Surgical Management of Petroclival Chordomas: Report of Eight Cases

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ABSTRACT

In the management of skull base chordomas, surgical treatment is essential to achieve long-term control. A petroclival chordoma growing laterally in the skull base is one of the most challenging tumors for neurosurgeons. We have treated petroclival chordomas based on the principle of maximal surgical resection of the tumor with minimal morbidity. Lateral skull base approaches were used to approach petroclival chordomas in eight patients. The surgical procedure involved removal of soft tumor tissue and extensive drilling of adjacent bony structures. Gross total resection of the tumor was achieved in six patients. Subtotal resection in the remaining two patients was associated with acceptable morbidity. In cases of petroclival chordomas, lateral skull base approaches can be used as a primary procedure, although those approaches may be associated with high rates of morbidity and mortality.

KEYWORDS: Chordoma, petrous bone, skull base, transpetrosal approach

Chordomas are rare primary bone tumors originating from remnants of the primitive notochord at the two extremes of the vertebral axis, the skull base and sacrum. Skull base chordomas usually arise as encapsulated tumors, but they infiltrate adjacent bony structures with destructive alteration of the skull base. They grow slowly but can manifest characteristics of a malignant tumor, being locally aggressive with a tendency for regrowth. Although

it is possible to concentrate high radiation doses to the lesion since the development of stereotactic radiosurgery (SRS), surgical treatment is essential for the definitive management of skull base chordomas.¹⁻⁴ The best prognosis appears to be associated when maximal tumor resection is achieved at the first operation.^{1,5}

The surgical approach for skull base chordomas should be based on their extension patterns

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through the skull base.² Skull base chordomas can grow anteriorly, inferiorly, or laterally. Chordomas growing anteriorly, but restricted to the central compartment of the skull base, can be removed maximally through a midline ventral approach such as a transsphenoidal or transoral approach. Chordomas growing anteriorly with soft tissue invasion or growing inferiorly to the craniovertebral junction require two or more skull base approaches such as the midfacial degloving approach to achieve gross total resection.^{6,7}

Chordomas growing laterally in the skull base, so-called petroclival chordomas, are also challenging tumors for neurosurgeons. We now report our surgical experience with eight patients with petroclival chordomas who had not undergone previous surgery.

METHODS

Patient Population

Between 1986 and 2004, we treated 16 patients with skull base chordomas. Patients who had undergone primary surgery or radiation therapy were excluded from the study. Of the 16 patients, 8 patients (mean age, 40 years; range, 22 to 67 years) had petroclival chordomas (Table 1). All patients received a comprehensive preoperative evaluation that included computed tomography (CT), magnetic resonance imaging (MRI), and conventional angiography or digital subtraction cerebral angiography.

Management Protocol

The goal of surgery was to achieve maximal surgical resection of the tumor with minimal morbidity. Various surgical approaches were used, alone or in combination, based on the tumoral extension. The diagnosis of chordoma was confirmed pathologically in all patients. SRS was introduced as the

postoperative adjuvant therapy based on the extent of tumor resection and on postoperative MRI. After their initial treatment, all patients were evaluated regularly with MRI. If tumor recurrence was detected, repeat surgery or SRS was indicated. The mean length of follow-up was 110 months (range, 9 to 226 months).

Assessment of Function

Function was assessed before surgery and at the most recent follow-up. Patients were classified into five grades of disability as follows: Grade 0: normal or able to perform normal activity; Grade 1: normal activity with effort, but without assistance; Grade 2: requires occasional assistance, or considerable assistance and frequent care; Grade 3: disabled or severely disabled; Grade 4: very sick or moribund.

RESULTS

The eight patients underwent 12 surgical procedures, including the orbitozygomatic infratemporal approach, transzygomatic subtemporal approach, extended middle fossa approach, total petrosectomy approach, and anterior and posterior combined transpetrosal approach. The surgical procedure involved removal of the tumor tissue and extensive drilling of adjacent bony structures. Gross total resection of the tumor was achieved in six patients (Table 1). The two patients with subtotal resection underwent SRS to treat their residual tumors 1 month after surgery. At the last follow-up review, only one patient had undergone repeat surgery, and three other patients had undergone SRS for tumor recurrence. Three patients have no evidence of tumor recurrence. The rate of surgical mortality was 0%. The mean preoperative disability grade was 0.5. At the most recent follow-up review, the mean postoperative disability grade was 0.6.

