Primary Osteosarcoma of the Skull Base Treated with Endoscopic Endonasal Approach: A Case Report and Literature Review

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Abstract

Introduction Giant cell-rich osteosarcoma (GCRO) is a rare pathologic diagnosis, and most cases have involved the appendicular skeleton. We present a challenging diagnosis of GCRO of the skull base treated with an endoscopic endonasal approach.

Case Presentation An 18-year-old female patient presented with acute monocular visual loss. Imaging revealed a large clival mass encasing the internal carotid arteries bilaterally with pituitary and optic nerve compression. The lesion was resected via a staged endoscopic endonasal approach and the patient's vision normalized postoperatively. The final pathological diagnosis was challenging and, after consultation with multiple North American centers, was concluded as GCRO. The tumor recurred and further surgery was performed, followed by adjuvant chemoradiation.

Conclusion We highlight diagnostic challenges of GCRO of the skull base, and describe, with intraoperative pictures, successful surgical resection via an endoscopic endonasal approach. Based on our literature review, this is the first published case report of GCRO of the skull base.

Keywords
- giant-cell rich osteosarcoma
- skull base
- endoscopic endonasal approach
- clivus

Introduction

Primary osteosarcoma of the head and neck represents 6 to 13% of osteosarcoma cases,[1] and 1.7 to 5% of head and neck cancers.[2] Males and females are equally affected, and patients usually present in the third and fourth decades of life. Contributing factors include prior chemotherapy, radiation, and underlying pathological conditions such as fibrous dysplasia and Paget disease.[2]

Osteosarcoma is subdivided based on histopathologic characteristics. These subtypes include conventional, telangiectatic, small cell, epithelioid, osteoblastoma-like, chondroblastoma-like, fibrohistiocytic, and giant cell-rich (GCRO). GCRO, first described by Bathurst et al, is a rare variant accounting for 1 to 3% of all osteosarcoma cases.[3] Most cases have been reported in the appendicular skeleton with two cases arising in the maxilla and the mandible.[4,5] Due to overlapping histopathological features, this subtype can be difficult to differentiate from other pathological entities including GCR variety of malignant fibrous histiocytoma, chondrosarcoma, and giant cell tumor. Because of the different prognostic features and management strategies of these pathologies, it is imperative to make the correct diagnosis in a timely fashion.

To date, there have been no published cases of primary GCRO of the skull base. We report a case of GCRO arising

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from the clivus and describe its radiologic and histopathologic characteristics, as well as our surgical treatment through an endoscopic endonasal approach. We will also discuss some of the challenges in establishing the pathologic diagnosis.

Case Description

An 18-year-old female patient presented with a 3-day history of progressive decreased left visual acuity. She also reported worsening headaches over the preceding 4 to 5 months. She denied any symptoms suggestive of endocrinopathies or increased intracranial pressure. Her medical and family histories were noncontributory.

Physical examination revealed a marked absence of left visual acuity. Imaging revealed a large enhancing and expansile central skull base mass involving the upper two-thirds of the clivus and eroding the dorsum sella (►**Fig. 1**). Both the pituitary gland and the chiasm were displaced superiorly. There was possible erosion of the medial aspect of the carotid sulci bilaterally at the level of the paraclival internal carotid arteries (ICAs). The lesion was also extending to the paraclival, parasellar, and paraclinoid ICAs bilaterally. The radiological differential diagnosis included chordoma, chondrosarcoma, interosseous meningioma, and osteosarcoma.

