tumor it might take 15 or 20 years to grow to a size that could cause real problems, we often will just do an MRI scan once a year to follow the tumor. If it does not grow, we do not need to do anything.
- Wilson How long is a patient typically in the hospital after the surgery?
- Michaelides After a typical surgery to remove an acoustic neuroma, a patient usually stays in the hospital for 4 or 5 days. Obviously, right after a major surgery, they will require intensive monitoring, but sometimes the patient just needs to stay a few extra days in the hospital because they might be dizzy after the surgery as we are taking the tumor off of the balance nerve, and that can lead to a temporary condition of vertigo.
- Foss Elias, how often do these tumors recur after you excise them?
- Michaelides Since they are benign tumors, if you remove the entire tumor, they do not come back. However, sometimes, we will leave a tiny bit of tumor on the facial nerve to avoid injury to the nerve. Usually, when you take most of the tumor, you take away its blood supply, so the tumor will no longer grow. It is very uncommon to see these tumors recur if the majority of them are removed.

22:28 into mp3 file [http://valecancercenter.org/podcasts/2011_0918_YCC_Answers -
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Wilson This might be a difficult question to answer, but if someone say, for example, has paralysis of their facial nerve and they are having difficulty using the muscles of their face, how do you generally make the determination about when to speak to a patient that you might be able to reverse this, versus a situation where you see certain factors that you are fairly convinced you are not going to be able to reverse that process? What are some of the clues?

Michaelides Luckily, it is very unusual to see facial weakness from an acoustic neuroma. Only when the tumor becomes very large and untreated for many years will the compression of the tumor cause problems of the facial nerve. If there is partial weakness of the facial nerve, there is a very good chance that removal of the tumor or treating with radiation will halt the progress of the damage to the nerve, and sometimes we actually may see some improvement. If there is no function of that nerve left, it also means that there is very deep involvement of the nerve and it may not recover with removal alone. However, we are able to nerve-graft it, so, take a nerve that is not needed from someplace else in the body and replace the damaged part of the nerve so that over time the nerve fibers can grow back and reanimate the face so that it can move more normally.

Wilson Do these tumors ever occur on both sides in a particular patient or is it usually just one side?

Michaelides Very rarely there is a rare genetic condition that can cause these tumors to occur in both sides. We call that neurofibromatosis, but it is a very uncommon thing to see it on both sides. In almost all circumstances, we see it just on one side and it does not have a genetic cause, it is a spontaneous tumor.

Wilson Meaning that you do not really see it in family members or anything like that?

Michaelides Correct.

Foss Elias, how often do you see other kinds of tumors in this area? We talked about acoustic neuroma, but how often do you see say metastatic disease in this area and how often do you see just benign meningiomas in this area?

Michaelides The area where acoustic neuromas form, within the nerve canal of the ear, I'd say about 95% of the tumors in this area are acoustic neuromas. We can see meningiomas, which behave exactly the same as acoustic neuromas and we treat them the same. There are other very rare types of tumors in this area including small fatty tumors and there are several instances where

25:14 into mp3 file [http://valecancercenter.org/podcasts/2011_0918_YCC_Answers -
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there can be metastatic disease from cancers elsewhere in the body including breast cancer that can spread to this area. Typically though, we do see different patterns of growth that we can pick up on MRI scan and CAT scan that help us tell the difference between these types of tumors.

- Wilson You mentioned working sometimes with a neurosurgeon, for example. Tell us a little bit about the first visit for a patient, how does it work at Yale, and here at Smilow Cancer Hospital, do you have a multidisciplinary team? Have you assembled a team to approach these problems? Give our listeners some of those details.
- Michaelides I typically am one of the first physicians to see a patient with hearing complaints. So, when a patient is diagnosed with one of these types of tumors, I typically begin the workup. If it appears that it is a tumor that might need treatment, I involve an entire skull base team. We have assembled group physicians, all with interest in taking care of tumors in this region including neurosurgeons, radiation oncologists, neuroradiologists that specialize in diagnosing tumors with CAT scan and MRI scans in this area, and we also will involve medical oncologists for the rare circumstances where we might need chemotherapy also.
- Foss Are there any research areas of interest in this particular area of the body now in terms of looking at skull base tumors? Are there ongoing studies? Are there research questions that are being explored?
- Michaelides We are always interested in finding out what the best treatments are for these tumors. Many factors go into deciding what is the best approach, be it just watching the tumor over long periods of time, treating with Gamma Knife therapy, radiation, or one of the surgical approaches. We still have some controversy in our field about which one is the best one, and I think it really takes a multidisciplinary team to look at these tumors to come to a consensus on what the most appropriate treatment is, and we continue to look at this to learn from our experiences and decide what the right treatment is for every patient.
- Wilson Is there any role for robotic surgery in the kind of work that you do?
- Michaelides I certainly would like to see the development of robotic surgery to help us in these delicate areas, but right now the technology I think is not quite good enough to allow us to use robots in these areas. Eventually, in the future, I think we can see that it would be helpful to us to use such devices to more accurately remove the bone around the inner ear but some work still needs to be done.

28:09 into mp3 file [http://yalecancercenter.org/podcasts/2011_0918_YCC_Answers -
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Wilson I figured since you are working in a pretty small place, so even with the robots, it is hard to get instruments in place.

Michaelides Hopefully we will see some advances in technology, anything that can help us in this delicate area would be very helpful.

Dr. Elias Michaelides is Assistant Professor of Surgery and Pediatrics, Director of the Yale Hearing and Balance Program and Otolaryngology Program Director at the Yale School of Medicine. If you have questions or would like to add your comments, visit yalecancercenter.org, where you can also get the podcast and find written transcripts of past programs. You are listening to the WNPR health forum on the Connecticut Public Broadcasting Network.