ECRONICON OPEN ACCESS

EC NEUROLOGY
Case Report

Giant Cell Tumor of the Sphenoid Bone Extending into the Ethmoid Sinuses, Orbit, Both Cavernous and Intracranial Extension in an Adolescent

Shamsul Alam[1], Bipin Chaurasia[2]*, Sujan Sharif[3], Tauhidur Rahman[3], Abu Naim Wakil Uddin[4], Satish Kumar Shah[3] and Dhiman Chowdhury[5]

[1]Assistant Professor, Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

[2]Chief Resident, Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

[3]Resident, Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

[4]Medical Officer, Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

[5]Associate Professor, Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

***Corresponding Author:** Bipin Chaurasia, Chief Resident, Department of Neurosurgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.

Received: July 02, 2018; **Published:** August 30, 2018

Abstract

Giant cell tumor (GCT) constitutes 3 - 7% of primary bony neoplasms which rarely affect the sphenoid bone. Computed tomography (CT) scan shows expansile lytic lesion in the body of sphenoid with marked contrast enhancement. Magnetic resonance imaging (MRI) reveals a low to moderate signal intensity on T1- and T2- weighted images (T1WI and T2WI). "Soap bubble" may be seen on T2WI. It displays aggressive local behavior. Dural penetration, cerebral invasion and sarcomatous changes may occur. We present a case of GCT of the sphenoid bone with emphasis on its variable biological behavior, radiological features and management.

Keywords: *Giant Cell Tumor; Sphenoid Bone; Intracranial Extension; Staged Operation; Radical Extirpation*

Introduction

The skull base GCT has a predilection for the sphenoid and temporal bones. It most commonly involves the sphenoid bone followed by the petrous temporal bone [1]. GCT is regarded as a benign lesion. Rarely it has been found to be metastasizes to the lung. The most appropriate treatment is radical surgical extirpation which may not be possible due to involvement of vital structures. Recurrence depends upon the extent of tumor removal and adjuvant therapy.

Case Report

A 15 years old boy came to us with the history of headache since six months. Left sided weakness, right sided ptosis, loss of vision and epistaxis were present for 14 days. On examination, he had right sided third nerve palsy.

CT scan of head in axial and bone window (Figure 1B) sections demonstrated a lytic, brilliantly contrast enhanced soft-tissue mass was centered on the body of the sphenoid. The mass was extended posteriorly into the clivus, posterolaterally into the right petrous apex with erosion of the anterior surface of the bilateral anterior clinoid process, inferiorly into the sphenoid sinus, anteriorly into the sella turcica, ethmoid cells, lateral and posterior nasal cavity (Figure 1B).

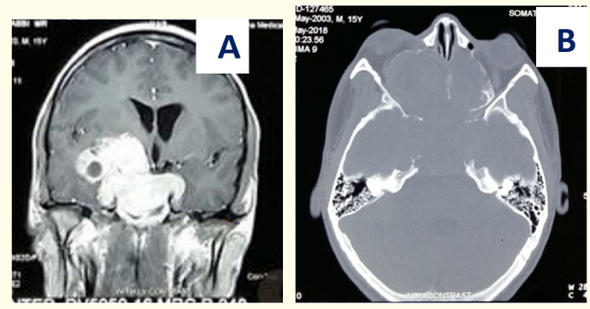

Figure 1: *(A) MRI of brain coronal section showing contrast enhancing lobulated mass in suprasellar, right parasellar extending to sylvian fissure, right thalamus compressing the third ventricle as well. (B) CT scan bone window view: Expansile lytic lesion causing thinning of medial wall of both orbit.*

MRI (Figure 1A) was performed with T1 and T2 weighted images in coronal, axial and sagittal planes before and after gadolinium injection. It demonstrated the mass arising from the sphenoid bone, displacing the normal pituitary gland upwards and eroding the floor of the sella turcica. The lesion had invaded the clivus, narrowed the pontine cistern, displaced the basilar artery backwards and had compressed the brain stem. It had also extended to both cavernous sinus, right thalamic and internal capsular region and had eroded the left petrous apex. The lesion was mostly isointense in T1WI and intermediate to hyper intense in T2WI.

There was T1 hypo intense and T2- hyper intense well circumscribed lesion present in suprasellar part of tumor which denoted the cystic component of the tumor. The tumor gave signal similar to that of the cortical grey matter on T1-weighted images and increased signal on T2- and proton density-weighted images. It showed homogeneous enhancement with gadolinium. Six months back, there was no existence of cystic component.

