YALE CANCER ANSWERS

Skull Based Tumors: Diagnosis and Treatment

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Welcome to Yale Cancer Answers with doctors Anees Chagpar and Steven Gore. I am Bruce Barber. Yale Cancer Answers features the latest information on cancer care by welcoming oncologists and specialists who are on the forefront of the battle to fight cancer. This week, it is a conversation about the diagnosis and treatment of skull-base tumors with Dr. Bulent Omay. Dr. Omay is an Assistant Professor of Neurosurgery at Yale School of Medicine and Dr. Gore is a Professor of Internal Medicine in Hematology at Yale and Director of Hematologic Malignancies at Smilow Cancer Hospital.

Gore I did not even know until about an hour ago that there was such an area as skull-base tumors and I bet our listening audience does not have a clue, are you a surgeon?

Omay Yes. I am a neurosurgeon. Neurosurgery is a specialty that deals with diseases of the nervous system and the surgical treatment of them.

Gore And they are also the brunt of jokes, this isn't neuroscience right, this isn't neurosurgery?

Omay That's true. And a skull base surgeon is actually a subspecialty of neurosurgery and it defines the neurosurgeon who deals with pathologies and this is most of the time tumors that arise at the base of the skull, and the reason it became a subspecialty is because it is a very complex area of the body where the brain interacts with the rest of the body and the skull base actually holds the brain and it has a lot of foramina that cranial nerves, major vessels pass through and it creates this very complex anatomy that requires another subspecialty to get trained and to have experience in it.

Gore I can imagine because I know that I had been to a medical school where I had to actually take a test on those various holes in the base of the skull. For sure, I would not be where I am today. Fortunately, I was a Yale Medical Student where there was an open book test. But I remember how complicated that was. Are these tumors arising out of the bone or are they arising out of the brain tissue or the lining outside the brain or all of the above?

Omay I guess the answer would be all of the above. The skull base is a complex area filled with different types of tissues, and that creates different types of cancers, essentially. Tumors can arise from the brain itself, from the covering of the brain, the dura, they can be outside the brain or dura and can arise from the bone, cartilage at the skull base

level, it can arise from the cranial nerves that have just exited the brain and sometimes there can be cysts associated in this area and sometimes there are some embryologic remnants that grow over time and create problems and act like tumors.

Gore You mean like leftovers of tissue that should have developed when the person was in embryo or fetus and somehow that primitive tissue remained?

Omay Correct. Skull base and especially midline skull base is a common area that these kinds of tissues can grow in and create problems.

Gore Now I am kind of nervous. If I had a problem at the base of my skull, would I know it?

Omay Yes and no.

Gore I was thinking it was an easy question.

Omay The way I end up seeing patients in my clinic is there are 2 different groups; one of them is the incidental diagnosis, where a patient ends up having a brain scan either a CT or an MRI of the brain for a minor trauma, gets involved in an accident, they scan the brain and find a lesion.

Gore Or have a bad headache or something like that?

Omay Yes. Sometimes they have symptoms and that kind of prompts the imaging and a lesion is found that way. And that can be as you said headaches or again this area, because is kind of a tightly spaced area with different tissues and critical structures, nerves that carry vision – optic nerves or like nerves that involve movements of the face, facial muscles or the sensation to the face, all these nerves are like closely packed in the skull base area, and any lesion, even small lesions that start growing in that area can interact with these structures and create symptoms like headaches, facial pain, vision loss and sometimes tumors that involve the frontal sinuses or again the sinuses in the face, sphenoid sinus, can create problems with functions of the sinuses and the nerves like a blockade of the nerves. Actually, there are a variety of ways that these tumors can present themselves.

Gore In general, patients would be referred to you because somebody else has done a scan that showed a problem?

Omay Yeah, most of the time when I end up seeing a patient, they already had a scan done and have a preliminary diagnosis of a skull base tumor.

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Gore But we do not know at that point whether it is a malignant tumor or potentially a benign tumor?

Omay We do not. At least we do not know with accuracy. Usually, it is possible to make an educated guess about what a tumor would be, but without pathological diagnosis, it is impossible to tell for sure whether the tumor is malignant or benign.

Gore Sounds like that must be a very terrifying diagnosis for a patient to find out you have got something bad at the base of your skull, go see this guy at Yale, he is the fix-the-skull surgeon. Seems like that would like really terrifying.

