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Petrous Chondrosarcoma: Case Study of Two Patients with Surgical Nuances and Literature Review

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Abstract

Primary chondrosarcoma of the skull base is a malignant tumor made up of cells developed from aberrant chondrocytes that occurs in 0.1 to 0.2% of all skull base lesions. Skull base lesions are challenging not just because of their location, but also because critical local neurovascular structures are involved. Thus, we will examine two cases with varying presentations and study the literature to evaluate various surgical management options and their nuances.

Keywords
- petrous bone
- chondrosarcoma
- surgery

Introduction

Chondrosarcoma of the skull base is an abnormally malignant mesenchymal tumor formed of cells produced from altered chondrocytes with a coverage of 0.1 to 0.2%.[1] Slow development, destruction of cranial fossae, and encasement of local neurovascular structures are all characteristics of this type of tumor. High-grade lesions, on the other hand, suggest rapid development and early metastasis. It is distinguished by a wide variety of clinical symptoms and morphological characteristics that vary according to the involvement of bone, neural, and vascular structures. Chondrosarcoma affects the skeletal system as well, most commonly the pelvis, upper part of humerus, or upper metaphysical and diaphyseal portions of the appendicular skeleton.

We are going to discuss two cases of petrous chondrosarcoma with multicompartmental and posterior petrous involvement in this case report. In addition, we performed a literature review to explore the many surgical nuances involved in its surgical management.

Case 1

A 22-year-old female presented with complaint of subtle weakness in left upper and lower limb with left eye axial proptosis with right-sided facial palsy for 10 to 15 days. Gadolinium-enhanced magnetic resonance imaging (MRI) brain showed extra-axial lobulated mass with T1-weighted (T1W) hypo, T2W hyper with soap bubble appearance on gadolinium enhancement causing expansion of the left Meckel's cave and displacing the left temporal lobe and extending posteriorly with infiltration of petrous causing its erosion and extending in the perimesencephalic cistern with shifting of brain stem posteriorly and toward right side (►**Fig. 1A**). Preoperative diagnosis of left trigeminal schwannoma was made. Patient underwent left retro mastoid suboccipital craniectomy and subtotal decompression with involvement of posterior fossa removed (►**Fig. 1B**). After 3 months, patient was operated via extradural subtemporal approach and excision of the tumor involving the cavernous sinus removal till petrous apex; near total excision was done (►**Fig. 1C**). Tumor was soft, suckable, grayish color with gelatinous consistency. Soft part was removed and hard part adhered to petrous bone was left.

Postoperatively, patient had no new neurological deficits. Patient developed wound infection and meningitis, for which she was managed conservatively. Patient condition gradually improved. Histopathology was suggestive of chondrosarcoma grade I. The microscopic features were of a low-grade tumor without mitosis or necrosis as shown in microphotograph (►**Fig. 1D**).

