Aggressive Osteoclastoma of Sphenoid Sinus: A Rare Surgical Case Report

¹G Sundhar Krishnan, ²Narendra Kumar

ABSTRACT

The giant cell tumor (GCT) or osteoclastoma is considered to be a locally aggressive benign tumor. The GCTs of the cranium represent only 1% of all GCTs and preferentially affect the sphenoid and temporal bones. We report a case of an 18-year-old male who presented with headache and diplopia. Radiological investigation shows a destructive midline mass involving the body of the sphenoid. The tumor was debulked endoscopically and histopathology reported as osteoclastoma of sphenoid sinus. Radiotherapy and zoledronate was given. We report this case due to its extreme aggressive nature of growth, which is a challenge to treat, and unique presentation in teenaged male, which is rare.

Keywords: Giant cell tumor, Osteoclastoma, Skull base, Sphenoid sinus.

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INTRODUCTION

Giant cell tumor (GCT), also called as osteoclastoma, involves the epiphysis of long bones. Giant cell tumor involving the skull tends to arise in the sphenoid or petrous part of the temporal bone and its incidence is less than 1% of all bone GCTs.¹ Giant cell tumor occurs most commonly in the 3rd and 4th decades of life and is predominant in females.² Osteoclastoma of the sphenoid bone is usually detected when the lesion is large and has expanded into the adjacent anatomical structures and cause symptoms. We report a case of an 18-year-old male who presented with headache and diplopia, and was diagnosed as osteoclastoma of sphenoid sinus and its aggressive nature of growth.

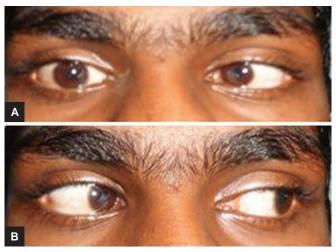
CASE REPORT

An 18-year-old male came to our hospital with complaints of headache for 6 months and diplopia for 15 days. On

¹Professor, ²Resident

^{1,2}Department of ENT, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai, Tamil Nadu, India

Corresponding Author: G Sundhar Krishnan, Professor Department of ENT, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai, Tamil Nadu, India Phone: +919841030992, e-mail: sundharkrishnan2@gmail.com examination, he had right lateral rectus palsy (Figs 1A and B), but all other ENT and neurological examinations were normal. The ophthalmologist gave an opinion as isolated right lateral rectus palsy, normal acuity of vision, and no papilledema. Diagnostic nasal endoscopy showed smooth bulge in the roof of the nasopharynx. In computerized tomography (CT), a destructing midline mass involving the body of the sphenoid is seen (Fig. 2). Magnetic resonance image showed a mass extending to the inferior aspect of the right cavernous sinus with possible involvement of the right abducent nerve. Floor of the sella and intersellar contents including pituitary were normal with no brainstem compression (Figs 3A to C).

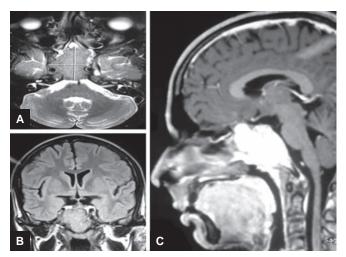


Figs 1A and B: Right lateral rectus palsy



Fig. 2: A destructing midline mass involving the body of the sphenoid is seen

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Figs 3A to C: Mass extending to the inferior aspect of the right cavernous sinus

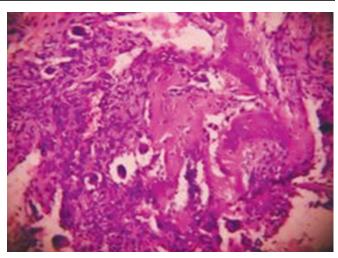
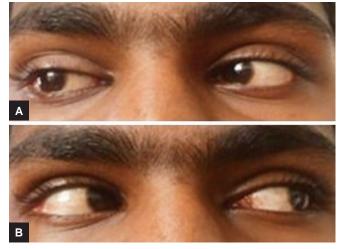


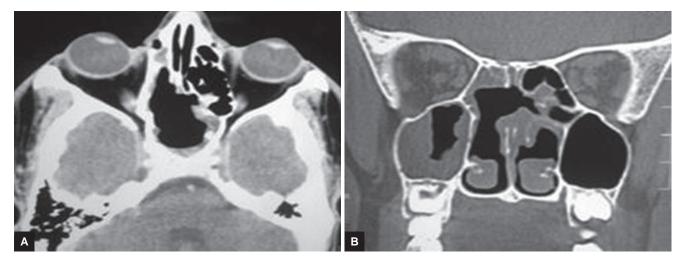
Fig. 4: HPE, evenly distributed multinucleate giant cells with coarse chromatin, prominent nucleoli



Figs 5A and B: Postoperatively, lateral rectus palsy improved

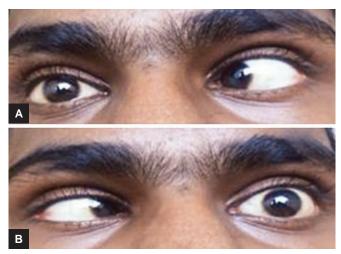
We did transnasal endoscopic excision of the mass. Histopathology report showed admixture of evenly distributed multinucleate giant cells and mononuclear cells having round-to-oval nuclei with coarse chromatin, prominent nucleoli, and display mitotic activity. Multinucleate giant cells show similar nuclear pattern, suggestive of osteoclastoma (Fig. 4). Postoperatively, lateral rectus palsy improved (Figs 5A and B). Postoperative scans showed disease-free images (Figs 6A and B).