Omay When I interact with patients, because having a tumor diagnosis is bad on its own but also having a tumor in this area, which is very hard to reach adds another level of complexity and anxiety on the patient level. And I do my best to be clear to the patients that this is going to be a journey and they need help from their family and also they need to understand what we are dealing with and how we are going to deal with that in detail. Because I think that is key to dealing with anxiety of the situation, just understanding what is going on.

Gore I imagine the first step must be to get a biopsy and find out what you are dealing with or is that not the case?

Omay Each case is evaluated individually and somewhat tailored to the patients. Some of them may require a biopsy, but sometimes we have enough evidence that we pretty much can guess what the tumor would be. In that case, we may proceed with a goal of resecting the tumor, not doing the biopsy first.

Gore Taking it out completely?

Omay Correct, taking it out completely and then have a pathologic evaluation afterwards and we will know what it is. But regardless, when we start the operation and first encounter the tumor, we send samples to the pathologist even before proceeding with removal of the tumor.

Gore So, while you are actually there and the patient's head is open?

Omay Yes. The pathologist quickly reviews the slides and gives us a mostly reliable answer about what he or she thinks this tumor is. And that gives another level of confidence to moving forward with the full removal of the tumor.

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Gore Is there any circumstance under which you would not take the tumor out?

Omay Yes. There are some scenarios where we may decide to stop the operation at that point because the tumor that we think is diagnosed at this point may be treatable with medications. Ini that case, then the patient will not be subjected to the potential complications of a resection, removal of the tumor, and we might just stop at that point and start medical therapy.

Gore So, that might be if it is something like a lymphoma or something that is very responsive to treatment.

Omay Correct. Lymphoma is a typical scenario that happens. This is not a common situation, it is very rare that it happens because there are ways that we can guess that a lesion may be lymphoma before we plan this and we might just do a biopsy at that point.

Gore I see. So, I am still trying to get my head so to speak around this complicated area, and I just cannot imagine, how do you go in there? How do you do that safely because it is connected to the brainstem which is connected to the spinal cord and all those nerves that feed, swallowing, tongue, taste, hearing, and all that stuff – that is like a such a poorly designed area evolutionarily in terms of if anything is going to bad, everything goes bad right there, right?

Omay That is correct.

Gore I mean it is a crazy place.

Omay That is totally true. Unfortunately, the anatomy is very complex and unpredictable in a way. So, each patient is different. First we start with evaluating the patient's images – the MRI scans and have an idea about where the tumor is, what are the neighboring structures, what is it interacting with and why are the symptoms occurring the way they occur and how can we approach this? In skull base surgery, the approach is kind of a very common theme like how are we going to approach this tumor. Because that might be very critical structures between the surgeon and the tumor as we are approaching the tumor and we have to approach it.

Gore You cannot get there from the top of the head right?

Omay No. We do not want to go through the brain for sure.

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Gore That sounds terrible right?

Omay Over the years, there have been very innovative ways developed to approach these lesions and these are described as skull base approaches. There are the conventional skull base approaches, which require big incisions over the scalp or the face sometime and big bony openings that can give access to the surgeon by navigating around these critical neural and vascular structures and the brain without manipulating them too much.

Gore It would make a big window in the bone?

Omay Correct. So, that is the conventional way of skull base approaches. But especially in the last 10 years, another method has arrived. So, this is the endoscopic method to skull base approaches. So, the idea or the strategy of the endoscopic method is to use natural corridors that already exist in the body in the face and the cranium and bring the actual light source and the lens that is required to do these surgeries close to the pathology where the tumor is using these corridors and make smaller bony openings and use the endoscope, which is a very innovative way of approaching and visualizing these lesions and do the resections that way.

Gore You are talking about going through some of those holes you talked about in the base of the skull or through the sinuses or what are you doing?

Omay Through the sinuses. So, the typical approach is the endoscopic endonasal approach, which means that the endoscope goes through the nose and goes through the sphenoid sinus most commonly.

Gore That is a sinus that is behind the nose, kind of?

Omay Correct. Sphenoid sinus is in the center of the head, under the brain and behind the nerves. So, it is almost like a natural opening to get access to skull base without going through the brain, without the need to retract brain. So, this is done jointly with an ENT surgeon.

