Chapter 2
Surgical Approaches to the Craniocervical Junction for the Resection of Chordomas

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Introduction

The anterior aspect of the craniocervical junction consists of the lower clivus (beginning at the level of the jugular foramen) and C1. Surgical approaches to the craniocervical junction have historically been one of the most challenging and highly morbid procedures (10, 17, 25, 26, 37). The challenge of obtaining adequate surgical exposure in a small operative corridor coupled with the anatomic depth of surgery has historically limited many attempts at tumor resection (1, 5, 15, 44). Many advances in the surgical approaches, imaging, and better understanding of the surgical anatomy have improved the management of these tumors (1–3, 9, 12, 13, 21, 24, 30, 36, 37). The surgical approaches are influenced by several factors (5). These include: tumor type, size, and precise location, as well as the experience and individual preferences of the surgeon. The approaches can be broadly divided into anterior, anterolateral, and posterolateral types, depending on the main trajectory of the approach. All of these approaches are useful either individually or in combination with each other as dictated by the individual case at hand. It is therefore important for the surgeon to be well versed with the entire spectrum of the available approaches to tackle the problem at hand. The chordoma is the prototypical cranial base tumor and is an excellent model to study the complexities of managing tumors that are located at the craniocervical junction.

TUMOR PATHOLOGY AND DIFFERENTIAL DIAGNOSIS

Pathological lesions affecting the craniocervical junction are quite diverse, requiring a detailed radiological workup (4, 16, 19, 28, 31, 39, 46). The radiological workup of fibrous dysplasia consists of computed tomographic (CT) scans that show the cranial base as expansile and sclerotic, in a characteristic "ground glass," appearance. On magnetic resonance imaging (MRI), fibrous dysplasia is low-to-intermediate signal intensity on both T1- and T2-weighted sequences, often with small areas of hyperintensity on T2-weighted sequences, which are thought to be caused by cyst formation and hemorrhage.

The most common osseous tumors in this area include: chordomas, chondrosarcomas, benign osteomas, metastatic tumors, multiple myeloma, plasmacytoma, and ossification of the posterior longitudinal ligament. The radiological workup of chordomas and chondrosarcomas consists of fine-cut CT imaging in which there is demonstrated variations in bone density and bone erosion with an outline of a destructive expansile lesion (23, 25). Areas of punctuate calcification can also be present in up to 50% of studies. On MRI, chordomas are low or intermediate in signal intensity on T1-weighted sequences and are high in signal intensity on T2-weighted sequences. On MRI, areas of signal loss may be associated with calcification. The clivus can show areas of heterogeneous marrow signal. With contrast, chordomas can enhance variably, from low to high brightness. Differentiating chordomas from chondrosarcomas is difficult, although chordomas are traditionally described as occurring more in the midline, compared with chondrosarcomas (7). Benign soft tissue tumors include: meningiomas, schwannomas of the lower cranial nerves, and neurofibromas. Meningiomas may cause hyperostosis and bone remodeling that is best

visualized on CT bone-window imaging. On MRI, meningiomas are isointense on gadolinium-enhanced T1- and T2-weighted sequences.

SURGICAL ANATOMY OF THE CRANIOCERVICAL JUNCTION

Understanding the surgical anatomy of the bony structures of the lower clivus–C1, the articular processes involving these structures, the muscular attachments, the vertebral artery, and the caudal cranial nerves is critical for management of tumors in this region. The osseous anatomy of the craniocervical junction involves predominantly the basioccipital bone (via the foramen magnum), the clivus, the occipital condyles, and the atlas. The jugular foramen is bordered by the occipital bone medially and the petrous temporal bone laterally. Cranial nerves IX, X, and XI, and the jugular bulb are transmitted through the jugular foramen. The foramen magnum is actually the opening of the occipital bone, which is wider posteriorly than anteriorly. The occipital bone itself is divided into a squamosal segment, a basal segment, and the paired occipital condyles. Lateral to the paired condyles lays the jugular process of the occipital bone, which forms the posterior segment of the jugular foramen, and connects the squamosal and basal parts of the occipital bone. The jugular process is separated from the mastoid process by two grooves lateral to the jugular process. Within these grooves, the occipital artery runs medially and the posterior belly of the digastric muscle runs laterally.

