

## Case Report

# A rare case of giant cell tumor involving the clivus resected through Le Fort I Osteotomy and median maxillotomy

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## Abstract

**Background:** Giant cell tumors (GCTs) are bone tumors that seldom involve the skull. Skull GCTs preferentially occur in the sphenoid and temporal bones with few reported cases involving the clivus. Due to the rarity and complex location, surgical management is not well established for clival GCTs.

**Case Description:** A 49-year-old male presented with headaches and blurred vision in the right eye for 2 weeks. Computed tomography (CT) with contrast revealed a sellar mass eroding through the sphenoid sinuses with compression of optic chiasm. Biopsy was consistent with GCT. Patient underwent tumor resection by Le Fort I Osteotomy and median maxillotomy for an extended transsphenoidal approach. Upon discharge, patient showed no neurological deficits and intact cranial nerves.

**Conclusion:** This case contributes to the limited amount of skull-based GCT cases worldwide. Additionally, the extended transoral approach can be performed safely in the context of a GCT within the clivus with acceptable morbidity and cosmesis.

**Key Words:** Clivus, giant cell tumor, Le Fort osteotomy, skull-based neoplasm

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## INTRODUCTION

Giant cell tumors (GCTs) comprise 5% of skeletal tumors.<sup>[8]</sup> GCTs involving the skull tend to arise in the sphenoid or petrous part of the temporal bone and account for less than 1% of all bone GCTs.<sup>[8]</sup> The typical patient is a female who is aged 20–40 years.<sup>[7,9]</sup> Because skull GCTs are extremely rare, there is little established surgical management. Reported cases of clival GCTs describe a transnasal, transmaxillary, or endoscopic endonasal approach (EEA) for resection.<sup>[5,7-8,11]</sup> We describe a GCT involving the clivus removed by a Le Fort I osteotomy (LFO) and median maxillotomy (MM) approach.

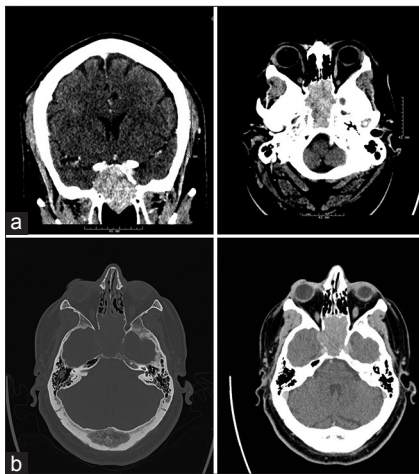
## CASE REPORT

A 49-year old male presented to the emergency department with headaches and blurred vision in the right eye for 2 weeks. Computed tomography (CT) with contrast revealed a 4.9 × 3.2 cm sellar mass with extension through the sphenoid sinus and posterior ethmoid air cells, compression of the optic chiasm and right optic nerve, and bony destruction of the clivus [Figures 1 and 2]. The patient was unable to have magnetic resonance imaging because of an implantable cardiac defibrillator. Laboratory studies revealed panhypopituitarism and slight hyperprolactinemia, consistent with stalk effect.

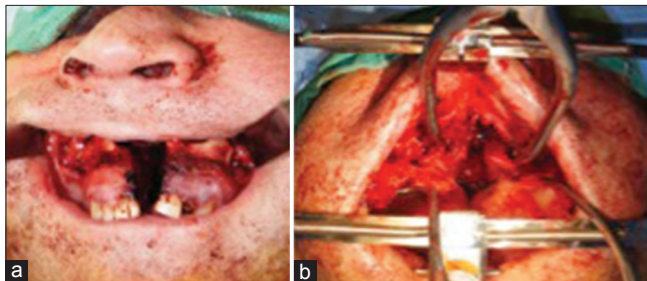
## Management and outcome

Given the atypical appearance of this lesion, a transnasal endoscopic transsphenoidal biopsy was performed for tumor diagnosis. The transsphenoidal biopsy was technically challenging due to the patient sustaining five previous nasal fractures and associated septal deviation. Biopsy revealed neoplasm composed of polygonal mononuclear cells with intermixed uniformly distributed giant cells (some containing more than 40 nuclei). Mitotic count was low (less than 5 mitoses per 10 high power fields) with no identifiable atypical mitoses. There was no tumor necrosis or vascular invasion in form of intravascular plugs by tumor cells. Morphologic findings were consistent with bone GCT.<sup>[2]</sup> A multidisciplinary surgical plan was developed for the patient to undergo resection of the GCT with a LFO and MM approach.