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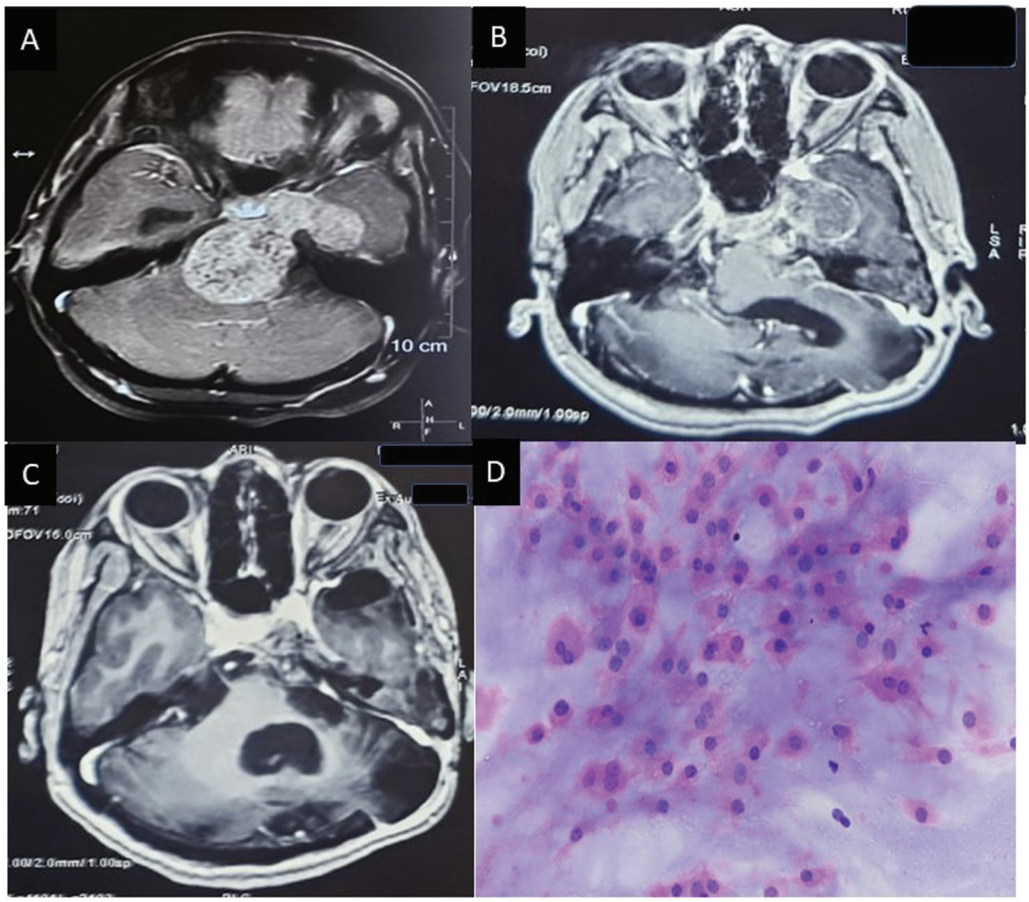

Fig. 1 Preoperative magnetic resonance imaging (MRIs) of left anterior petrous lesion (**A**) showing large dumbbell-shaped mass lesion at left prepontine cistern extending across left Meckel's cave bulging into left cavernous sinus and adjacent medial temporal lobe. MRIs after first surgery (**B**) showing excision of prepontine part with residual lesion in left cavernous and temporal lobe with postoperative changes. MRIs after second surgery (**C**) showing small residual lesion over the left petrous apex. (**D**) Microscopic photograph—Formalin-Fixed Paraffin-Embedded (FFPE) section showing classic histomorphology of an infiltrative chondrosarcoma low-grade tumor that appears as a bluish matrix lesion amid bone and fibrous tissue with monotonous round to oval cells with bland round nuclei and abundant eosinophilic cytoplasm, few binucleated forms, in the bluish chondroid matrix background, hematoxylin and eosin, $4 \times 10 = 40X$.

Case 2

A 46-year-old female presented with complaints of tinnitus in right ear and facial weakness of right side of face for the last 1 month. On examination, there was right-sided facial palsy, House–Brackmann grade II. Mild sensorineural hearing loss found in right ear. Rest cranial nerve examination was normal. Motor and cerebellar examination was normal. Computed tomography scan showed a heterogenous extra-axial soft tissue lesion with multiple peripheral and central calcific foci in right posterior fossa, centered toward right petrous bone, having approximately $3 \times 3 \times 2.9$ cm size. Lesion causing infiltrative erosion of retrolabyrinthine part of right petrous bone and posteromedial mastoid part of right temporal and jugular foramen was found (►**Fig. 2A**). Gadolinium-enhanced MRI brain was suggestive of well-defined mild expansile lytic lesion at mastoid portion of right temporal bone. Lesion is abutting and displacing the mastoid segment of facial nerve anteriorly. Heterogenous hyperintense on T2W imaging and intermediate signal T1W imaging. Heterogenous mild-to-moderate enhancement was seen. Lesion was also compressing the sigmoid venous sinus and partially obliterating the jugular foramina (►**Fig. 2B**). Differential diagnosis was kept as paraganglioma, chondrosarcoma, and aneurysmal bone cyst. Right retromastoid suboccipital craniotomy and gross total excision of tumor was done. Tumor was extradural arising from right posterior petrous region. Tumor was bony in consistency with intermittent soft to firm in consistency with mild vascularity. Impression-Chondrosarcoma (►**Fig. 2C**). Postoperatively same lower motor neuron type facial palsy grade II was found.

Postoperatively, patient was discharged in satisfactory condition with no new neurological deficit.