The clivus rises at a 45-degree angle from the foramen magnum to join the sphenoid bone anteriorly. The occipital condyles articulate with the atlas, and are located lateral to the anterior half of the foramen magnum. The lateral masses of C1 articulate with the occipital condyles. The shape of the lateral masses of C1 allow for movement through their cup-shaped contour. This design allows for flexion and extension primarily. The transverse process of the C1 level projects anteriorly and laterally. Many important muscles attach to this transverse process, including the superior and inferior oblique, the levator scapulae, and the rectus capitis muscles. The longus capitis muscle attaches to the lower clivus and the rectus capitis muscle attaches anterior to the front of the occipital condyle. The C1–C2 articular surfaces are flat to facilitate translational motion and, more importantly, rotational motion. Approximately 50% of axial rotation in the cervical spine occurs at the C1–C2 level. The adaptation evolved to prevent anteroposterior and lateral translation of C1 on C2 is the odontoid peg, or the dens and its associated ligaments. The cruciate, alar, and apical ligaments maintain the stability of the odontoid peg to the occiput. The alar ligament of the odontoid process attaches to the medial tubercle of the occipital condyle. The ligaments maintaining the stability between the odontoid peg and C1 are the transverse atlantal, atlantoalar, and atlantodental ligaments. This design is unique in the spinal bony architecture.

Cranial nerves IX and X arise as multiple fascicles from the lateral aspect of the medulla, whereas cranial nerve XI arises from the medulla and the upper cervical spinal cord and travels upward behind the dentate ligament. All then enter the jugular foramen medial to the jugular bulb. Anterior and superior to the occipital condyle is the hypoglossal canal, which transmits the hypoglossal nerve. The third segment of the vertebral artery begins at the foramen transversarium of C2 and ascends through the C1 transverse process. Surrounded by a rich venous plexus, it proceeds above the arch of C1 and crosses medially behind the articular capsule of the atlanto-occipital joint to enter the dura in an oblique manner, traveling upward and anteriorly. This segment of the vertebral artery above C1 runs in the anatomic suboccipital triangle. This triangle is delimited by three muscles: the superior oblique, extending from the occipital bone to the C1 transverse process; the inferior oblique, extending from the C1 transverse process to the C2 spine; and the rectus capitis posterior major, extending from the C2 transverse process to the occipital bone.

SURGICAL APPROACHES

The surgical approaches can be divided into anterior, anterolateral, and posterolateral. All of these approaches can be useful either individually or in combination in the management of chordomas (2, 7, 10, 26). It is important to understand the areas of access provided by each approach and, more importantly, to understand the limitations and potential risks imposed (29).

The anterior approaches that are useful in accessing the anterior craniocervical junction can be broadly divided into transcranial (subfrontal transbasal) and transfacial. These are noted in Table 4.1.

TRANSMAXILLARY/LEFORT I, WITH MEDIAL MAXILLECTOMY

The transmaxillary approach is best suited for extradural lesions extending into the sphenoid and the upper and middle clivus with minimal lateral extension (7, 10, 18, 22, 32, 38, 40, 41). This approach provides the most direct access to the clival tumors in the ventral midline, such as chordomas and chondrosarcomas (11, 42). The transmaxillary approach further expands the conventional transnasal approach and is well suited for lesions that are too rostral on the clivus for a transoral approach (6, 8, 9, 43). In the transmaxillary approach, the lateral boundaries are defined by the medial pterygoid plates, the internal carotid arteries (ICAs) at the level of the foramen magnum, the cavernous sinus, the hypoglossal canals, and the jugular foramen. An extension of this approach can be performed via a midline sagittal split of the maxilla and the soft palate to provide a further caudal exposure to the craniovertebral junction.