Gore That would be a head and neck surgeon?

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Omay Correct. The head and neck surgeon actually does the nose part of the operation and brings the surgical team like intrasphenoid sinus where the skull base is visible. So, the top of the sphenoid sinus is essentially the skull base.

Gore This is very exciting because not only do you get your tumor resected but it sounds like you might get a nose job, but right now we need to take a short break for a medical minute. Please stay tuned to learn more about skull base tumors with Dr. Omay.

Medical Minute

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This is a medical minute about genetic testing, which can be useful for people with certain types of cancer that seem to run in their families. Patients that are considered at risk receive genetic counseling and testing, so informed medical decisions can be based on their own personal risk assessment. Resources for genetic counseling and testing are available at federally designated comprehensive cancer center. Interdisciplinary teams include geneticists, genetic counselors, physicians and nurses who work together to provide risk assessment and steps to prevent the development of cancer. More information is available at YaleCancerCenter.org. You are listening to Connecticut Public Radio.

Gore Welcome back to Yale Cancer Answers. This is Dr. Steven Gore. I am joined tonight by my guest, Dr. Bulent Omay and we are discussing the diagnosis and treatment of skull base tumors. Bulent, just before the break I made this unfortunate joke about the head and neck surgeon, I guess it is really a plastic surgeon that would do the nose job classically, so it is not really funny to make light of what is a really serious problem, so I apologize if I offended anybody as I was trying to make light, but so you are giving us this fascinating story about how the head and neck surgeon puts the endoscope through the nose into this big sinus and they are called the sphenoid sinus and what you said right before the break was that the top of the sphenoid sinus is the skull. Is that right?

Omay That is true.

Gore Scary. Do you take the roof of the sphenoid sinus off?

Omay Yes, at that point, once we bring our endoscope into the sphenoid sinus which is this air-filled cavity which already exists in every patient.

Gore I mean, are we talking about something that is an inch or half an inch or 3 inches, or how big is this sinus?

Omay Probably an inch.

Gore That is tiny, cannot even imagine.

Omay It is actually very variable, like from patient to patient, but it can be small, it can be large, but it is large enough so that we can bring our endoscope in and get our instruments in there so that we can access the skull base.

Gore Are the instruments part of the endoscope or they are separate?

Omay They are separate.

Gore So, they are a flexible tuby thing?

Omay They are actually straight, they are not flexible, but they are elegant, long structures designed instruments that help us do these surgeries.

Gore They must be tiny right?

Omay They are..

Gore Millimeters or?

Omay For example, the scissors that we use are probably a couple of millimeters in length.

Gore Wow and how wide is the clip part of the scissors?

Omay So, they are attached to this like long base that comes out of the nose and the surgeon is actually operating out of the nose.

Gore I imagine with a microscope or something?

Omay The endoscope, and the endoscope is actually in the sphenoid sinus and it brings the light source and the lens very close to the pathology.

Gore You got some goggles or some optical ways to look in right?

Omay It creates a high-definition image and it is projected into our monitor.

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Gore So, you are doing TV surgery?

Omay Correct.

Gore Did you hear folks, your base of the skull is on TV. Wow! I guess you can really blow it up big right? High- def TV no doubt.

Omay Correct. I mean it creates very impressive images. And so once we are there, we also use what we call a navigation system which is used in very different types of brain surgery, but it is very important in skull base surgery as well. This is almost like a typical navigation device.

Gore Like a GPS kind of thing?

Omay Exactly. That tells us where we are in the brain, in the skull and we can check on another monitor to make sure that we are actually in the place that we want to be on an MRI. So, we kind of correlate what we are in actual reality to where we are, where we would be on an MRI scan.

Gore But there is not an MRI machine in the room.

Omay No there is not at that point. So, after all these checks are done, we use a drill, almost like a dentist drill that is like very small and elegant instrument that can create a hole in the skull base. So, once we create that hole, now we have access to the brain essentially and what is like under the brain, between the skull and the brain. And then we proceed with the operation, and once we get access, we remove the pathology and at that point we deal with the problem of closing the defect. So, in skull base surgery, there is always this strategic problem of closing a skull base defect.

Gore You got a hole in there.

Omay Correct, and that hole is communicating with the nose. So, that needs to be closed and we usually use a vascularized tissue that is derived from the nose and almost like a plastic flap that is created within the nose.