Discussion

Chondrosarcomas are malignant tumors originating from chondrocytes that are infrequently linked to Ollier disease, Maffucci syndrome, or Paget disease.[2] According to Rosenberg et al,[3] the temporo-occipital region (66%) is the most frequent site of skull base chondrosarcoma, followed by clivus (28%) and sphenoethmoid complex (6%). They are extradural space occupying lesions with a jelly like consistency that damage bony structures.[4] Focal pain and edema are frequent complaints. Symptoms are produced by the enlargement of skull bones,

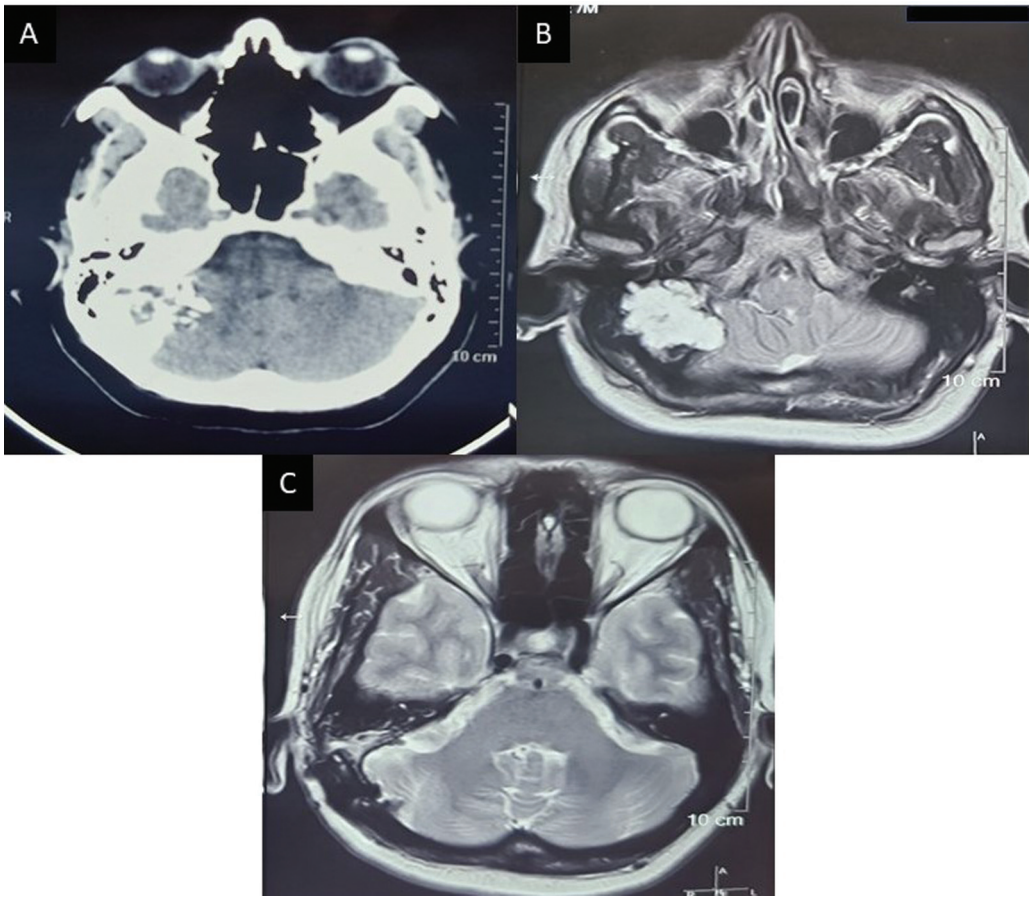

Fig. 2 Preoperative computed tomography (CT) and magnetic resonance imaging (MRI) of right posterior petrous lesion (**A, B**). (**A**) Preoperative CT scan showing conglomerate calcification with erosion of right petrous bone. (**B**) T2-weighted axial MRI showing T2 hyperintense well-defined lesion of $3 \times 3 \times 2.5$ cm size. (**C**) Postoperative MRI scan showing total removal of lesion with post operative changes.

pressure effect, or involvement of the surrounding brain, cranial nerves, and paranasal sinus.[5] Petrous chondrosarcomas can involve the middle and posterior cranial fossa. Petrous apical lesions affecting the middle cranial fossa and spreading medially can result in pituitary symptoms, cavernous symptoms, and temporal lobe dysfunction. This was seen in patient 1. When involving the posterior fossa in midline, unilateral/bilateral cranial nerve palsies occur, and laterally, cerebellopontine angle syndrome (seen in patient 2), jugular foramen, and/or foramen magnum syndrome occur.[5] On radiology, they are isointense to hypointense on T1 and hyperintense on T2, with low T2 signal foci matching to calcified chondroid matrix.