Table 1 Patient Characteristics and Clinical Results

Pt	Age (yrs), Sex	Initial symptom	Tumor side	Preop ADL	Primary surgery	Extent of resection	SRS early after surgery	Recurrence	Repeat surgery	SRS for recurrence	Follow-up (mos)	Latest ADL
1	59, F	Diplopia	R	0	Orbitozygomatic infratemporal	Gross	-	+	Combined transpetrosal	+	226	1
2	22, F	Diplopia	L	0	Orbitozygomatic infratemporal	Gross	-	-	-	-	186	1
3	54, F	Asymptomatic	R	0	Extended middle fossa	Gross	-	+	-	+	153	1
4	67, M	Diplopia	R	1	Orbitozygomatic infratemporal	Gross	-	+	-	+	144	0
					Extended middle fossa							
5	33, F	Diplopia	R	1	Extended middle fossa	Gross	-	-	-	-	113	1
6	25, M	Hearing disturbance	R+L	1	Left transzygomatic subtemporal	Sub-total	+	-	-	-	24	0
					Right total petrosectomy							
7	41, F	Facial numbness	R	0	Extended middle fossa	Gross	-	-	-	-	23	0
8	22, F	Dizziness	R+L	1	Right combined transpetrosal	Sub-total	+	-	-	-	9	1
					Left lateral suboccipital							
Mean				0.5							110	0.6

ADL, activity of daily life; SRS, stereotactic radiosurgery; +, positive; -, negative.

ILLUSTRATIVE CASES

Patient 6

A 25-year-old man gradually developed a hearing disturbance and dysesthesia on the right side of his face and mild motor weakness of his right upper and lower extremities. Neurological examination revealed paralysis of the right abducens nerve, mild hyperalgesia on the right side of his face, moderate peripheral facial palsy (modified House-Brackmann grade 3/6), hearing loss on the right side, and grade 4/5 right hemiparesis. CT showed a large tumor extending from the clivus to the petrous bone, predominantly on the right side, associated with calcification and bony destruction (Fig. 1A). The tumor was hypointense on T1-weighted MRI and hyperintense on T2-weighted MRI. On postcontrast T1-weighted MRI, scattered foci of increased intensity extended into the brain stem (Figs. 1B, C).

The patient first underwent a left transzygomatic subtemporal approach to remove the tu-

mor growing into the brain stem. A thin fibrous membrane, identified as stretched dura mater, intervened between the tumor and brain stem. The tumor was successfully removed from the brain stem by following this membrane. After surgery, the patient's right hemiparesis recovered with no deterioration of other neurological signs. Postoperative MRI confirmed gross total resection of the tumor. Pathological examination confirmed the diagnosis of chordoma.

The second surgery, a right total petrosectomy approach, was performed to remove a tumor extensively invading the skull base. The total petrosectomy approach involved extensive drilling of the petrous bone using a transcochlear technique in addition to a combined anterior and posterior transpetrosal approach.⁸ The tumor was removed both epidurally and subdurally. The facial-auditory nerve complex was severely elongated by the tumor, with dura mater between the nerve and tumor tissue (Fig. 2A). The trigeminal nerve was elongated superiorly by the tumor (Fig. 2B).

Figure 1 Patient 6. (A) Preoperative CT using a bone algorithm shows bone destruction in the skull base. (B) and (C) Preoperative postcontrast T1-weighted MRIs show an invasive tumor in the petroclival area extending into the brain stem. (D) Postoperative CT using a bone algorithm shows the right total petrosectomy. (E) and (F) Postcontrast T1-weighted MRIs obtained 3 months after surgery show the subtotal resection of the tumor and possible residual tumor in the lower clivus. CT, computed tomography; MRIs, magnetic resonance images.

Figure 2 Patient 6. Intraoperative photographs show the surgical steps of the right total petrosectomy approach. (A) The facial-auditory nerve complex (VII and VIII) is severely elongated by the tumor from which it is separated by the dura mater (asterisk) in between. (B) The trigeminal nerve (V) is elongated superiorly by the tumor, which was removed epidurally. (C) Basal dura mater in the clivus was perforated by a calcified portion of the tumor (double asterisks). (D) Careful tumor removal during the final stage of surgery shows the basilar artery, both AICAs, and both sixth cranial nerves (VI). AICAs, anterior inferior cerebellar arteries.

