



AHNS Skull Base Surgery Section Sinusal Patient Information

AHNS Sinonasal Patient Information

INTRODUCTION

Cancers of the nose and sinuses are a rare group of diseases which make up 3% of head and neck cancers and affect about 1-3 per 100,000 people per year. There are many different types of cancers that occur in this location, and they usually arise from the lining of the sinuses or nasal cavity. If tumors become large, they can extend to structures including the brain, cranial nerves, large blood vessels, and the eyes. Given the complex anatomy in this region, care for these cancers is best done at a specialized center with an experienced team.

SYMPTOMS

The most common symptoms of cancers originating within the nose and sinuses are nose bleeding and difficulty breathing through the nose. Additional symptoms can mimic sinusitis and include facial pain, nasal drainage and changes in sense of smell. Sudden symptoms on just one side of the nose or face raises the suspicion of a tumor. Because these tumors can grow into structures near the sinuses, additional symptoms include numbness of the face, vision changes, bulging of an eye, difficulty or pain with opening the mouth, change in how the teeth meet, facial droop, neck masses, and hearing loss.

EVALUATION

A specialist should perform a complete head and neck examination, including evaluation of ears, eyes, nose, mouth, throat, neck and cranial nerves. Evaluation with an endoscope (a thin camera that fits into the nose and sinuses) is strongly recommended. Evaluation by an ophthalmologist (eye specialist) may also be needed to test vision.

If a tumor is suspected, imaging studies are required to fully evaluate the size and location. A CT scan is most commonly the first step. CT scans are particularly helpful at evaluating the sinuses and bones of the skull. An MRI may also be useful to evaluate nerves, brain, blood vessels and soft tissue in the area. It is common for patients to require both an MRI and CT scan to get a full picture of the tumor.

A biopsy (removal of a small piece for diagnosis) of the tumor is required to determine what type of tumor is present. There are many different types of tumors that can be present within the nose and sinuses, and the treatment plan will vary depending on what the type of tumor is present. There are many ways to obtain a biopsy, and the best method will be determined by the tumor size, location and other characteristics.

If a cancer is confirmed, staging the tumor is a critical step. Staging includes determining the size and location of the sinonasal tumor and if the tumor has spread anywhere else in the body. The two most common ways to evaluate if the cancer has spread is with a CT scan of the neck and chest or a positron emission tomography-computed tomography (PET-CT). Cancerous tumors are staged by the TNM staging (T = tumor size, N = lymph nodes with cancer, M =



AHNS Skull Base Surgery Section Sinus Patient Information

metastasis to other sites in the body) and converted to an overall stage 1-4, which will guide treatment.

Once the tumor has been biopsied and all the imaging is complete, your team will determine the best treatment plan. Your treatment team will review medical records from your other/previous doctors. You should bring any scans (on CD or have sent beforehand) and test results (including biopsy reports) to your appointments. Discussion is commonly held at a multidisciplinary “tumor board” attended by the head & neck surgeons, radiation oncologists and medical oncologists. The plan may involve a combination of surgery, radiation therapy, chemotherapy or immunotherapy. Consultations with a neurosurgeon, reconstructive surgeon, eye surgeon will be ordered if needed. Clinical trials may also be appropriate.

COMMON TYPES OF SINUS CANCER

Squamous Cell Carcinoma. Squamous cell carcinoma (SCC) is the most common cancer in the sinuses. Men are affected twice as often as women. Risk factors for SCC include tobacco use, as well as nickel, asbestos, formaldehyde and textile industry exposures. The role of longstanding sinusitis is not clear. SCCs can arise from previously noncancerous tumors called inverted papillomas. Treatment for sinus SCC is often surgical, followed by radiation treatment for more advanced disease. For tumors that cannot be surgically removed, radiation or chemotherapy with radiation is standard (depending on stage). Lymph nodes in the neck may need to be treated with radiation or surgical removal (a neck dissection).

Adenoid Cystic Carcinoma. Adenoid cystic carcinoma is the second most common sinus cancer. It usually presents as a slow growing mass with nerve symptoms (numbness or weakness of the face, for example). Tumor grade (how aggressive it looks under the microscope) impacts how we think about likelihood of spread and aggressiveness of treatment. Nerve involvement is common for these types of tumors, and there is a high rate of recurrence. Complete surgical removal may be challenging because of a tendency to spread along nerves towards the brain. Adenoid cystic carcinoma is generally slow growing but has a tendency to spread to other parts of the body (most commonly the lungs) even years after diagnosis and treatment of the sinus tumor. Treatment is often with surgery plus radiation. Proton beam radiation has been used in some cases. Chemotherapy is generally not recommended though there is ongoing research on chemotherapy and immunotherapy options.

Adenocarcinoma. Adenocarcinoma accounts for a third of sinus cancers. Adenocarcinoma arises from the mucous glands in the nose and sinuses. Occupational exposure in woodworkers to both wood dust and chemicals is a well-documented risk factor for sinonasal adenocarcinoma. Adenocarcinomas commonly start in the upper nasal cavity and ethmoid sinuses. Tumor grade (how it looks under the microscope) impacts the likelihood of spread and aggressiveness of treatment. Treatment is often surgical removal with postoperative radiation, though treatment is grade- and stage-dependent.

Lymphoma. Lymphoma occurring in the sinuses is typically non-Hodgkin lymphoma. Lymphomas of B-cell, T-cell, T/Natural Killer (NK)-cell types may occur and have distinct



AHNS Skull Base Surgery Section Sinusal Patient Information

behavior. Surgical intervention is typically limited to biopsy. Treatment varies by the type of lymphoma but commonly involves local radiation therapy with possible chemotherapy depending. Care is most often directed by a medical oncologist specializing in lymphomas.