Histologically, they are of four subtypes: classical, mesenchymal, clear cell, and dedifferentiated. Classical subtype is predominantly found in the cranial region. The World Health Organization classification of head tumors classified classical chondrosarcomas into different grades depending upon the mitotic activity and cellular and nuclear polymorphism.[2,6]

1. Grade I: Atypical chondromatous tumor: most common type, is found in approximately 80% cases. Microscopic features are similar to benign cartilaginous tumor like chondroma. Mitotic activity is less than or equal to 2 mitotic figures / 10 HPF.
2. Grade II: It is found in approximately 17.8% cases, less common in cranium. Extracellular matrix myxoid alterations with significant nuclear polymorphism. Mitotic figures more than or equal to 2 mitotic figures/10 HPF.
3. Grade III: It is found in approximately 1.8% cases. It exhibits high mitotic activity due to extensive cellular and nuclear polymorphism.

Surgical Nuances

Preoperative examination includes determining the exact location of origin, direction and local dissemination of the tumor, amount of dural and skull base bone involvement, tumor vascularity and consistency, and the arachnoid plane between tumor and the brain.

Depending on the extent of the lesion, skull base approaches are split into midline and lateral. Sellar/suprasellar and clival lesions form midline lesions.[5] Temporal base and infratemporal fossa lesions, as well as lateral petroclival lesions, form lateral lesions. Lesions of the petrous bone usually affect the middle and posterior skull bases; midline lesions can be managed endoscopically, whereas lateral/ posterior skull base lesions are managed laterally.

Extradural subtemporal approach is the most often used lateral skull base approach for lesions of the middle fossa and

cavernous sinus. Petrous ridge obstructs the entry to the petroclival region through the subtemporal route. In such case, anterior transpetrous approaches are combined with subtemporal approaches to improve exposure of lesions extending to the cerebellopontine angle and the petroclival region.

To access lesions involving the cerebellopontine angle region and posterior surface of temporal bone, posterior approaches are used, which include far lateral or extreme lateral approaches, and retromastoid suboccipital craniotomy. Lesions around the jugular foramen are often treated with a transjugular approach with or without petrosectomy.

Endoscopic Surgery

Endoscopic surgery is minimally invasive approach for biopsy and lesion removal. Endoscopes allow access to lesions in the skull base with minimal brain and neurovascular manipulation.[7,8] Lack of binocular vision makes it difficult to have three-dimensional vision, increases the chances of cerebrospinal fluid leak, even makes it difficult to manage bleeding in case of a hemorrhagic complication.

Neuronavigation

Bony landmarks are altered and/or obliterated in skull base lesions, thus surgery becomes difficult owing to anatomical orientation loss. In such scenario, neuronavigation becomes a useful guiding tool.[9,10] It was used in our second case for achieving complete excision.

Radiotherapy

Goal of surgery consists of maximum safe resection, supplemented by radiation therapy. Histology and relieving pressure over cranial nerves were main targets of surgery. Radiotherapy is recommended as second line of postoperative management.[4,11] According to Bloch and Parsa, 5-year mortality rate is 25% in only surgery group and 9% in surgery followed by radiotherapy group.[1] However, it has also some risks. Risks include damage to cranial nerves, brainstem, major vessels, pituitary gland, and other neural structures.

As they are very slow-growing tumors, they respond well to radiosurgery, and we believe that partial excision followed by radiation exposure is the best technique for avoiding neurological deficits.

Chemotherapy had not shown any significant advantages.[2,12] It is used in some cases with grade III lesions as last option, in which patients had shown no response to combined treatment.[4]

Conclusion

Surgical approach, grade of resection, aiming for maximal tumor resection with functional preservation must be adapted specifically for each patient, taking into account the surgical team expertise and preferences. Best approach consists of maximum safe resection followed by radiotherapy based on histological pattern, with focus on patient's quality of life and neurological function preservation. Combining neuronavigation with microsurgical and endoscopic procedures will help to improve surgical results and decrease morbidity.

Authors' Contribution
All the authors prepared and reviewed the manuscript. A.S. and M.J. reviewed the literature. V.S. examined the surgical specimen for pathology.

Conflict of Interest
None declared.

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