The basal dura mater in the clivus was perforated by a calcified portion of the tumor. The flattened facial nerve, which was sectioned for radical resection of the tumor, was reconstructed with fibrin glue. Careful tumor removal revealed the basilar artery, both anterior inferior cerebellar arteries (AICAs), and both sixth cranial nerves (Figs. 2C, D). The tumor close to the jugular foramen was carefully removed so as not to damage the jugular bulb. Dural and bone reconstruction was then completed.

After the second surgery, the patient developed paralysis of the right trochlear nerve. His right facial palsy and right hearing loss were aggravated but there were no other deteriorating neurological signs. Postoperative CT and MRI showed the total petrosectomy and residual tumor in the lower clivus, which was treated with SRS (Figs. 1D–F).

Patient 8

A 22-year-old woman presented with the gradual onset of dizziness and mild motor weakness of her left upper and lower extremities. Her neurological examination revealed horizontal nystagmus and grade 4/5 left hemiparesis. CT showed a large tumor extending in the skull base and associated with a cyst formation, calcification, and bony destruction of the clivus and petrous bone (Fig. 3A). The tumor was hypointense on T1-weighted MRI and hyperintense on T2-weighted MRI. On post-contrast T1-weighted MRI, scattered foci of increased intensity extended into the brain stem (Figs. 3B, C).

She first underwent a right anterior and posterior combined transpetrosal approach.⁸ The tumor appeared to be partially adhesive to the brain stem. The facial and auditory nerve complex was displaced laterally by the tumor. Careful dissection of the tumor from the brain stem revealed the cyst in the brain stem. The cyst appeared to be covered by neural tissue, and not by tumor tissue. Pathological examination confirmed the diagnosis of chordoma. Postoperatively, she suffered facial dysesthesia on the right side and bilateral abducens palsy. Postoperative CT and MRI showed the partial petrosectomy, shrinkage of the cyst in the brain stem, and the residual tumor in the left cerebellopontine angle (Figs. 3D–F). She subsequently underwent a left lateral suboccipital approach and then developed hydrocephalus. A ventriculoperitoneal shunt was placed. Residual tumor in the lower clivus was treated with SRS.

DISCUSSION

Although chordomas are usually regarded as slow-growing tumors, they often recur soon after surgical treatment. The malignant potential of chordomas lies in their tendency to infiltrate bone, which leads to their high recurrence rate after treatment. Most repeat operations for tumor recurrence have been

Figure 3 Patient 8. (A) Preoperative CT using a bone algorithm shows the bony destructive alterations in the skull base. (B) and (C) Preoperative postcontrast T1-weighted MRIs show an invasive tumor in the petroclival area extending into the brain stem. (D) CT using a bone algorithm obtained after the first surgery shows the right partial petrosectomy. (E) and (F) Postcontrast T1-weighted MRIs obtained after the first surgery show subtotal resection of the tumor. Residual tumor in the left cerebellopontine angle was subsequently removed via a left lateral suboccipital approach during the second surgery. CT, computed tomography; MRIs, magnetic resonance images.

performed 2 or 3 years after primary treatment; few repeat operations are performed more than 5 years after initial surgery.⁵ Permanent postoperative functional deterioration and improvement have been reported in 40% and 20% of patients with chordomas or chondrosarcomas, respectively.¹ The estimated overall survival rates for patients with chordomas are 51% to 86% and 35% to 69% at 5 and 10 years after resection, respectively.^{1,5,9,10}

In the management of skull base chordomas, surgical treatment has a definitive role. Longer survival rates have been associated with more extensive tumor removal, although aggressive surgical removal can also be associated with higher rates of morbidity and mortality than more conservative resections.^{1,2,4} The deep position (at the skull base) and the tendency to infiltrate bone make total removal of skull base chordomas difficult. Chordomas are usually extradural midline tumors and hence displace the neuraxis dorsally or dorso-laterally. Anterior midline extradural approaches such as the transsphenoidal or transoral approach

are preferred.^{5,10,11} In cases with an extensive lateral extension, however, an anterior midline extradural approach is inadequate to obtain maximum safe resection of the tumor. Extension through the skull base is highly characteristic of skull base chordomas and is the main limiting factor in their surgical management. Al-Mefty and Borba emphasized that the use of various skull base approaches based on the degree of tumor extension is associated with low rates of mortality and morbidity.²