Case Illustration of the Transmaxillectomy Approach

A 27-year-old, left-handed woman presented with headaches since 1995. The headaches were severe and progressive and were worse during her menstrual cycles. During the workup, she had an MRI scan performed, which revealed a tumor in her lower clivus (Fig. 2.1). A transnasal endoscopic biopsy was performed and revealed the tumor to be chordoma. Aggressive treatment was planned. Because of the tumor's midline location, with no lateral extension, a maxillectomy approach was undertaken (Fig. 2.2). She underwent a transfacial approach to the clivus, which consisted of a bilateral maxillotomy and a medial maxillectomy. In this approach, the prevertebral muscles were taken down and the opening in the sphenoid sinus was visualized. Using the navigation system, the rostral, caudal, and lateral limits of the tumor were defined. The clival bone was carefully drilled down. The retropharyngeal portion of the tumor was clearly visible after the longus colli muscles were removed. The tumor was visualized and removed in a piecemeal fashion. The tumor had eroded through and gone through the periosteal layer of the dura. The inner layer of the dura towards the brainstem was, however, intact. The retropharyngeal portion of the tumor was also removed in a progressive fashion. The remainder of the clivus-involved bone was drilled until no more abnormality was identified. Intraoperative image-guided navigation and endoscopic inspection was very helpful. It appeared that we were almost at the level of the hypoglossal canal, which was left intact and not interfered with. The pharyngeal flap was sutured with the Mitek anchor in place. A gross total removal of the tumor was achieved along with resection of the surrounding bone.

Postoperatively, the patient did very well and was kept on a soft diet. She was discharged to her home a week after the operation. Postoperatively, she had minor facial swelling, although there was no evidence of a cerebrospinal fluid (CSF) leak. Her bite and swallowing were normal. A postoperative MRI scan demonstrated no residual tumor, and proton-beam therapy was withheld until necessary Fig. 2.3).

TRANSMANDIBULAR APPROACH

The transmandibular approach is similar to the transoral and transmaxillary approach. It provides midline access to the lower clivus and the upper three vertebrae. Compared with the transmaxillary approach, the transmandibular approach provides a more caudal exposure (27, 36, 45). It is best suited for extradural tumors. In the transmandibular approach, a midline incision is made from the lower lip to the hyoid bone, involving the midline mandibulotomy and often with a median glossotomy. The dissection in this approach follows the floor of the mouth posteriorly toward the glossopharyngeal sulcus. The mandible and tongue segments swing laterally. The removal of the posterior hard palate can expose the bony nasal septum and the anterior inferior surface of the sphenoid sinus. An inverted U-shaped pharyngeal flap is hinged downward, and the prevertebral muscles are reflected caudally. Intraoperative image guidance is helpful in guiding the approach, resection, and bone drilling.

Case Illustration of Transmandibular Approach

A 59-year-old, right-handed man presented with a month-long history of numbness and heaviness of his tongue that progressed to slurring of his speech. For the previous month, he complained of upper neck pain and a chronic sinus problem. He reported difficulty breathing for approximately 1 year previously. More recently, he developed difficulty swallowing with both liquids and solids, although he denied any choking episodes. His neurological exam was

significant for slurred speech and the remainder of his cranial nerve exam was normal. He had some difficulty with tandem gait testing, although no clear ataxia was noted.

The patient's preoperative MRI scan demonstrated a large lower and mid clival mass that seemed to be extradural (see Fig. 2.4). The tumor was adjacent to the right hypoglossal canal and jugular foramen. A CT scan showed bone destruction in the area of the lower clivus and the hypoglossal canal.

On the basis of the tumor location and morphology, a transfacial transmaxillary approach to the clivus in the craniocervical junction on the right side was performed. During the surgery, it was noted that the tumor extended from the level of the mid clivus all the way down to the level of C1. The tumor appeared to be epidural in location as well as retropharyngeal, and the lower part of the tumor could not be accessed through the maxillotomy approach. To avoid disruption of the palate, this approach was not converted to an open-door maxillotomy. Because of the limitations to this initial approach, a second staged surgery via a transmandibular approach was planned 4 weeks later. In this surgery, the patient underwent a tracheostomy, and a transmandibular approach through a midline mandibulotomy and glossotomy was performed. The posterior pharyngeal wall was exposed and no obvious bulge of the tumor could be seen. The longus colli muscle was reflected laterally and the tumor was clearly exposed. The lower pole of the tumor was at the level of C1–C2 on the patient's right side. The bone edges of the clivus that were eroded through were carefully defined, and the tumor was removed and separated from the bone and dura. The tumor extended further on the right than the left. The jugular foramen and the hypoglossal canal were not seen during this resection. There was no breach of the dura. Using an endoscope, the dura and the edges of the resection were examined. Although an area of dural invasion was noted, it was not excised because of concerns of postoperative CSF leakage. The addition of the transmandibular approach allowed the lower pole of the tumor to be clearly seen, enabling safe resection of this portion of the tumor under direct vision. The complete surgical resection could not have been performed through a single transmaxillary approach without dividing the hard and soft palate.

