

Skull Base Chondrosarcoma: Is there a Role of Endoscopic Excision?

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Abstract

Skull base chondrosarcomas are rare and account for approximately 0.15% of all intracranial tumors. Management of these tumors is challenging because of their potential to recur, and their proximity to vital structures such as major vessels and cranial nerves. We describe five patients with chondrosarcoma of the skull base who were managed at our institute between January 2002 and December 2009.

Keywords: Chondrosarcoma, skull base, endoscopic, craniotomy.

INTRODUCTION

Chondrosarcomas are a rare group of tumors that arise in any bone that is preformed by cartilage. It accounts for 11 to 19% of all primary bone tumors. Skull base chondrosarcomas usually arise *de novo* and account for approximately 0.15% of all intracranial tumors.¹ Cranial chondrosarcomas are more frequently located at the sphenoccipital fissure. These tumors represent a challenge for surgeons because of their tendency to infiltrate bone, their potential to recur, and their proximity to vital structures such as major vessels and cranial nerves. Thus, negative resection margins around the entire tumor specimen may be difficult to obtain, and even gross total resection often does not result in oncologically complete microscopic resection.^{2,3}

Therapeutic options include surgical removal, radiotherapy, or combination thereof. The rarity of chondrosarcoma restricts surgical experience and conclusive analysis of different treatment strategies. Conventional microsurgical approaches are limited to exposing the lateral and bilateral regions because of surgical trauma. Endoscopic excision provides better visualization of the deeper anatomical structures in the skull base and affords a means to 'look around corners'. It is both a safe and efficient procedure. The prognosis of these tumors was considered to be generally poor,² although the results of the recent largest series indicate 5- and 10-year survival rates of 83 to 99%.^{4,5} In the present article, we report our experience

with management of skull base chondrosarcoma and discuss the role of endoscopic excision in these cases.

MATERIALS AND METHODS

A prospective study was conducted in the Department of Otolaryngology, Postgraduate Institute of Medical Education and Research, Chandigarh, India. The study included five patients with chondrosarcoma of the skull base between January 2002 and December 2009. A detailed history was taken and a thorough clinical examination carried out in all the cases including ophthalmic examination and a neurological examination. Tumor extension and bone destruction were evaluated by performing magnetic resonance imaging (MRI) and/or computed tomography (CT) studies. MRI was performed in the axial, coronal and sagittal planes to help define the relation of the tumor to the surrounding brain, blood vessels and other vital structures.

The patients were taken up for surgery. In three cases, transnasal transsphenoid endoscopic approach was used. In two patients disease was removed by craniotomy open approach by two teams of ENT-head and neck surgeons and neurosurgeons working simultaneously. The patients were followed up by clinical examination and radiologic examination. Follow-up period ranged from 24 months to 5 years. Recurrence of symptoms resulted in revision surgery. Postoperative adjuvant radiotherapy was not administered routinely.

Surgical Technique of Transnasal Transsphenoid Endoscopic Approach

Surgery was performed under general anesthesia with orotracheal intubation. The nasal cavity was decongested by 4% xylocaine and adrenaline wicks. Ethmoidotomy is performed to expose the sella and upper portion of the clivus. The bone of the sella and clivus is drilled to expose the tumor. Complete tumor removal was achieved. Hemostasis was achieved and merocel packing was done.

RESULTS

Between 2002 and 2009, five patients chondrosarcoma of skull base were treated at Postgraduate Institute of Medical Education and Research, Chandigarh, India.

Clinical Features

The patients consisted of three males and two females. The mean age at presentation was 38.9 years with age range of 10 to 55. All the patients reported headache. Visual dysfunction or blindness was present in 4 cases and diplopia in 3 cases. Two patients had diplopia. Nasal obstruction or hyposmia was present in 4 cases. Facial numbness was reported in 1 case.

Radiologic Examination

The computed tomographic (CT) scans performed in five patients showed high-density lesions with calcification. Skull base chondrosarcoma are hyperintense on T2 and hypo- to isointense on T1-weighted sequences, with homogeneous or inhomogeneous contrast uptake. In this study, the tumor involved clivus in 4 cases, nasopharynx in 3 cases, sellar floor in 3 cases, sphenoid sinus in 3 cases, ethmoid sinus in 2 cases, cavernous sinus in 2 cases, orbit in 1 case and infratemporal fossa in 1 case.

Surgery

In three patients, transnasal transsphenoid endoscopic approach was used. In two patients, craniotomy open approach was used (frontotemporal in one and bifrontal in one). There was no major intraoperative and postoperative complication.

Follow-up

Follow-up period ranged from 24 months to 5 years. Three patients (two after endoscopic surgery and one after craniotomy) developed recurrence after mean 3.5 years. In one of the patients, after four years of follow-up radiology showed some doubtful lesion in right posterior ethmoid.

