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Case Report

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Unusual giant cell tumor of the clivus: a case documentation and analysis

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ABSTRACT

Giant cell tumors (GCTs) primarily occur in the epiphyses of long bones and are generally considered histologically benign. However, they can be locally aggressive, with a high rate of local recurrence, and have the potential for distant metastasis. Primary GCT of the clivus is extremely rare and is often misdiagnosed as pituitary macroadenoma or clival chordoma, making treatment controversial. The present report describes the case of a 17-year-old male with GCT located in the skull base originating from the clivus, with the involvement of adjacent cranial nerves, which was successfully treated with trans-nasal trans-sphenoidal surgery. This report contributes to the limited literature regarding GCTs of the skull.

Keywords: Cranium, Giant cell tumor, Clival mass, Skull base mass

INTRODUCTION

Giant cell tumors (GCTs), also known as osteoclastomas of the skull, are rare, benign yet locally aggressive tumors originating from bone tissue. These tumors feature multinucleated giant cells amidst a backdrop of mononuclear stromal cells. While GCTs typically arise in the epiphyses of long bones, especially around the knee, they can occasionally affect the skull, accounting for less than 1% of all bone GCTs, primarily involving the sphenoid and temporal bones in the middle of the cranial fossa

Most studies on skull GCTs are case reports, with the tumors found in bones such as the temporal, petrosal, sphenoid, and occipital bones. Primary GCTs of the clivus are exceptionally rare. In radiology, the imaging characteristics of skull GCTs are typically influenced by the tumor's location and extent. These tumors almost always occur after the growth plate has closed, usually appearing in early adulthood. About 80% of cases are reported in individuals aged 20 to 50, with a peak

incidence between ages 20 and 30.^{5,6} This study reports a case of a GCT in the clivus in a 17-year-old male, who presented with headache, double vision, and drooping eyelid. He was successfully treated with minimally invasive surgery. The study also reviews the literature on the diagnosis and treatment of such tumors. Written informed consent was obtained from the patient's family.

CASE REPORT

A 17-year-old male presented with a 15-day history of frontal headache and left eyelid drooping, with no underlying comorbidities. He had previously experienced double vision on the left side 1.5 months earlier, which improved with medication after being diagnosed by an ophthalmologist with left lateral rectus palsy and ptosis.

The ophthalmological examination showed diplopia due to left abducens nerve (CN VI) palsy, but his vision, visual fields, and corneal reflexes were normal, and there was no papilledema. The ophthalmologist recommended an MRI and a consultation with a neurologist. The neurological

examination, including cerebellar tests, was normal, with the patient fully cooperative and oriented. The hormone profile indicated elevated prolactin levels and decreased cortisol levels. A contrast MRI of the brain revealed a well-defined, extra-axial, lobulated mass lesion measuring approximately 3.1×3.6×3.2 cm (AP×TR×CC), located in the clivus and sellar region, with the clivus as its epicenter.

The mass extended to the suprasellar and bilateral parasellar regions, encasing the cavernous and clinoid segments of the internal carotid arteries on both sides. The pituitary gland was not identified separately from the lesion.

The lesion abutted the optic chiasma superiorly and the basilar artery and P1 segment of the left posterior cerebral artery posteriorly, extending laterally to the medial temporal lobe without causing significant mass effect in underlying brain parenchyma.

The tumor tissue appeared isointense on T1-weighted imaging (WI), T2WI, and fluid-attenuated inversion recovery (FLAIR), with moderate heterogeneous enhancement on the post-contrast scan and areas of GRE blooming indicating calcification (Figure 1, 2, 3).

A CT scan further delineated the involvement of the skull base, showing erosion of the clivus, anterior and posterior clinoid processes, and sella turcica, with rarefaction of the posterior wall of both sphenoid sinuses. No definitive extension into the sphenoid sinuses was found (Figure 4). Based on the symptoms and MRI findings, a pituitary macroadenoma was considered the most likely diagnosis, although the possibility of a chordoma or another clival tumor could not be excluded.

The tumor was removed using an endoscopic transnasal transsphenoidal approach under general anaesthesia. Intraoperative findings revealed a grey, soft, friable, hypervascular mass arising from the clivus. The pituitary gland was seen bulging separately from the lesion. Consequently, near-total resection of the tumor was performed along with the floor of the sella turcica.

Postoperative histopathology diagnosed the patient with a giant cell tumor. The histopathology revealed the tumor was composed of benign oval to polyhedral mononuclear cells mixed with numerous evenly distributed osteoclast-like giant cells with more than 15 nuclei. There was evidence of woven bone formation within the tumor. No evidence of sarcomatous transformation, haemorrhagic lakes with clustering of giant cells or pituitary tissue was identified (Figure 6).

Postoperative MRI revealed a residual lesion involving the left cavernous sinus (Figure 5). The patient was asymptomatic and discharged after a few days. He is currently on regular follow-ups and is scheduled for adjuvant radiotherapy.