One-month later, the patient presented with bilateral lateral rectus palsy (Figs 7A and B) and CT scans showed soft tissue density involving sphenoid sinus extending into clivus, petrous apex, clinoid process, and right cavernous sinus (Figs 8A to C). The patient was given palliative radiotherapy – 300 centi gray × 10 fractions and the tumor was debulked. Biopsy report again showed features suggestive of osteoclastoma. Postoperatively, there was mild improvement in lateral rectus palsy (Figs 9A and B) and CT scan showed the presence of residual disease (Figs 10A and B). We started with bisphosphonate, such as zoledronate 4 mg IV over 30 minutes once a month for 4 months and the patient symptomatically improved.

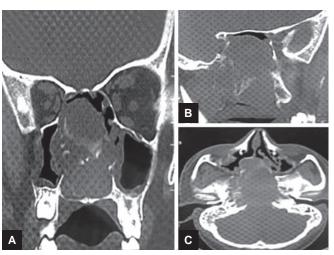


Figs 6A and B: Postoperative CT shows disease-free images

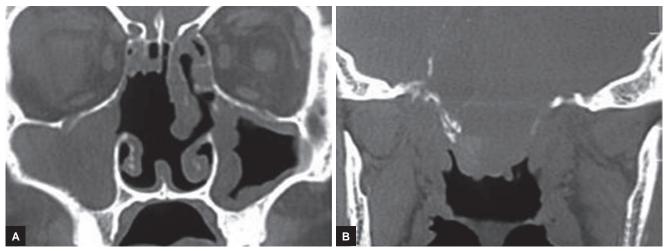
G Sundhar Krishnan, Narendra Kumar



Figs 7A and B: Bilateral lateral rectus palsy



Figs 8A to C: Repeat preoperative CT scan



Figs 9A and B: Repeat postoperative CT scan



Figs 10A and B: Final postoperative picture

REVIEW OF LITERATURE AND DISCUSSION

Giant cell tumors of the sphenoid bone, despite their benign histologic appearance, are clinically fatal, due to their location and capacity for local destruction of the vital structures.³ Embryologically, the sphenoid and the temporal bones are produced by an endochondral bone formation, except for the greater wings and the pterygoid processes of the sphenoid. While the other skull bones are generated by an intramembranous bone formation. This clearly explains the relative absence of osteoclastoma in the other calvarial bones.^{3,4}

The most common symptom is headache, followed by diplopia. The expansile nature of GCT to the cavernous sinuses is responsible for the frequent involvement of the third and sixth cranial nerves.⁵

Giant cell tumors are extradural and do not invade the intradural compartment. In CT scanning, osteoclastoma appears homogeneous hyperdense mass, which was highly enhanced with contrast medium. Bony erosions are also demonstrated.⁶ Magnetic resonance image clearly demonstrates the soft tissue extension and association with the surrounding structures.³ Compared with GCTs of long bones, X-ray and CT scan of skull GCTs lack the characteristic sign of the "soap bubble" appearance.^{1,7} Differential diagnosis of the osteoclastoma includes bone-invading tumors, such as chondrosarcoma and chordomas, and mostly nontumor diseases, such as aneurysmal bone cyst, giant cell reparative granuloma, "Brown tumor" of hyperparathyroidism, and fibrous dysplasia.^{4,6}

The surgical treatment of GCTs is radical removal of the lesion. The most preferred route is anterior approach, because it allows more direct access to the clivus as they place the pathology at the center of the surgical field while leaving neurovascular structures peripherally.

Radiotherapy is also indicated in osteoclastoma that cannot be resected completely, patients who cannot undergo surgery because of comorbid factors, or for patients with progressing or multiple recurrences after surgical excision.⁸ Bisphosphonate administration is also recommended for the treatment of GCT which slows down bone resorption, allowing the bone-forming cells time to rebuild normal bone and allowing bone remodeling.¹

In conclusion, advantages of endoscopic endonasal approach include the utilization of natural corridors, improved visualization by using a light source closer to the pathology, the possibility to look laterally with angled endoscopes, and the ability to approach the clivus without manipulating critical neurovascular structures and thereby removing the tumor easily.⁹

A significant amount of experience, coordination, and teamwork by surgeons can treat clival lesions easily through endoscopic endonasal approach.¹⁰

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