Postoperatively, after 3 months, the patient had normal tongue movement and swallowing function (Fig. 2.5). His neck pain also resolved.

ADVANTAGES AND DISADVANTAGES OF THE ANTERIOR APPROACHES

The anterior transfacial approaches provide direct access to the anterior craniocervical junction. They are best suited for tumors that are strictly in the midline and have no intradural extension. The surgeon avoids brain retraction and manipulation of cranial nerves using these approaches. The tumor is reached before the brain stem and, thus, manipulation of the brainstem is also minimized. The relation of the tumor to the hard palate must be assessed on the sagittal images to determine the limitation posed by this structure in the vertical dimension. Craniocaudal access can be maximized by selecting the proper approach or by using the approaches in combination. Tumors with intradural extension are usually not suitable for these approaches, because the options for closure of the dural defect and reconstruction are very limited, thus, predisposing to the risk of a CSF fistula (37). Limited access to the lateral portion of the tumor is a major drawback of these anterior transfacial approaches. The lateral limitations are imposed by the pterygoid plates, the parapharyngeal space, the jugular foramen, and the hypoglossal canals. The vascular structures are covered by the pharyngeal mucosa and should not be exposed to the pharyngeal contamination. Intraoperative image guidance is very useful in planning the approach and during the tumor and bony resection.

ANTEROLATERAL APPROACHES

The Preauricular Infratemporal Fossa Approach

The preauricular infratemporal fossa approach allows access to the clivus and infratemporal fossa. The caudal limit of this exposure is the hypoglossal canal (33, 35). The preauricular infratemporal fossa approach starts with a frontotemporal craniotomy with a supraorbital and zygomatic osteotomy. The mandibular condyle is dislocated from the joint, and the greater sphenoid wing is cut down with rongeurs to remove the roof of the foramen ovale and rotundum, such that there is direct access to the pterygoid plate (33). Next, the roof of the petrous carotid is completely removed and displaced laterally, after establishing proximal control in the neck. The clival bone medial to the carotid is then drilled away to provide access to the clival pathology.

Advantages and Disadvantages of this Approach

The main advantage of the preauricular infratemporal fossa approach is that it allows an extrapharyngeal access to the area in front of the brainstem, working anterior to the cranial nerves (36). It is primarily useful for extradural tumors (7, 9). The disadvantages are that it provides a unilateral route that is quite limited in width (9). This approach involves an extensive amount of dissection and mobilization of the entire petrous internal carotid artery (9). The temporomandibular joint is disrupted and the Eustachian tube is necessarily divided.

Case Illustration of the Preauricular Infratemporal Fossa Approach

An 11-year-old boy presented with a 4-month history of lack of coordination and weakness involving the left hand. Additionally, he was noted to have a left-sided facial droop. On neurological examination, his extraocular movements were full and no nystagmus or diplopia was noted. He had a slight facial droop on the left side that seemed to be a central facial weakness. Motor examination suggested mild weakness of his left upper extremity of approximately grade 4/5. He also had weakness of his left lower extremity of approximately grade 4/5. His MRI scan showed a very large tumor involving the clivus. It was isointense on T1-weighted sequences and hyperintense on T2-weighted sequences (see Fig. 2.6). The basilar artery was pushed back into the brainstem and there was a severe amount of brainstem compression. The CT scan showed some bone destruction in the clivus, greater on the right side, and the destruction went all the way down to the level of the hypoglossal canals. There did not seem to be destruction of the occipital condyles. There was some retropharyngeal tumor also noted more on the right side of the patient.

