Excision of a Clival and Upper Cervical Pheochromocytoma by an Extended Maxillotomy Approach: A Case Report

Abstract—By use of an extended maxillotomy approach, greater surgical access is provided to the midline skull base in comparison with lateral approaches. We describe this technique used to debulk a tumor extending from the sphenoid sinus across the craniocervical junction to the second cervical vertebrae. (*Skull Base Surgery*, 3(2):87–90, 1993)

Midline skull base tumors are rare and, because of the regional anatomy of the major blood vessels and cranial nerves, are difficult to expose without hazard. We describe an anterior midline approach, a modification of the conventional transoral route, that provided excellent exposure of an extensive pheochromocytoma emanating from the clivus and craniocervical junction.

CASE REPORT

A 40-year-old man with multiple pheochromocytomas since the age of 10 years had undergone numerous intra-abdominal excisions. Three years prior to this presentation, he developed dysphagia, dysphonia, and hemiatrophy of the tongue and was found on investigation to have an upper cervical mass extending into the clivus. At that time, the lesion was considered not to be surgically resectable and he was treated with embolization of the mass and subsequent radiotherapy with significant shrinkage of the mass and some alleviation of his neurologic symptoms.

Six months prior to his current admission, he presented with diplopia as a result of a left VIth nerve palsy. Magnetic resonance imaging (MRI) scans showed that the tumor had increased in size and was deforming the pons. He had another course of radiotherapy with little evidence of any further clinical improvement or radiologic regression (Fig. 1a,b), so he was then referred for a surgical opinion.

On examination, he had a mild left-sided VIth nerve palsy. Pharyngeal sensation was impaired and the gag reflex was absent on the left. He was dysarthric and had left hemiatrophy of the tongue. The left trapezius muscle was somewhat atrophic.

MANAGEMENT

On admission, alpha and beta blockade was instituted using atenolol and phenoxybenzamine to obtain cardiovascular stability. An angiogram was performed and this revealed an extremely vascular tumor, so prior to surgery, embolization of the tumor was performed.

Figure 1. Magnetic resonance image of tumor extending from the sphenoid to the base of C2 of a coronal section (a) and transverse section (b) at the craniocervical junction.

Operation

An elective tracheostomy was fashioned to provide airway control and a pharyngogastrotomy was performed to allow postoperative gastric alimentation.

A standard Le Fort I horizontal mucoperiosteal incision was made above the mucogingival reflection (Fig. 2). The proposed bone cuts were marked out and titanium miniplates (Liebinger, Germany) were fixed into position. Placement of these bone plates prior to bone section enables precise relocation of the maxilla and prevents postoperative malocclusion of the teeth. A midline sagittal section was then made in the labial and palatal mucoperiosteum and two miniplates were fitted across the midline. All the miniplates and screws were then removed and carefully labeled for correct replacement. A standard Le Fort I osteotomy¹ was performed with a reciprocating saw. A midline sagittal split of the hard and soft palate was then made and both components were fractured downward (Fig. 3) and laterally. The inferior turbinates and the vomer were excised and the nasal mucosa was reflected laterally.

The tumor was exceedingly vascular and generally soft. It was debulked with suction and rongeur. During dissection, blood loss of upward of 500 ml/min was encountered on occasion. During these episodes, rapid dissection was carried out until approximately 2 liters were lost, than a "rest" period was instituted to regain hemostasis. In all, blood loss was in excess of 11 liters. Considerable debulking of the tumor from the sphenoid sinus to below the craniocervical junction was achieved.

Hemostasis was difficult in the tumor cavity and so a

Figure 2. The soft and hard palates are divided in the midline.

Figure 3. The hard palate is down-fractured after dividing the vomer. Both parts of the palate are separated laterally to expose the midline structures. Reprinted with permission from [author].⁹

the ends being brought out through the nostrils. Gauze packing to secure hemostasis is infrequently used in neurosurgery, but is common in other branches of surgery. Five days later, attempts were made to remove the gauze pack, but considerable bleeding was encountered. Angiography was performed and this excluded a major vascular injury. Ten days after the initial operation, the pack was removed without significant hemorrhage and the cavity was packed with muscle and Tisseal. The latter is a thrombin fibrin glue used to seal tissues. Three weeks later, the palatal wires from his extended maxillotomy were removed.