In our series, the surgical approaches were tailored according to their extension patterns in the skull base. An extended middle fossa approach or orbitozygomatic infratemporal approach was used to remove tumors that extended laterally into the petrous bone but were restricted anteriorly to the internal auditory meatus. The more complex anterior and posterior combined transpetrosal approach was used to remove a tumor that grew more extensively in the lateral direction in the skull base. In Patient 6, a total petrosectomy approach was used to remove the tumor safely. The tumor invaded

subdurally and appeared to be encapsulated by a thin fibrous membrane. It was released from the brain stem using the lateral extradural corridor.

Lateral skull base approaches such as the extended middle fossa approach or the total petrosectomy approach to the petroclival area were pioneered by Hitselberger and House as the "transcochlear approach" and by Morrison and King as the "translabyrinthine-transtentorial approach."¹²⁻¹⁴ Fisch and Pillsbury described the infratemporal approach, which combined a partial posterior and inferior petrosectomy with a cervicofacial approach.¹⁵ The combination of the suboccipital-subtemporal approach to improve exposure of the petroclival area was popularized by Malis as the "petrosal approach."¹⁶

The lateral skull base approaches via the temporal bone have been modified to enhance exposure of the petroclival area.^{8,17-24} With bony resection of the petrous bone, several modifications have been useful to obtain wide exposure of the petroclival area. The temporomandibular joint was exposed to increase the width and ease of surgical access to the anterior petrous region.^{22,23} The labyrinthine and tympanic segments of the facial nerve can be mobilized posteriorly to obtain wide exposure of the anterolateral aspect of the petroclival area. The technique of posterior mobilization of the facial nerve in the extended middle fossa approach was detailed by Goel.²⁴ These additional maneuvers provided the necessary increase in exposure to access lesions in the petroclival area.

Spetzler and colleagues divided these approaches through the temporal bone into three variations. The retrolabyrinthine technique involves petrous bone resection and preserves hearing. The translabyrinthine technique requires more extensive petrous bone resection and hearing is sacrificed. Finally, the transcochlear technique involves maximum petrous drilling, the sacrifice of hearing, and the transposition of the facial nerve.²⁵ These technical evolutions have helped to provide a wide and horizontal exposure to the petroclival area with minimal brain retraction.

Although the number of cases presented here is limited, petroclival chordomas can be removed maximally and safely using lateral skull base approaches. Postoperative close follow-up of the patients is mandatory. In place of adjuvant proton-beam therapy, SRS may be enough to control the tumor in the long term.

CONCLUSION

In the management of skull base chordomas, one of the most important factors is maximal tumor removal during the first operation. Considerable expertise is required to achieve maximal surgical resection of the tumor with minimal morbidity. In cases of petroclival chordomas, a midline ventral approach such as the transsphenoidal or transoral approach may be inadequate. More complex lateral approaches can be used as a primary procedure.

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Commentary

The authors present eight patients who underwent surgical treatment for petroclival region chordomas. Their approach to these complex patients incorporates many aspects critical to the treatment of chordomas: maximal surgical resection, use of a variety of skull base approaches tailored to the location of tumor, extensive resection of bone surrounding the soft tissue tumor component, and adjunctive radiation therapy. The authors used stereotactic radiosurgery (SRS) to treat residual and recurrent tumors. We prefer the use of proton beam therapy, which has the longest treatment record and highest success rate associated with the treatment of chordomas. SRS is more readily accessible than proton beam therapy, which would make treatment with this modality advantageous. However, a large series of patients treated with SRS would be required to ascertain the role of this modality in the management of chordomas.

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Commentary

In this article, Takami and colleagues have presented their experience with chordomas involving

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the petroclival area. Patients who had previously been operated on and/or irradiated, and who are the most difficult cases to treat, were excluded from the study. Therefore, the number of cases presented is limited. The authors' intent was to perform complete tumor resection, and they did so in six of eight patients. At the last follow-up three patients had no tumor recurrence; the remainder underwent surgery, irradiation, or both.

For the treatment of chordomas, I do not follow the same policy as the authors of this article. I perform an aggressive resection followed by radiotherapy if any tumor remains. The quality and appropriateness of the illustrations provide useful information for the readership.

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