The patient underwent a planned two-stage endoscopic endonasal approach to obtain tissue diagnosis and decompress the optic apparatus. Briefly, we raised a right standard nasoseptal flap[6] and performed a posterior septotomy, bilateral maxillary antrostomies, ethmoidectomies, and sphenoidotomies for access and to widen the nasal corridor (►**Fig. 2**). The floor of the sphenoid sinus was drilled to the level of the clivus. A left-sided transpterygoid approach was undertaken.[6] The vidian canal was identified and delineated to the level of the petrous ICA (►**Fig. 3**). This step was essential to allow for proximal control of the vessel on that side. We elected not to perform the same maneuver on the right side to avoid compromising the vascular supply to the nasoseptal flap. Tumor debulking was initiated centrally and progressed laterally on the left and then right side in a counter-clock wise fashion. The right medial optic carotid recess, lateral optic carotid recess, and right paraclinoid ICA were identified. Following the plane between the tumor and the right paraclinoid ICA proximally allowed for complete detachment of the tumor from the dura of the medial compartment of the cavernous sinus and the paraclival ICA. The micro Doppler probe was used frequently throughout the process to identify and gauge proximity to the ICA. The tumor was well encapsulated in most areas and elevated well off the lateral recesses of the sphenoid sinus and the planum sphenoidale. However, it had clearly eroded the middle third of the clivus at the posterior wall of the paraclival recess as well as the floor and anterior wall of the sella turcica. Because of the fibrosity of the tumor and its large size, an automated side suction-cutter aspirator (Myriad) (NICO Corp., Indianapolis, Indiana, United States) was used for a significant part of the debulking process. ►**Fig. 4** illustrates the final reconstructed surgical defect. An intraoperative frozen biopsy suggested a benign process. Although there was dural tumor involvement, the surgeons opted not to resect the dura because of the benign intraoperative pathology report.

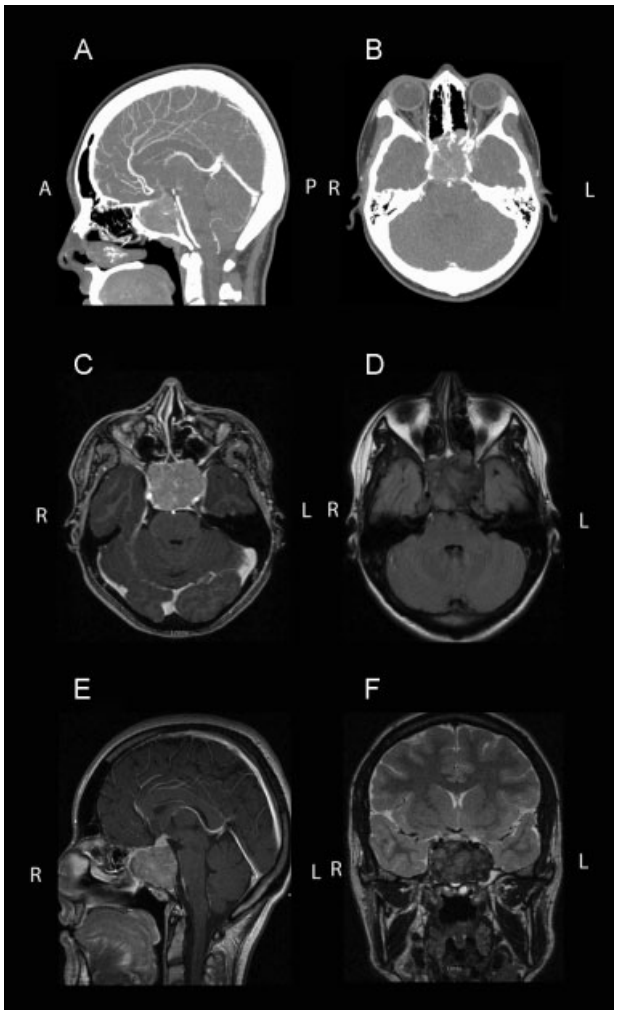

Fig. 1 Computed tomography angiogram scans with bone window sagittal (A) and axial (B) showing large central skull base lesion with mass effect over the internal carotid arteries and basilar artery. There is erosion of the floor of the sella turcica with extension into the sphenoid sinus. The mass caused cranial displacement of the pituitary stalk. (C) Axial enhanced magnetic resonance imaging disclosing invasive expansile mass in the central skull base with signal intensity most consistent with partial osteoid matrix. (D–F) Magnetic resonance imaging axial (D), sagittal (E), and coronal (F) disclosing large central skull base mass with mass effect on internal carotid arteries and encasing the left carotid artery.

Immediately, postoperatively, the patient's vision improved and then normalized by postoperative day 7. There were no intraoperative complications. A postoperative magnetic resonance imaging (MRI) showed more than 98% tumor resection, with a small remnant at the dura (►**Fig. 5**).