Gore Sort of like a graft, like almost like a skin graft but it is using nose tissue?

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Omay Correct, exactly. And then, we use that graft to close this defect in addition to other methods, but the graft is kind of key in these kinds of closures. So, brain ends up being separated from the nose.

Gore And those tissues like those membranes that surround the brain, do they grow back or you do not need them?

Omay They eventually heal, and especially this vascularized flap is the key to that healing process.

Gore Wow. So, I know that in a lot of or at least in some brain surgeries from what I understand, they keep the patient partially awake so that they can make sure they stimulate a part of the brain and stuff, are these patients awake with this base of skull surgery?

Omay No. That awake surgery is commonly done with primary brain tumors, tumors that arise from the brain tissue itself that are close to critical areas of function like speech or movement. But in the skull base, this is never required and we want the patient to be stable, not moving at all because we are dealing with very, very small areas, so actually it is not wanted that the patient would be awake and have a potential of moving.

Gore How will you know if you by accident nick one of these nerves or other structures that you were telling us about - the optic nerves for vision or these nerves that control the tongue or anything like that? I mean can you see them very clearly?

Omay So, we are using various methods and technologies to kind of navigate through the skull base; one of them is the GPS system that I mentioned. Direct visualization is ideal of course, but it cannot always happen because we are dealing with very small openings.

Gore There is a tumor there in the way right?

Omay Yes and there is a tumor there. So, we are using the navigation system that bases itself on the MRI, that is one thing. We use small Dopplers that kind of tell us where the main vessels will be in real time.

Gore That is like an ultrasound machine or something?

Omay That is like an ultrasound machine that can track movement of blood.

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Gore I see. So you are looking for the blood vessels that way.

Omay Correct, so we stay clear from them. That is one method. Knowledge of basic anatomy about how things look at the skull base is also extremely valuable to the skull base surgeon because for example, the optic chiasm has an impression on the skull base.

Gore You mean where the optic nerve separates?

Omay Yes, and how the optic nerves move in the skull base, they leave an impression and the surgeon can see and track that impression and know that the nerves would be there if the surgeon went deeper in that area.

Gore Well, this seems to be like pick the hardest job you can pick as a neurosurgeon right? I mean, what was it that made you say, I want to do taht, I want to be there with a microscope and the endoscope and these teeny weenie scissors, I mean how did you decide to do this?

Omay It was kind of one thing after the other that brought me into being a skull base surgeon, once I started my neurosurgical training, I always was interested in brain tumors and as I progressed through my training.

Gore Which is long training right?

Omay Yes, which is long, 7 years of neurosurgical training. Then, I did 2 years of brain tumor fellowship and I did another endoscopic skull base fellowship after that.

Gore My gosh that is a lot of years.

Omay Yeah, it was. But it is worth it and that is the reason people end up doing it, although it is a very challenging field in surgery and it is very challenging for the patients, the rewards are also very high. If one can help a patient from getting rid of a tumor in the skull base, it is a very rewarding feeling.

Gore Oh! I can only imagine. And can I ask how many surgeons or surgical assistants are involved in one of these. Is it just you or it sounds like you have got your endoscope, you got your other thing, you got your ultrasound, you do it all yourself or do you have an assistant?

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Omay So, I am working at an academic center and we have neurosurgical residents that help us do these operations and as I mentioned, there is a surgical team, which is made up of ENT surgeon and neurosurgeon and we have our own residents, so there are 4 people who make the surgical team. Then, there is the anesthesiology part of it. That is another team in the OR.

Gore And they are right nearby right, they are all in the face. Cozy?

Omay Yes. Actually, we turn the patients, so they are actually at the feet of the table with their monitors. So, that is kind of how we work it out and of course, there are the surgical techs, the nurses and all the other supporting staff in the OR which are very critical. And we have the pathologist, who is there looking at the slides.

Gore Right there?

Omay In the same area.

Dr. Bulent Omay is an Assistant Professor of Neurosurgery at Yale School of Medicine. If you have questions, the address is canceranswers@yale.edu and past editions of the program are available in audio and written form at YaleCancerCenter.org. I am Bruce Barber reminding you to tune in each week to learn more about the fight against cancer here on Connecticut Public Radio.