Patient underwent a tracheostomy followed by Mayfield head-holder immobilization and registration of frameless stereotactic neuronavigation (Medtronic Stealth System). A full-thickness palatal split was performed with median incision spanning from the left of the uvula to between the incisors. Bilateral gingival buccal sulcus incisions were made that extended into the subperiosteal plane



**Figure 1: Preoperative Postcontrast CT Scans of the Head.** Axial and coronal CT scans were used to assess margins of the sellar mass. (a, b) The tumor extended into the sphenoid sinuses and eroded through the clivus and medial aspect of the cavernous sinus on right side (b)



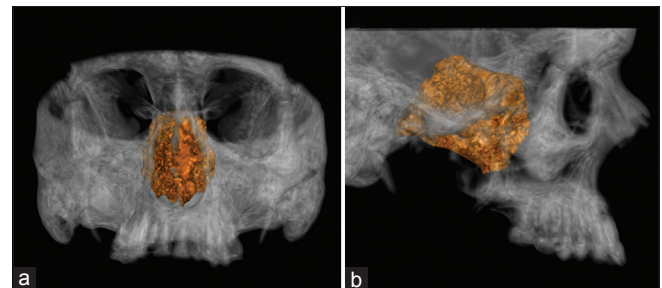
**Figure 3: Median maxillotomy was performed with preservation of anterior nasal spine (a).** Each segment of hemi-maxillae was retracted inferiorly and laterally with stabilization of the clivus (b)

and subsequently the bilateral pterygoid plates were dissected.

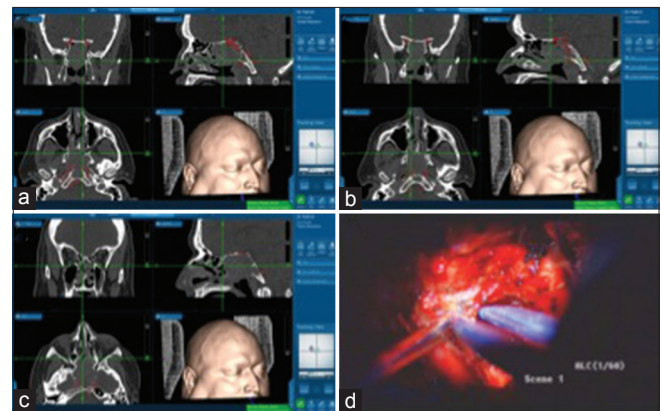
Prior to osteotomy, plates were preregistered along the nasomaxillary, zygomaticomaxillary buttresses and across the maxilla inferior to the anterior nasal spine. A standard LFO was performed with midline maxillotomy to separate the maxilla between the incisors [Figure 3a]. The separated maxillae were rotated and retracted laterally [Figure 3b]. The clivus was identified and an additional mucosal incision was made for proper tumor exposure.

Following exposure of the sella turcica and inferior clivus, microdissection and stereotactic navigation was used to dissect the tumor off the medial walls of the cavernous sinuses bilaterally, optic chiasm and optic nerves superiorly, and the dura behind the clivus [Figures 4 and 5]. The mass was soft, friable, and yellowish-red in color. Following exploration and ensuring hemostasis, Surgifoam followed by Gelfoam was placed in the posterior margin of the resection [Figure 6]. Subsequently, an abdominal fat graft was placed in the resection cavity. The maxilla segments were reconstructed with preregistered plates and soft palate was reapproximated in layers [Figure 7].