The patient underwent nasal endoscopy which revealed fibrotic tissue in the region of right posterior ethmoids. One patient died after 26 months. The 5 years survival rate for skull base chondrosarcoma observed was 80%.

DISCUSSION

Cranial base chondrosarcomas are rare tumors and are difficult to manage. Complete surgical resection of these tumors is precluded by their location, and the rate of the local recurrence is dependent on the adequacy of the surgical resection.

Chondrosarcomas are thought to originate from primitive mesenchymal cells or from the embryonal portion of the cartilaginous matrix of the cranium. The sites of origin of chondrosarcomas include the clivus, sacrococcygeal region and vertebral column. Cranial chondrosarcomas most commonly originate in the region of the clivus, but are rarely found to be located at the midline.⁶ They are usually paramedian, extending most frequently posteriorly or posterolaterally. Although chondrosarcomas grow slowly and metastasize infrequently, they are histologically malignant and locally aggressive.

Surgical excision of chordomas and chondrosarcomas has been performed using numerous approaches, ranging from the transsphenoidal approach to craniotomy open approaches.⁷ The choice of the surgical approach depends mainly on the extension and especially on the anatomic location of these tumors. Skull base chondrosarcomas are basically midline tumors to which anterior midline approaches are generally preferred which include the transsphenoidal approach, midfacial degloving approach and transoral approach. The anterior midline approaches provide excellent exposure of the sphenoid sinus, sella turcica, and upper and middle clivus. Its disadvantages include limited lateral and inferior exposure, a deep and narrow field and the lack of proximal control of the cavernous carotid artery.⁸

Lateral approaches through the petrous bone provide a relatively direct access to the clivus but are not adequate to expose the region lateral to the upper clivus and bilateral extensions of clival chondrosarcoma into the parasellar region cannot be managed by a unilateral approach.

The endoscopic transsphenoidal approach has been reported in the literature as a useful tool to treat sellar and parasellar lesions.⁹ Chordomas and chondrosarcomas usually are good candidates for endoscopic approaches because of the extradural origin and their predominantly extradural growth, the soft tissue of the tumors, and the minor bleeding.¹⁰ An endoscopic endonasal approach can expose a region from the sellar floor to the foramen magnum in the vertical dimension and between the mandibular joint

areas in the transverse dimension.¹¹ The lateral exposure can be made up to the ovale foramen and rotundum foramen. Therefore, this approach can be considered appropriate for skull base tumors. The endoscope permits a panoramic view instead of the narrow microscopic view, and it allows the inspection of sellar, parasellar, and suprasellar compartments by means of angled lens endoscopes.¹² Moreover, the use of devices such as the microdebrider, the microvascular ultrasound Doppler probe, and computer assisted navigation have improved safety in the resection of tumors using the endoscopic transsphenoidal approach.¹³

Frank et al¹⁴ have described an endoscopic transnasal ethmo-pterygoid-sphenoidal approach to treat cranial base chordomas and chondrosarcomas. This approach provides a more lateral view of the cavernous sinus. It achieves the creation of a surgical route involving less risk: A medial route between the pituitary gland and the ICA, and the lateral route between the ICA and the IIIrd, IVth, V1, and V2 nerves protected by a periosteal and dural layer.

The main limits of the endoscopic technique are represented by extensive dural invasion of the tumor and the tumor being primarily centered on the inferior clivus and extended to the occipital condyle. These latter conditions are better managed by craniotomy open approaches. The future will show whether endoscopic techniques provide the same results as shown by classic cranial base approaches but there are studies which report that the morbidity of transsphenoidal approaches is very low with a high-rate of improvement of symptoms.⁸

Long-term survival with good quality of life may require repeated surgeries or radiotherapy in case of tumor recurrence or regrowth. The use of radiotherapy is indicated for surgically inaccessible, postoperative residual or recurrent tumors.¹⁵

Different forms of radiotherapy claim to achieve high rates of local tumor control and survival like proton-beam radiation therapy,¹⁶ fractionated proton-beam radiation therapy,¹⁷ helium ion treatment¹⁸ and stereotactic radiosurgery.¹⁹ For the most efficient use of these therapeutic modalities, the tumor should be as small as possible, and these modes of therapy should be reserved for a residual tumor after the surgical resection of as much volume as possible.

CONCLUSION

The advantageous use of nasoendoscopes is not limited only to the minimally invasive surgery. They also offer better visualization of the deeper anatomical structures in the skull

base, provide the wide exposure of the field and provide a rapid access to the clival and parasellar region. The main limits of the endoscopic technique are represented by extensive dural extension of the tumor and the tumor being primarily centered on the inferior. The endoscopic approach is also repeatable in cases of recurrence or in cases of residual tumors.

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