Review in the outpatient department revealed the patient's voice to be very much stronger and coherent, his swallow was better, and his double vision had settled. Examination revealed resolution of his left-sided VIth nerve palsy and improvement in his palatal movement, although his XIIth nerve palsy persisted.

Repeat MRI scans showed a considerable reduction in tumor volume (Fig. 4a,b).

DISCUSSION

Tumors of the clivus usually present with lower cranial nerve dysfunction and later neuroaxial compression. Because of the relative inaccessibility of the clivus, various approaches have been advocated to deal with these lesions, each with its inherent risks and drawbacks. A transcervical retropharyngeal approach has been de-

scribed by Stevenson et al.² The problem with this route is the risk of damage to the carotid artery and lower cranial nerves. Other approaches include a transseptal sphenoidal route,³ a transpetrosal route,⁴ and an approach lateral to the orbit.⁵ These approaches are inappropriate for large lesions of the clivus, which extend inferior to the cranio-cervical junction. In the aforementioned case, all would have required several difficult operations and proximal and distal vascular control would have proved difficult.

A transoral approach^{6,7} may be used for high midline cervical lesions with additional palatal splitting incorporated for access to the foramen magnum. However, it provides limited access to the upper third of the clivus; lateral extensions are also limited by the alveolar ridges of the maxilla. A combined transoral, transnasal procedure provides increased access to the clivus, but lateral access remains limited. A combination of maxillotomy⁸ and a transoral approach has been used by us in osteogenesis imperfecta.

With the extended maxillotomy approach involving a transverse osteotomy at the Le Fort I level and a sagittal split of the maxilla and palate, an uninterrupted exposure of the floor of the sphenoid sinus, the clivus, the cranio-cervical junction, and at least the first two cervical vertebrae is achieved. Lateral visibility remains somewhat limited, however.

The major perioperative complication from this method of approach is hemorrhage during mobilization of the maxilla and bleeding from the mucosa and cut surfaces

Figure 4. Postoperative magnetic resonance image showing the considerable reduction in tumor bulk in a coronal section (a) and transverse section (b) at the cranio-cervical junction. (Figure continued on next page)

Figure 4, cont.

during the neurosurgical procedure. In our particular case this was minimized by teamwork involving physicians and radiologists in the preoperative phase and expert anesthesia. Postoperative packing, as illustrated in our case report, may be used as an adjuvant in the control of hemorrhage and did not result in infection.

Late complications, which are rare, include palatal fistulization, dental malocclusion (prevented by preplacement of the bone miniplates), restricted mouth opening due to stiffness of the temporomandibular joints, and very occasionally velopharyngeal incompetence.

CONCLUSION

The extended maxillotomy approach confers good exposure of midline skull base tumors. Although different tumors will require different approaches, the extended maxillotomy offers another approach that may be useful to the skull base surgeon.

REFERENCES

1. Brommer RB: The history of the Le Fort I osteotomy. *J Maxillofac Surg* 14:119-122, 1986
2. Stevenson GC, Stoney RJ, et al: A transcervical approach to the ventral surface of the brain stem for removal of a clivus chordoma. *J Neurosurg* 24:544-551, 1966
3. Bailey P, Bagdasar D: Intracranial chordoblastoma. *Am J Pathol* 5: 439-449, 1929
4. Fisch U: *Microsurgery of the Skull Base*. Stuttgart: Thieme Medical Publishers, 1988
5. Derome P, Caron JP, Hurth M: Indications de la voie trans-bucco-pharyngee et malformations de la charniere craniovertebrale. *Neurochirurgie* 28:282-285, 1977
6. Crockard HA: The transoral approach to the base of the brain and upper cervical cord. *Ann R Coll Surg Engl* 67:321-325, 1985
7. Crockard HA, Bradford R: Transoral transclival removal of a schwannoma anterior to the craniocervical junction. A case report. *J Neurosurg* 62:293-295, 1985
8. Archer DJ, Young S, Uttley D: Basilar aneurysms. A new transclival approach via maxillotomy. *J Neurosurg* 67:54-58, 1987
9. James D, Crockard HA: Surgical access to the base of skull and upper cervical spine by extended maxillotomy. *Neurosurgery* 29: 411-416, 1991