Operative management of skull base malignancies: choosing the appropriate approach

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Malignant tumors of the skull base are complex lesions. Identifying the indications and contraindications for resection is essential for the successful treatment of these lesions. This requires an understanding of the pathology, principles of resection, and nonsurgical therapeutic modalities. Choosing the appropriate surgical approach requires an understanding of the tumor and its association with the anatomy of the skull base. Preoperative assessment and preparation of the patient for the postoperative course, including functional and cosmetic deficits, are reviewed in the context of the specific approach. Anatomical variations encountered in the preoperative planning are discussed. A review of reconstructive alternatives is presented that is specific to the approach and anatomical violation. Finally, the use of a multidisciplinary team both in and out of the operating room is recommended, emphasizing a team approach during the resection itself.

KEY WORDS • clivus • infratemporal fossa • skull base • temporal bone • transfacial approach

Malignancies of the skull base account for a small but significant percentage of skull base lesions. These tumors may arise primarily from the skull base itself or invade secondarily. Those tumors secondarily invading the skull base may do so either through local invasion or by metastatic spread. From the perspective of the skull base surgeon we have found classification by anatomy to be the most useful in terms of predicting pathological composition and in treatment planning. From a structural point of view the malignancies are divided into three groups: 1) anterior skull base, 2) clivus, and 3) lateral skull base (infratemporal/middle fossa and temporal bone). As with any classification scheme it represents a continuum, and in some cases a combination of two or more of these groups is more applicable to the patient’s presentation.

As with any lesion involving the skull base, consideration of cranial nerve function is essential. A thorough examination of the cranial nerves is necessary to determine the patient’s premorbid status as well as to prepare the family adequately for any postoperative deficits that may affect the patient and the caregiver. Second, evaluation of the cerebrovascular anatomy, its involvement by the tumor, and its potential for injury and/or sacrifice must be understood by both the surgical team and the patient. Examination of the vascular anatomy should include the entire operating team, including the reconstructive surgeon(s), as the success of these cases requires a team approach. The preoperative assessment should consider the extent of resection and plan for the reconstruction.

PREOPERATIVE ASSESSMENT

Cranial Nerves

A thorough preoperative examination of the cranial nerves potentially involved by the tumor and/or the surgical approach is essential to assess any preoperative deficits that may either progress or require intervention during the patient’s recovery. Understanding the concomitant deficit and its effect on the patient either cosmetically or functionally and educating the patient and the caregiver(s) prior to surgery will allow the patient to make informed and appropriate decisions in their care. Although commonly overlooked, anosmia and its concomitant aguesia are not benign deficits and can be associated with significant postoperative lifestyle modifications for the patient and his/her family.
**Cranial Nerves Two, Three, Four, and Six.** The loss of vision, especially if present preoperatively can be traumatic for the patient. Many of these patients will require some form of radiotherapy postoperatively; thus, the potential for further diminution of vision over time must be considered in the context of the patient’s survival and the potential benefit expected from sacrifice of the optic apparatus. A formal visual field examination to identify even subtle involvement of optic apparatus should be performed preoperatively.

Extracranial dysmotility can be a significant and even disabling deficit for the patient. Preservation of the optic apparatus in the affected eye is of little consolation and in some cases even detrimental, to the patient. An honest evaluation by the surgeon(s) as to their own surgical series/results involving permanent and temporary extracranial dysmotility deficits must be discussed with the patient.

**Cranial Nerve Five.** Involvement of the trigeminal nerve complex by the tumor is usually quite apparent during the examination. In many cases of malignancies involving the trigeminal complex the patient will typically present with an exquisite neuralgia. This is not always the case, however, and in the early presentation may present with only minimal hypesthesia, masticatory muscle weakness/atrophy, or no deficit at all. In all malignancies of the middle cranial fossa, consideration of these complications either pathology-related or iatrogenic must be discussed with the patient. In most cases recognition of a trigeminal nerve deficit and early intervention can minimize any functional deficit in the patient.

**Cranial Nerves Seven and Eight.** As with any of the cranial nerves, preoperative assessment by the surgical team as to the benefit of a structure’s sacrifice must be weighed against the benefit of the patient’s survival and quality of life. Although not life-threatening, the loss of the facial nerve can have a tremendous psychological impact on the patient’s quality of life. The cosmetic implications of facial nerve loss as well as the functional implications including oral incontinence and vision loss due to exposure keratitis should be addressed early on in the patient’s recovery. Loss of the trigeminal and facial nerve concomitantly pose an even greater threat of exposure keratitis. Early gold weight implantation and lower lid tightening procedures should be implemented. Nerve repair at the time of the initial procedure, whether by primary anastomosis, cable graft, or transposition (cranial nerves 12–seven) should be considered if sacrifice of the nerve is necessary. A hypoglossal–facial nerve anastomosis can be performed at 12 to 18 months if the nerve remains paralyzed but was left structurally intact. Temporalis or masseter muscle transpositions are alternative procedures for functional deficit in the patient.