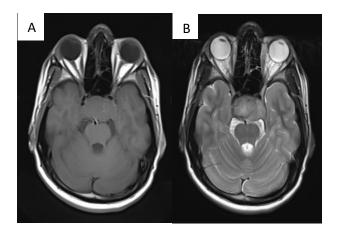


Figure 1: Axial section of T1(A) & T2 (B) weighted magnetic resonance imaging revealing a well-defined, extra-axial, lobulated mass lesion originating from the clivus and surrounding both cavernous sinuses in the sphenoid sinus area of the middle fossa.

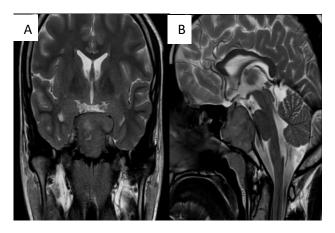


Figure 2: Coronal (A) and sagittal (B) section of T2 - weighted magnetic resonance imaging revealing a lobulated mass lesion originating from the clivus abutting optic chiasm and basilar artery and surrounding both cavernous sinuses.

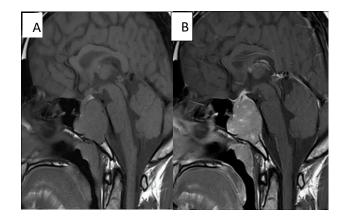


Figure 3: Sagittal section of fat suppressed T1-weighted magnetic resonance imaging (A) with post contrast study (B) revealing a significant post contrast enhancement of the mass lesion of clivus.

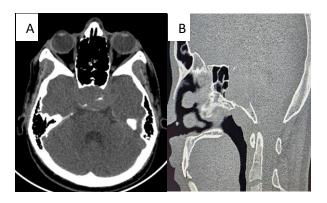


Figure 4: axial section of soft tissue window of CT scan (A) and sagittal section of bone window of CT scan (B) revealing few areas of punctate calcification in mass lesion, erosion of clivus and widening with erosion of Sella turcica.

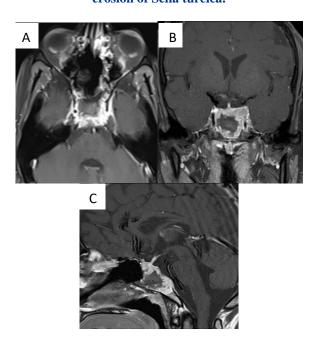


Figure 5: (A) Axial, (B) coronal and (C) sagittal section of postoperative T1-weighted magnetic resonance imaging with contrast revealing residual lesion involving the left cavernous sinus and normal enhancing pituitary gland.

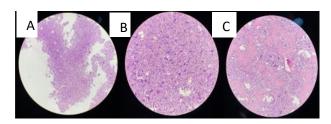


Figure 6: (A, B and C) Histopathological examination revealed that the tumor was composed of benign oval to polyhedral mononuclear cells mixed with numerous evenly distributed osteoclast-like giant cells with more than 15 nuclei. (stain, hematoxylin and eosin magnification, A scanner, B X40, C X40).

DISCUSSION

Giant cell tumor of the clivus is an exceptionally rare condition, with fewer than 40 cases documented in medical literature. These tumors usually develop in long bones, making their presence in the clivus quite unusual. Wolfe et al. first reported a surgically treated clival GCT in 1983. Major symptoms often include headaches due to increased intracranial pressure and diplopia caused by abducens nerve palsy. These tumors typically occur in the second and third decades of life and are slightly more common in women. Symptoms can include headache, visual field defects, blindness, and diplopia. Surgical treatment of these tumors at the skull base is challenging, making adjuvant therapy important. GCTs are locally aggressive, with a high local recurrence rate and a low potential for distant metastasis.

Modern imaging techniques, particularly MRI, are valuable for localizing skull base lesions, showing vascular structures without needing angiography.8 X-rays and CT scans of skull GCTs often show expansive and occasionally lytic bone lesions, typically without the classic 'soap bubble' appearance. MRI effectively demonstrates soft-tissue extension and relationships with surrounding structures. GCTs usually appear hypointense or isointense on T1-weighted images (WI) and T2WI with contrast enhancement, a pattern seen in the present case. 10,11 Major radiological differential diagnoses include chordoma, giant-cell reparative granuloma, aneurysmal bone cyst, fibrous dysplasia, 'brown tumor' hyperparathyroidism, eosinophilic granuloma, and plasmacytoma. The clinical behaviour of GCTs is unpredictable, making treatment controversial. The preferred treatment for cranial GCT is radical surgical removal of the diseased bone. However, complete removal may be impossible due to the tumor's anatomical location or involvement of vital structures, as in the present case, where a minimally invasive intralesional approach was used. This results in a high recurrence rate, highlighting the importance of adjuvant therapy. 12,13

CONCLUSION

In summary, giant cell tumors of the skull are rare conditions, but their radiological features are crucial for diagnosis, treatment planning, and follow-up monitoring. Advanced imaging techniques like CT and MRI provide vital information about tumor extent and characteristics. assisting clinicians in delivering optimal care to patients. However, imaging alone cannot definitively distinguish lesions, highlighting the necessity histopathological examination for a conclusive diagnosis. Histologically, GCTs are generally considered benign, but they can display locally aggressive behavior with a high recurrence rate of up to 60%. Additionally, they have the potential for distant metastasis, most commonly to the lungs, occurring in approximately 4% of patients with GCT.

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