Postoperatively, the initial pathological diagnosis was "giant cell lesion," and the differential diagnoses included giant cell tumor and GCRO. The final description found that

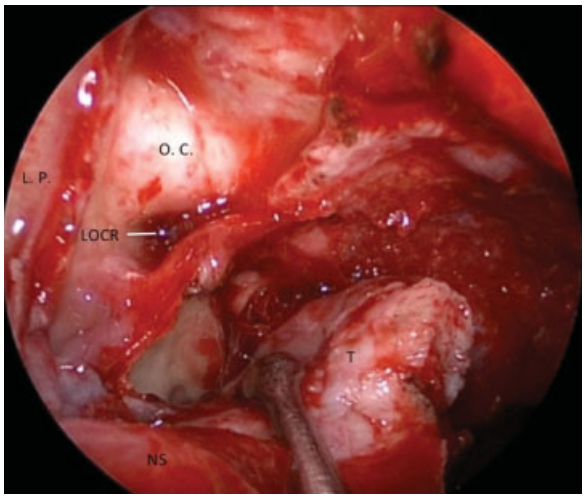

Fig. 2 Intraoperative endoscopic view of tumor resection from the posterior sphenoid sinus. The nasoseptal flap is tucked into the created right maxillary antrostomy. LOCR, lateral opticocarotid recess; L.P., lamina papyracea; O.C., right optic nerve; T, tumor.

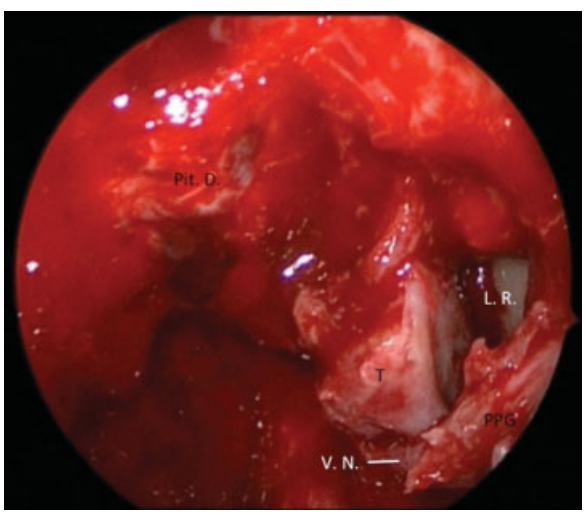

Fig. 3 View of the left posterior nasal corridor. The vidian nerve was identified and traced as a landmark for the carotid artery. L.R., lateral recess of the sphenoid sinus; Pit. D., pituitary gland; PPG, pterygopalatine fossa; T, tumor; V.N., left vidian nerve.

the tumor was composed of atypical epithelioid and spindle cells within fibrous tissue. The presence of osteoid matrix with focal mineralization, consistent with osteosarcoma, was identified. Nuclear pleomorphism and occasional mitoses were also noted (►**Fig. 6**). Eventually, after consultation with several North American pathologists, the final diagnosis of GCRO was confirmed.

After 3 months of surgery at the same time the pathological diagnosis was being finalized, there was evidence of tumor recurrence on a follow-up MRI (►**Fig. 7**). A metastatic workup was negative. Further surgical resection was performed to reduce tumor burden in preparation for adjuvant chemoradiation.

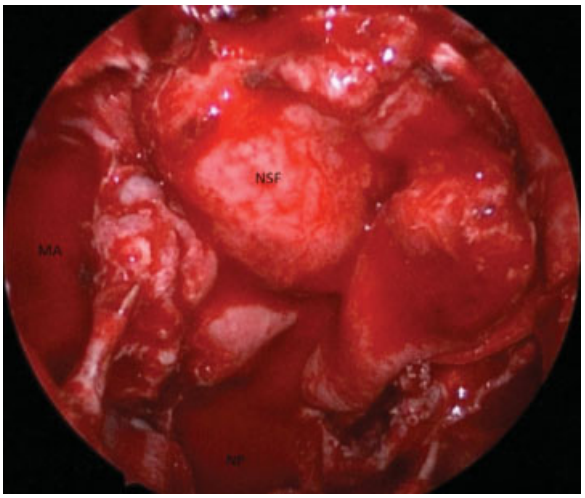

Fig. 4 Final view of the reconstructed surgical defect with the pedicled nasoseptal flap in place. MA, right maxillary antrostomy; NP, nasopharynx; NSF, nasoseptal flap.