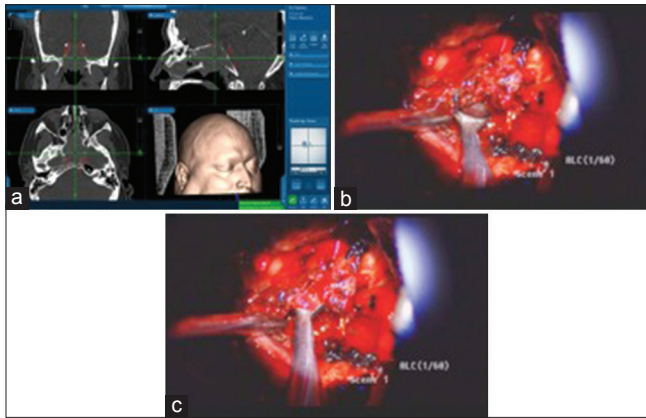
The patient was monitored for 2 days in the intensive care unit and transferred to the floor where his diet was



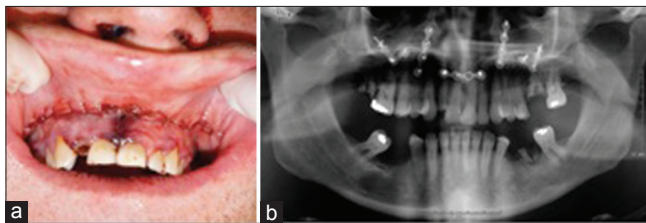
**Figure 2: 3-D images of the skull base tumor generated from the patient's preoperative CT (a, b)**



**Figure 4: Stereotactic neuronavigation was used intraoperatively to ensure adequacy of exposure before beginning microdissection (a-c).** Anterior surface of tumor was exposed followed by coagulation of tumor capsule (d)



**Figure 5:** Microdissection of the tumor continued in a piecemeal fashion (a-c)



**Figure 7:** Maxilla continuity was restored (a). Postoperative panoramic orthopantogram demonstrated healing of the maxilla segment (b)

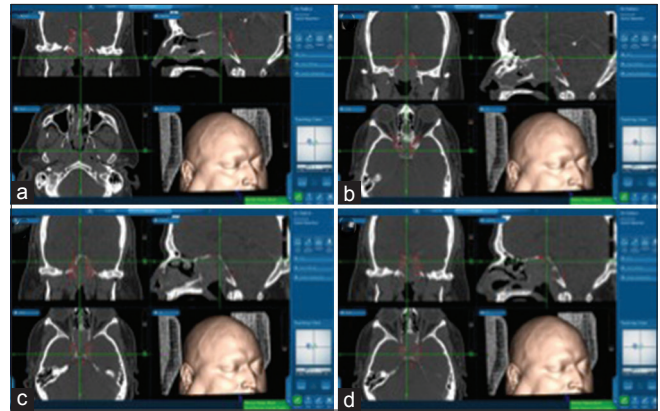
advanced 7 days after surgery. Postoperative imaging was obtained [Figure 8].

After surgery, he remained neurologically intact without cranial nerve abnormalities and reported vision improvement, confirmed by Humphrey Visual Fields testing. Histological examination revealed tumor morphologically identical to the previous biopsy [Figure 9]. Focal reactive bone formation was noted. No spindle cells with marked cytologic atypia were identified to indicate malignancy in the GCT.<sup>[6]</sup> Patient underwent external beam radiation therapy postresection to reduce the risk of tumor recurrence. He continues to be tumor-free and neurologically intact at 1-year follow-up.