Esthesioneuroblastoma. Esthesioneuroblastoma, or olfactory neuroblastoma, is rare (about 1 in a half million) and there are no known risk factors. It often comes from the small nerves that allow for sense of smell just under the brain or at the top of the nasal cavity and sinuses. Olfactory neuroblastomas often grow slowly. These cancers may extend into the eye socket causing a bulging appearance or vision changes. They also spread to lymph nodes in the neck about 20% of the time. Treatment is often accomplished with minimally invasive surgery upfront. Radiation is often recommended after the surgical removal to both the sinus area and the neck. For more extensive disease, an “open” operation that involves a larger external craniotomy may be required, along with neck dissection (removal of the lymph nodes of the neck). Local recurrences are common, and long-term follow-up is necessary. Olfactory neuroblastoma has the best prognosis of all sinus cancers, but again, recurrence is relatively common, even years down the road.

Sinonasal Undifferentiated Carcinoma. Sinonasal undifferentiated carcinoma (SNUC) is an uncommon, very aggressive cancer. It often shows up as a rapidly enlarging mass with nosebleeds and facial pain. It often has extension to or into the eye socket and brain. Treatment often involves chemotherapy, radiation therapy and surgical removal. Prompt treatment at an experienced center is crucial.

SURGICAL MANAGEMENT

Surgery for cancers of the nose and sinuses requires careful planning and understanding of the complex anatomy. Surgery will involve removal of the tumor plus reconstruction. Depending on the specific tumor, removal of tissue near the brain, nerves, large blood vessels and eyes may be necessary. When possible, minimally invasive techniques can be used to remove the tumor typically through the nose using scopes, often an otolaryngologist (ENT) and a neurosurgeon working together through both nostrils. However, there are situations in which surgery with scopes is not the best approach. Larger tumors that involve the brain may require a scalp incision and temporary removal of a skull bone by a neurosurgeon. Important factors to consider are the specific type of tumor, the size, location and stage of the tumor plus the reconstruction required after the tumor is removed. The ultimate decision about surgical approach and reconstruction should be made by an experienced team who are comfortable with all the available techniques. For further information, there is a patient education document about skull base surgery on the website <https://www.ahns.info/sections/skull-base/for-patients-skull-base-surgery/>.

RADIATION THERAPY

Radiation therapy (RT) is a commonly used in the treatment pathway for sinonasal cancers. Radiation treatments are often given by radiation oncologists after surgery (adjuvant therapy),



AHNS Skull Base Surgery Section Sinusal Patient Information

sometimes in combination with chemotherapy (chemoradiation). Tumors that are not amenable to surgery may be treated with chemoradiation as a first line of treatment. Despite improvements in disease control, RT can have significant complications due to injury to the brain, visual pathway, inner ear, and remaining local tissue. Modern advances in RT technology allow improvements in radiation delivery to tumor tissue and while avoiding the surrounding structures.

Intensity Modulated Radiation Therapy

Intensity modulated radiation therapy (IMRT) is an advanced form of three-dimensional (3D) radiotherapy. The patient undergoes a treatment planning CT scan, and the data from this scan is fed into a computer program that constructs a 3D model, allowing the radiation oncologist and radiation physicist make a treatment plan that delivers the full of radiation to the tumor and minimizing the radiation dose to the nearby structures. Most commonly, IMRT is delivered by daily treatment (Monday through Friday) for 4-7 weeks. Prior to the treatment, the patient has a mask custom made specifically to his or her facial structure. During the treatment, the mask is worn to hold the same position for each treatment. During the treatment, the radiation beams are conformed to the precise shape of the tumor.

Proton Beam Irradiation

Proton beam radiation protons differs from IMRT above in that protons (particles that are smaller than an atom) are used instead of photons (x rays). Proton radiation can be delivered even more precisely than photons, which means that radiation can be customized to the irregular shape of the tumor, avoiding sensitive nearby structures even better than IMRT. Proton irradiation is effective in patients with chordomas, chondrosarcomas and other skull base malignancies requiring high dose radiation in close proximity to vital neurovascular structures. Proton beam radiation has been studied in skull base adenoid cystic carcinoma and may allow higher dose radiation to the tumor with better local control without changing overall survival.

Stereotactic Radiosurgery

Stereotactic radiosurgery (SRS) is delivered in a single dose in a one-time setting. During this procedure, the patient undergoes a CT scan that is uploaded onto a computer to determine the area of the tumor in three dimensions. Next, a head frame is secured to the patient's skull using pins. The positioning of this is determined by CT scan. Stereotactic radiosurgery is most often used in treating benign tumors and conditions in the skull, such as vestibular schwannomas, meningiomas, and arteriovenous malformations (AVMs). The role of SRS in cancer has expanded in treating primary malignant neoplasms and metastases of cancers from other areas of the body to the skull. After treatment, the tumor may shrink or remain stable in size.

CHEMOTHERAPY AND IMMUNOTHERAPY



AHNS Skull Base Surgery Section Sinusal Patient Information

Chemotherapy and immunotherapy are cancer medications given by medical oncologists, usually by an infusion through the veins. These medications may be given alone or with radiation therapy. Sometimes a chemotherapy port is placed under the skin of the chest to reduce the number of IVs a patient will require for infusions. The specific types of medications given depend on the type of cancer, the tumor stage and the prior treatments. Side effects depend on the specific medication or combination of medications given. The goal of any treatment plan is to treat the tumor as effectively as possible while minimizing any side effects. An experienced team of cancer doctors will be able to explain the treatment that is best for any particular cancer and help the patient understand what side effects should be expected during treatment.