Literature Review

We searched the Medline database through PubMed and Ovid interface using the terms “giant cell rich osteosarcoma” and “head and neck.” The resulting articles written in English were reviewed. This review yielded two case reports. One article presented a case of GCRO of maxilla and the other presented a case of GCRO of the mandible.[4,5] A retrospective review of 27 cases of osteosarcoma arising in the head and neck by Ha et al reported one case of GCRO of left posterior occiput.[7] There were no cases of GCRO of skull base.

Discussion

Primary osteosarcoma of the cranial base is a rare and challenging condition with fewer than 150 cases reported in the literature since 1945.[8] Clinical findings may include proptosis, cranial nerve palsies, visual impairment, and headaches.[2] The patient reported in this study presented with a decrease in visual acuity and chronic headaches.

The majority of osteosarcoma subtypes are high-grade conventional, which include osteoblastic, chondroblastic, and fibroblastic subtypes.[1] GCRO is an extremely rare morphological variant of high-grade conventional osteosarcoma. To our knowledge, this is the first case of GCRO of the skull base.

The differential diagnosis of giant cell lesions can present a diagnostic challenge, especially if they occur in an unusual location. In this case, it was difficult to differentiate between a GCRO and giant cell tumor. This case was sent out to pathologists in other North American centers, and the diagnosis of GCRO was eventually confirmed. An osteosarcoma may contain so many giant cells, and only focal atypia, that the malignant elements in the background are obscured, and the lesion is mistaken for a giant cell tumor.[9] Also, the presence of osteoid could be quite rare in an obviously malignant lesion. Malignant giant cell tumor is a controversial

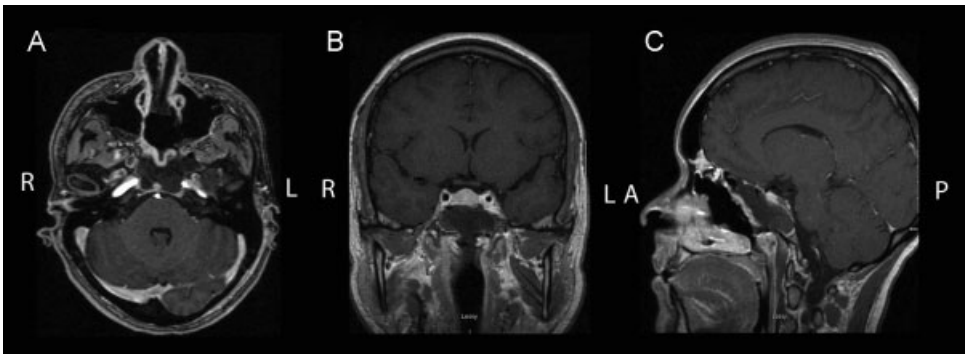

Fig. 5 Magnetic resonance imaging axial (A), coronal (B), and sagittal (C) showing significant reduction in size of lesion. There is decreased mass effect over the internal carotid arteries. The pituitary stalk is midline and there is no compression of the optic chiasm.

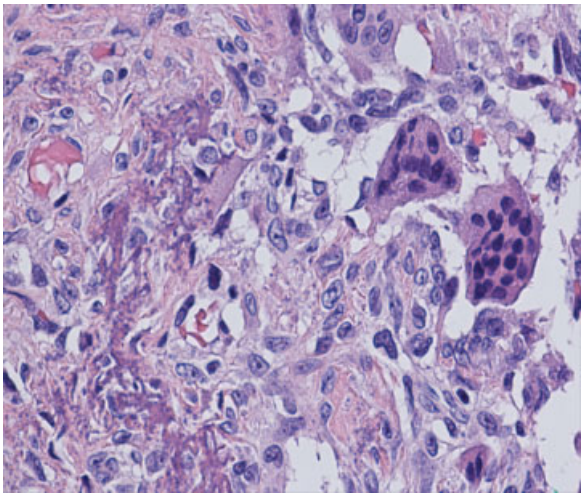

Fig. 6 Biopsy of lesion composed of atypical epithelioid and spindle cells within fibrous tissue and osteoid matrix which is focally mineralized. Nuclear pleomorphism and occasional mitoses are also noted.

entity that should be ruled out by a careful examination for osteoids, important for the diagnosis of GCRO.[1]