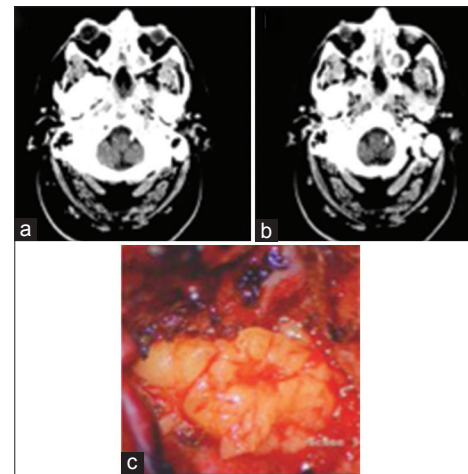
## DISCUSSION

Our case contributes to the few reported skull-based GCT cases worldwide. As demonstrated in previous case reports, the typical patient with skull GCT is a young female, while our patient was a 49-year-old male.<sup>[1,3,4,7,9,10,12,13]</sup> Although generally benign, GCTs can exhibit malignant features. Complete tumor resection is essential to prevent local recurrence, but is difficult for GCTs in this location due to the challenging exposure and the adjacent critical structures.

EEA is a newer, less invasive approach for clival GCTs, with reported advantages of better visualization by placing the lens and light source closer to the mass and lateral visualization with angled endoscopes.<sup>[5,11]</sup> Despite



**Figure 6:** Prior to completion, stereotactic neuronavigation was used to check the margins of the gross total resection including inferior margin (a), superior margin (b), and posterior margin (c). The medial aspect of the right carotid was inspected for sufficient decompression and decompression of the optic nerve (d)

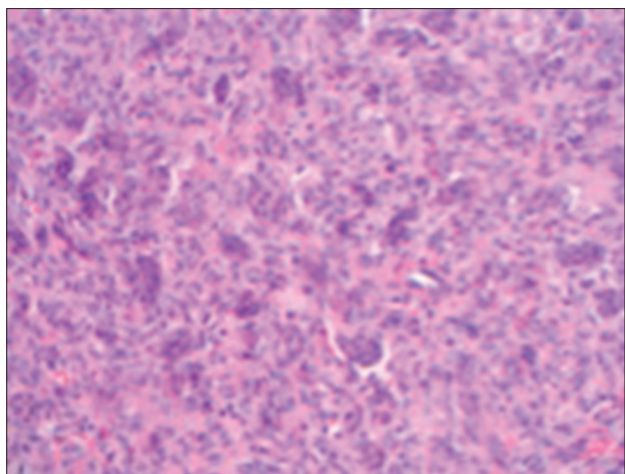


**Figure 8:** Postoperative Postcontrast CT Scans of the Head. CT images show fat graft in the center of resection cavity along with surrounding surfigoam and postoperative products (a, b). CT confirms midline fat graft placement during surgery (c)

these reported advantages, EEA would have been challenging in our patient with a history of five previous nasal fractures and a deviated septum. In addition, an advantage of the extended transoral approach includes direct visualization of the clival mass in the center of the surgical field with neurovascular structures placed laterally, maximizing our ability to safely obtain a gross total resection.

The transoral approach for clival lesions allows exposure alongside the midline of the inferior third of the clivus, the cervicovertebral junction and the C1/C2 complex. This allows a direct extradural approach without brain retraction. The extended transoral approach is used when the lesions extend beyond the exposure limits of a standard transoral approach and involves additional incisions and facial osteotomies to mobilize structures that may obstruct visualization of the lesion. Accurate





**Figure 9: The histopathology revealed the tumor was composed of mononuclear and giant cells. The giant cells were uniformly distributed throughout the tumor (H and E, ×400)**

reconstruction of the maxillofacial osteotomies is essential to achieve excellent cosmesis and avoid malocclusion. The following approaches allow for more superior exposure of the upper and middle clivus and sphenoid sinus: Transmaxillary (Le Fort I maxillotomy), transmaxillary palatal split, or the transpalatal. The LFO with MM approach was optimal for our exposure. Gupta *et al.* reported utilizing this approach for resection of a clival GCT in a 17-year-old female, but did not describe the surgical details.<sup>[3]</sup>

## CONCLUSION

Our case report demonstrates that the extended transoral approach can be performed safely in the context of a clival GCT with acceptable morbidity. Also, this case underscores the importance of multidisciplinary collaboration between craniofacial surgeons and neurosurgeons to optimize patient outcomes.

## ACKNOWLEDGMENT

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