**Vascular Assessment**

Preoperative evaluation of the relationship of the tumor to the associated cerebrovascular anatomy cannot be overstated. Evaluation can be performed using either a formal angiogram or by MR imaging (MR angiography or MR venography). In most cases, the MR imaging technique is preferred because it is noninvasive; however, in those cases in which it is necessary to obtain flow characteristics, as in assessment of the patency and adequacy of the circle of Willis or when preoperative embolization of the tumor is necessary, obtaining a cerebral angiogram is essential. Although controversial, when sacrifice of a CA is contemplated, a balloon test occlusion in conjunction with a Xe-CT or positron emission tomography scan is helpful in identifying those patients at risk of cerebrovascular insufficiency. It is known, however, that 3% to 8% of those patients in whom a successful preoperative assessment is completed will develop an ischemic stroke. Therefore, when sacrifice of the internal CA is necessary, vascular reconstruction should be considered.

Second, identifying hypervascular tumors and the origin of their vascular supply may significantly reduce the perioperative blood loss and associated complications. The relationship of the major cerebrovascular structures to the tumor may help to determine the extent of resection and identify those patients in whom sacrifice of the vessel is necessary. Therefore, we have found the interventional neuroradiologist to be an indispensable part of a multidisciplinary approach to the tumors.

**Anterior Skull Base**

Malignancies involving the ASB are divided into three groups. The first group is composed of those tumors arising from the skull base primarily (Fig. 1). The second group secondarily invades the ASB locally from a primary nasal or paranasal site of origin. The third group includes metastatic deposits from distant primary sites (such as the prostate gland) usually via a hematogenous route. Even with the significant advances made in both radiation oncology and medical oncology, resection remains the mainstay in the effective treatment of many of these.
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Fig. 1. Sagittal Gd-enhanced MR image demonstrating a low-grade chondrosarcoma arising from the clivus and extending superiorly involving the floor of the ASB.

Fig. 2. Intraoperative photograph showing a bifrontal cranial section including superior orbital rims and nasion (cranioorbital-nasal) in preparation for resection of tumor seen in Fig. 1.

Fig. 3. Intraoperative photograph showing a pedicle pericranial flap to be used for reconstruction after anterior craniofacial resection.

En bloc resection of the ASB is indicated in those tumors involving the cribiform plate or fovea ethmoidalis. Patients presenting with extensive intracranial/intradural involvement or distant metastasis must be evaluated on an individual basis, as any extensive skull base resection will be only palliative. Approaches to the ASB are typically classified as intracranial, extracranial, or combined. Intracranial approaches to the ASB typically include a bifrontal craniotomy as the basis for the approach. A subfrontal approach incorporates osteotomies of the superior orbital rim(s) and/or nasion. These osteotomies can be performed in a one-piece manner with the craniotomy (Fig. 2) or as a separate osteotomy. The subfrontal approach allows wider exposure of the ASB, as well as the clival region, while limiting the amount of retraction of the frontal lobes of the brain. The dura serves as a barrier to the intracranial spread of the tumor. Dural resection in those patients with limited dural involvement, but without transgression into the intracranial space usually provides an adequate oncological margin; however, if there is violation of the intradural compartment, the prognosis declines significantly.

As many of the malignancies of the ASB arise primarily from the nasal and paranasal regions, an extracranial route of access is commonly used either alone or in combination with the intracranial exposure. Extracranial routes to the ASB can be performed either through transfacial incisions or facial degloving procedures. We typically use transfacial incisions for malignancies confined to the ASB and reserve sublabial degloving procedures for those tumors involving the sella and/or the clivus.

Meticulous attention to reconstruction following resection of the ASB is focused on the separation of the intradural compartment from the sinonasal cavity. In cases in which the dura has not been intentionally violated, a careful evaluation of the dura’s integrity must be made. Inspection for the presence of CSF should be performed including provocative testing by means of a moderate Valsalva maneuver. If there has been any violation of the dura either due to its resection or during provocative testing a meticulous watertight closure is necessary. This can be performed primarily or supplemented with grafts (autologous or dural substitutes) (Fig. 3). The use of onlay or “sutureless” dural grafts should be avoided in this area, as the integrity of the graft may fail resulting in a CSF leak and concomitant contamination of the intradural space. We have not found it necessary to reconstruct the skull of the ASB, and in fact, we recommend against this practice because any substance (autologous calvarial grafts or artificial constructs) that is exposed to the sinonasal cavity provides a nidus of infection (Fig. 4).