The patient underwent a preauricular subtemporal and infratemporal fossa approach that consisted of a right frontotemporal craniotomy and orbitozygomatic osteotomy. The approach to this predominantly extradural tumor necessitated decompression and mobilization of the entire petrous carotid artery, including the vertical segment. The entire tumor and involved bone of the clivus was drilled down from the petrous bone to the foramen magnum. The clival dura was opened, and it was noted that the sixth nerve was displaced high up by the tumor, and the grayish tumor was visualized. It was clearly apparent that the tumor was invaginating from the mid clivus into the brainstem, and that the upper pole of the tumor was completely capped by the upper brainstem. The tumor had the typical grayish soft consistency of a chordoma. The tumor was debulked and removed in a piecemeal fashion. After tumor removal, the clival dura was excised in a systematic fashion and the cranial nerves were visualized all the way down to C1. The anterior portion of the cervicomedullary junction and the anterior spinal artery were seen. Both vertebral arteries were seen and the vertebrobasilar junction was clearly exposed. Postoperatively, the patient's left-sided hemiparesis resolved and he had a temporary right VIth cranial nerve palsy. Postoperative MRI scans showed evidence of residual tumor along the anterior portion of the clivus as well as the cerebellopontine angle and the foramen magnum (Fig. 2.6). This residual portion was treated with proton-beam therapy, and, 3 years since surgery, the patient is currently without evidence of tumor growth on serial MRI scans.

LATERAL APPROACHES

Lateral approaches include the transtemporal approaches and the extreme lateral approach.

The Extreme Lateral Approach

In the extreme lateral approach, the conventional posterior and suboccipital exposure is modified with additional bone removal of the mastoid process and the articular pillars to truly obtain a lateral view of the brainstem (3, 14, 34, 36, 37). In this approach, the muscles of the posterior cervical triangle are elevated in layers. The muscles are detached from the occipital and the mastoid region and reflected inferiorly, anteriorly, and posteriorly. The transverse process of C1, the lateral mass of C2, and the spinous process of C2 are crucial landmarks in conducting the soft tissue dissection. The vertebral artery is exposed between C2 and C1 and then above C1. The artery is extracted from the foramen transversarium of C1 and displaced posteriorly. Through this dissection, there is a direct corridor created to the entire ipsilateral lateral mass of C1, the C1 arch, the occipital condyle, the odontoid process, and the body of C2. A posterior fossa craniectomy and a mastoidectomy are performed, typically with preservation of the labyrinth. The roof of the sigmoid sinus is removed and the sigmoid sinus is followed into the jugular bulb. The jugular bulb is

exposed to the point where it joins the internal jugular vein, uncovering the mastoid segment of the facial nerve. Access to the anterior aspect of the craniocervical junction is gained by drilling the occipital condyle and lateral mass of C1. Resection of the occipital condyle for chordomas destabilizes the joint and necessitates an occipitocervical fusion at the same time or a separate sitting.

Advantages and Disadvantages of the Lateral Approaches

The transtemporal and extreme lateral approaches allow an extrapharyngeal access to both intradural and extradural tumors with equal ease (20, 34). These approaches provide for full control of the vertebral artery, sigmoid sinus, and jugular bulb, and the cranial nerves VII to XII on one side. The surgeon is able to visualize the tumor–brainstem interface tangentially, facilitating its separation. The disadvantages are that these are unilateral approaches, requiring an extensive amount of soft tissue and bony dissection that can be quite time consuming (9). These approaches require a thorough knowledge of the temporal bone anatomy. Exposure of the contralateral structures across the anterior midline becomes quite limited.

Case Illustration of the Extreme Lateral Approach

A 60-year-old, right-handed man presented with pain, tingling, and numbness in his right upper extremity with associated progressive weakness and atrophy of the right half of his tongue. His neurological exam was significant for very poor hearing in his right ear with inability to hear a whisper on the right side. He had atrophy of the right half of his tongue and a decreased gag reflex on the right side. He had paralysis of his right sternocleidomastoid muscle, with right upper extremity weakness in all muscle groups, especially the right deltoid, which was 3/5, whereas the remaining muscle groups in the right upper extremity were 4/5. The left upper extremity and both lower extremities showed normal strength. Deep tendon reflexes showed hyperreflexia in all four limbs in a symmetrical fashion.