Contributing to the diagnostic challenges, the radiological features of GCRO differ from that of conventional osteosarcomas. Bathurst and Sanerkin described a typical radiographic pattern in long bones: an ill-defined margin surrounding a predominantly lytic lesion of the diaphysis or metaphysis. A soft tissue mass is usually not present and the periosteal reaction is weak.[10] In the reported case of GCRO of the maxilla, a contrast-enhanced CT scan showed an expansile heterogeneous mass with areas of speckled calcification within the tumor and adjacent sclerosis.[4] In the reported case of GCRO of the mandible, the radiographic appearance showed an osteolytic lesion.[5] Radiographic changes in conventional osteosarcoma typically show rapid invasion and destruction of metaphyseal area of the long bone with periosteal reaction and soft tissue extension.[3]

Osteosarcoma of skull base is rare and evidence for its treatment is consequently sparse. Although, there is a lack of consensus on the optimal treatment, surgical resection is the mainstay of treatment.[11] The most important prognostic factor is the feasibility of complete tumor resection with negative margins.[11] Our patient presented with an extensive but well-encapsulated tumor of the skull base, almost completely encasing both carotid arteries medially. With advances in endoscopic surgical techniques, image-guidance, Doppler ultrasound, and the expertise of the skull base team, we successfully performed surgical resection of all gross tumor, with minimal morbidity.

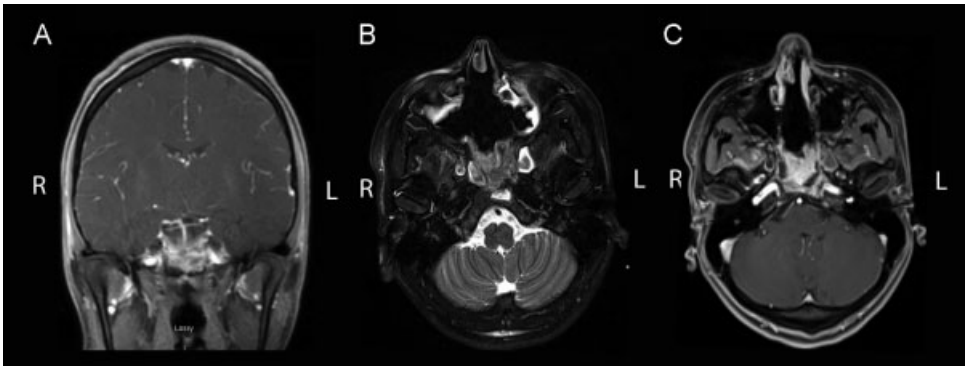

Fig. 7 Enhanced magnetic resonance imaging coronal (A) and axial (C) disclosing recurrence of the clival mass. Solid enhancing component displacing the pituitary gland to the overlying optic chiasm. (B) Axial magnetic resonance imaging disclosing marked interval expansion of the mass. The cavernous sinuses are displaced laterally. The lesion lies adjacent to bilateral internal carotid arteries without apparent encasement.

The tumor site has a significant impact on survival, and extragnathic tumors are usually associated with worse prognosis due to incomplete resection owing to the complex anatomy of that region. The anatomy of the head and neck and especially skull base is complex and complete resection without causing morbidity may not be feasible. In addition, surgeons may be reluctant to undertake complete resections that affect function in younger patients.[12] In osteosarcoma of long bones, the addition of adjuvant chemotherapy and radiation has resulted in improved survival. However, its use in head and neck osteosarcoma has not been established.[12] In a study by Guadagnolo et al, the role of radiotherapy with respect to outcome in head and neck osteosarcoma was analyzed. The authors found that radiotherapy in addition to surgery improved the overall and disease-specific survival in cases where resection margins were positive or uncertain.[13] In the case of GCRO of the maxilla, the patient underwent resection of the lesion followed by 60 Gy radiotherapy with five cycles of chemotherapy. She was disease free at her last follow-up.[4] GCRO of the mandible was treated with surgical resection followed by chemoradiation and patient was disease free at her 1-year follow-up.[5] Local recurrence is the main cause of fatality in head and neck osteosarcoma and distant metastasis is only seen in 7 to 17% of cases.[2]

Conclusion

This report details a patient presenting with a rare tumor, GCRO, in a difficult anatomic location, the skull base involving both carotid arteries, treated with surgical resection using expanded endonasal approach followed by chemoradiation. Our experience adds to the very few similar cases in the literature, and should help other surgeons who are presented with similarly challenging cases.

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