Clivus

As with malignancies of the ASB, malignancies of the clivus can arise 1) primarily (such as, chondrosarcomas...
chordomas, osteogenic sarcomas; Fig. 5); 2) secondarily from local invasion (such as nasal or paranasal sinuses); or 3) from distant metastatic deposits. As with ASB malignancies, contraindications to aggressive resection include poorly controlled systemic disease, extensive intradural involvement, or radiosensitive tumors (such as plasmacytoma). As such, an endoscopic biopsy of the lesion, if possible, should be considered prior to an extensive resection. As these tumors may secondarily involve the cavernous sinus, Dorello’s canal, jugular foramen, or the hypoglossal canal, a thorough cranial nerve examination is necessary preoperatively. In most cases obtaining both an MR imaging study for assessment of the soft tissues and a CT scan (Fig. 6) to assess craniofacial skeleton are necessary to determine the appropriate approach and its limitations. Additionally, when violation of the oropharynx is necessary, an examination of oral hygiene should be performed, and in some instances, a culture of the oral flora should be performed to identify any resistant bacterial strains that may be present. A preoperative assessment of the dentition is necessary, especially if sacrifice of the dentition is a possibility or when radiation therapy is considered for postoperative treatment.

Malignancies of the clivus can be approached through a direct anterior transfacial approach or a subfrontal approach (Fig. 2). The transfacial approaches as described previously can be performed via a facial incision or a sublabial degloving exposure. The choice of approach is determined by the area of the clivus involved: superior, middle or inferior third, and soft tissue of paranasal sinus extension; however, all of the anterior transfacial approaches are limited in their lateral extent of exposure. Superiorly the cavernous sinus limits the lateral extent of the exposure. Inferiorly, the jugular foramen and hypoglossal canal limit the lateral extent of exposure.

Those lesions confined to the sella turcica and superior third of the clivus can be adequately exposed through a sublabial transphenoidal approach. Tumors involving the lower two thirds of the clivus typically require a swing-door (extended) maxillotomy. This approach is also made through a sublabial incision. Once exposed, a LeFort II maxillotomy is performed. The hard palate and soft palate are then split either midline or paramedian allowing for the rotational lateral displacement of the maxilla. This provides excellent exposure of the entire clivus down to the level of the foramen magnum.

As with other reconstruction techniques associated with the ASB, the emphasis on obtaining a watertight barrier is essential. We typically use free tissue autograft rather than dural substitutes because of the exposure of the oropharynx and the propensity for infection (Fig. 7). Second, a meticulous closure of the oropharynx with attention to the
soft palate is required to provide both a vascularized tissue barrier and to provide the patient good return of nasopharyngeal function. We typically place a nasopharyngeal feeding tube at the time of surgery, and the patient remains on tube feedings until the oropharyngeal mucosa has healed. We do not routinely perform a tracheostomy with a maxillotomy, unless a concomitant mandibulotomy is performed.

Lateral Skull Base

Malignancies involving the LSB most commonly arise from lesions extending from the infratemporal fossa. In our series the majority of these tumors were neurofibrosarcomas associated with neurofibromatosis Type I (Fig. 8). Tumors arising from the parotid or from perineural spread of a paranasal carcinoma account for the majority of the remaining tumors in our series. A minority of these tumors arise directly from the temporal bone (such as squamous cell carcinoma). Another subset of tumors involving this region arises from the floor of the middle fossa. This latter group of tumors includes both solitary fibrous tumors as well as meningiomas secondarily invading the infratemporal fossa or temporal bone. Although histologically benign, the invasive nature of these aggressive tumors warrants comment in this text.24

The choice of the appropriate approach for removing malignancies of the LSB is made by knowing the anatomical location of the tumor and its extension and then obtaining vascular access both proximally and distally. Malignancies confined to the parapharyngeal area of the infratemporal fossa with involvement of the floor of the middle cranial fossa are further divided based on their location with respect to the cervical CA, either lateral or medial. Tumors primarily lateral to the CA or within the CA sheath are typically approached through a preauricular approach (Fig. 9). The approach is extended superiorly in a curvilinear fashion and is extended inferiorly along the anterior border of the sternocleidomastoid. We typically use a zygomatic osteotomy to allow better visualization of the floor of the middle fossa while reducing brain retraction. Exposure of the CA both along its cervical course as well as along its petrous course provides vascular control. As with en bloc resection of the ASB a similar approach can be performed along the middle fossa. Exenteration of the infratemporal–parapharyngeal space can be performed, including muscle and divisions of the trigeminal nerve if necessary. Early identification of the facial nerve is essential to avoid injury or, in cases of planned sacrifice, identify areas for nerve splicing.