A preoperative MRI scan demonstrated a large craniocervical chordoma (see Fig. 2.7). The tumor extended into the jugular foramen on the right side and was also intimately involving the articulation of the occiput with C1 on the right.

The patient underwent two staged surgeries, first an extreme lateral approach, where the tumor was resected in the retropharyngeal space and in the region of the jugular bulb and occipital condyle. In this surgery, the lower cranial nerves were found to be grossly involved and invaded by the tumor. A gross total removal of the tumor was achieved. A tracheostomy was also performed at this time to protect the patient's airway. Because the first surgery was a destabilizing procedure, the second stage was an occipitocervical fusion down to C3 using titanium rod and sublaminar wires and bone from the patient's left posterior iliac crest.

Postoperatively, the patient did well and was weaned off the tracheostomy (Fig. 2.8). His speech and swallowing studies improved with time. His hemiparesis rapidly recovered to normal. His postoperative course was complicated with meningitis that was treated with intravenous antibiotics.

SUMMARY OF OUR CRANIOCERVICAL CHORDOMA EXPERIENCE

Our experience in treating craniocervical junction chordomas was recently analyzed. A population of 20 patients with these lesions was identified from our database, and all charts and imaging were examined. Detailed neurological examinations and postoperative imaging were reviewed. Chordomas at the craniocervical junction commonly present with neck pain (71%), headaches (35%), and lower cranial nerve involvement (47%) (Fig. 2.9). Midline resections have resulted in residual tumor and progression. We mostly performed the extreme lateral (59%) and transfacial transmaxillary (24%) approaches (Fig. 2.10). In 55% of patients, a second surgery was necessary for a gross total resection, which was achieved in 80% of patients. Occipital-cervical fusion was performed in 71% of patients. Postoperative proton-beam therapy was performed in 65% of patients. There was a 29% tumor recurrence rate and 20% of cases incurred new cranial nerve morbidities. CSF leakage (29%) and wound infection (10%) were seen among the morbidities.

Surgical resections limited to midline approaches (i.e., transoral) result in higher incidences of residual tumor because of limited access to lateral portions of the tumor. We favor single or combined approaches in a single sitting

or in multiple stages to permit as complete a resection as possible. Neurosurgeons managing these tumors at the craniocervical junction must be well versed in a variety of surgical approaches that can be judiciously used to manage this difficult problem.

Table 4.1

Approach	Area of access
Subfrontal transbasal	Sphenoid sinus, clivus
Transsphenoidal	Sphenoid sinus, clivus
Transoral	Clivus, C1, C2, retropharynx
Transmaxillary	Clivus, retropharynx
Transmandibular	Clivus, C1, C2, retropharynx, parapharyngeal

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FIG. 2.1. Preoperative T2-weighted axial MRI showing tumor (arrow).
FIG. 2.2. A, intraoperative photograph of the surgical corridor created in the transmaxillary approach; and B, the

stereotactic navigational image.

FIG. 2.3. Postoperative T2-weighted axial MRI after tumor resection.

FIG. 2.4. A, preoperative T1-weighted sagittal MRI. B, T2-weighted axial MRI of the craniocervical tumor. Arrows indicate the tumor.

FIG. 2.5. A, postoperative T1-weighted sagittal MRI. B, T2-weighted axial MRI after tumor resection.

FIG. 2.6. A, preoperative T2-weighted sagittal MRI; arrow indicates the tumor. B, postoperative T1-weighted sagittal MRI with gadolinium contrast enhancement.

FIG. 2.7. Preoperative T2-weighted axial MRI demonstrating significant craniocervical tumor; arrow indicates the tumor.

FIG. 2.8. Postoperative T-weighted axial MRI demonstrating tumor resection.

FIG. 2.9. Preoperative presenting symptoms in the chordoma series.

FIG. 2.10. Initial surgical approach used in the chordoma series.