Neoplasms located medial to the CA are typically approached through a swing-door mandibulotomy.13 This provides excellent visualization of the infratemporal fossa...
superiorly to the level of the floor of the middle fossa with less risk to the facial nerve. If necessary an extradural middle fossa exposure, similar to that described above, can be incorporated to provide distal access to the CA within the petrous canal.

Malignancies arising in the region of the temporal bone or jugular foramen are approached by a postauricular incision extending inferiorly along the anterior border of the sternocleidomastoid (Fig. 10). As with other lesions in the infratemporal fossa, a neck dissection providing access to the vascular structures is performed. The extent of temporal bone drilling performed is dependent on the tumor origin (that is temporal bone, jugular foramen, endolymphatic sac) and its extent. Although we have tremendous respect for the preservation of hearing, when planning for an extensive resection of a malignant neoplasm in this area, the appropriate amount of drilling should not be compromised. In cases of extensive involvement of the temporal bone, oversewing of the external auditory canal is performed, especially when hearing is not present (Fig. 11). We typically approach these lesions through a transjugular approach and/or total petrosectomy depending on the extent of the temporal bone involvement (Fig. 12).9,25–27

Continuous neurophysiological monitoring is performed in most of the approaches for LSB resections.11,12,14,20,37 Facial nerve monitoring of the orbicularis oculi and oris are performed. In cases of hearing preservation auditory brainstem responses are continuously monitored to assess both hearing and brainstem function at the level of the cochlear nuclei. The 10th cranial nerve is monitored using either a specialized endotracheal tube or direct electrode insertion into the musculature of the vagus nerve. Direct electromyography recording in the trapezius muscle and the tongue is used for monitoring cranial nerves 11 and 12, respectively. During surgery we use dissecting instruments insulated to the tip and connected to the monitoring system to assist in the early identification of nerves during the dissection.

The resection of these LSB tumors typically involves a large dead-space cavity, which may or may not include the overlying skin and in some cases the pinna of the ear. Additionally, intradural extension of the tumor and/or dissection may also involve a direct communication with the CSF with insufficient dural borders to obtain a watertight closure. Finally, as the tumor may extend anteriorly involving the paranasal sinuses and/or the oropharynx, a barrier of well-vascularized tissue may be necessary. Therefore, a complete spectrum of free tissue transfers, free vascularized soft-tissue flaps and pedicle flap transfers must be available at the conclusion of the resection; pre-

Fig. 10. Coronal Gd-enhanced MR image revealing the jugular foramen/infratemporal fossa neurofibrosarcoma shown in Fig. 8.

Fig. 11. Coronal Gd-enhanced MR image demonstrating an infratemporal fossa neurofibrosarcoma with involvement of the pinna and temporal bone, which necessitated resection of the pinna and external auditory canal.

Fig. 12. Axial contrast-enhanced CT scan revealing the temporal bone/infratemporal fossa neurofibrosarcoma, which necessitated total petrosectomy.
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ferably this is planned preoperatively and perioperatively. Identifying the need for procedures palliating oropharyngeal dysfunction due to violation or lower cranial nerve dysfunction also should be recognized early on, preferably preoperatively. As with the mandibulotomy patient, the need for an accompanying tracheostomy and/or feeding tube should be recognized early during the resection or early on during the postoperative course.

CONCLUSIONS

Malignancies of the skull base account for a small percentage of skull base lesions overall; however, their complexity requires more than a casual understanding of the pathological and anatomical nuances specific to each. The fundamental premise to be considered in every patient in whom an extensive resection for malignancy is considered is the goal and risks of the planned procedure. Can a total resection be achieved or is the procedure only palliative? Are there nonsurgical therapeutic adjuncts specific for the tumor that can successfully treat any residual or recurrence? What deficits, either functional or cosmetic, is the patient or his/her support system able to accommodate? Once the goal of surgery is understood, the planning of the entire procedure preoperatively is necessary. The planning should include all members of the surgical team, radiologists, and oncologists. Familiarity with a wide spectrum of surgical approaches is essential for the resection of these tumors, and this requires a multidisciplinary team of surgeons. We have found that familiarity within the surgical team and working together on a consistent basis breeds a greater degree of success compared with casual interactions. Therefore, the importance of a multifaceted surgical team including neurosurgeons, neurootologists, craniofacial surgeons, and reconstructive surgeons cannot be overemphasized.

References


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