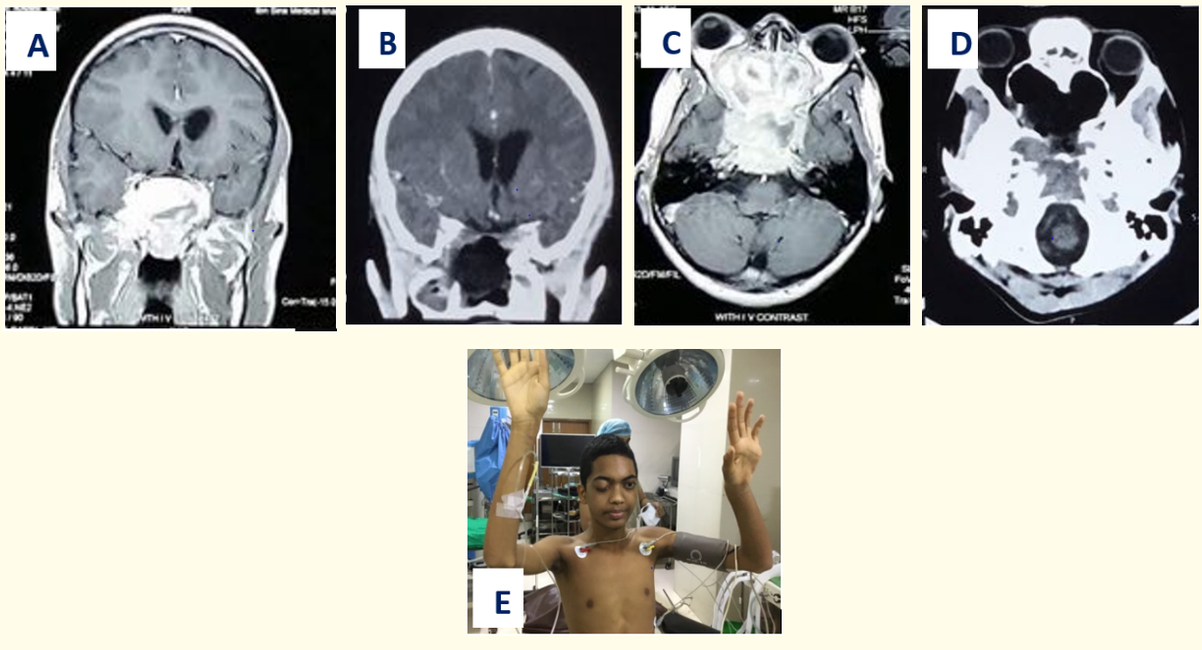

Figure 2: *(A) and (B) Pre and postoperative coronal images showing radical excision of sphenoidal, ethmoidal and orbital part of tumor. (C) and (D) Pre and postoperative axial images showing complete excision of sphenoidal, ethmoidal and orbital part of tumor through endoscopic, endonasal trans sphenoidal approach. (E) Consciousness level improved after first surgery but right sided ptosis still persisted (picture was published with patients kind permission).*

Surgical procedure

Staged surgery was performed for this rapidly growing giant cell tumor in a week apart. Endoscopic biopsy was done first followed by endonasal transethmoidal, trans-sphenoidal transmaxillary transorbital through lamina paparacia approach. Tumor was found soft to firm in consistency, highly vascular, mostly pinkish to whitish with some blackish in colour indicating necrosis. After removing ethmoidal air sinuses and tumor involving the sinus, anterior cranial base dura was seen which was kept intact. Medial wall of maxilla and orbit was drilled out. Both optic and carotid prominence was well appreciated, intracavernous part of carotid was navigated by doppler. Upper part of clivus and its involving tumor was taken out. Haemostasis was achieved. Three units of blood were transfused per-operatively. Postoperative period was uneventful (Figure 2E). Post-operative CT scan showed good removal of targeted part of tumor with residual cavernous, parasellar and intracranial part of tumor (Figure 2A-D).

One week later orbito-zygomatic single piece craniotomy (Figure 3A-C) was performed followed by extradural pilling and removal of cavernous part of tumor. After doing curvilinear durotomy and sylvian fissure dissection, tumor was taken out through optico-carotid, lateral to carotid, looking up view toward the frontal, temporal base, suprasellar, supratentorial and suprachiasmatic approach. Tumor was seen as whitish in colour. There was central necrosis in tumor which was of blackish in colour. The content inside cyst was blackish in colour which was sucked out and the wall of cyst was totally removed (Figure 3D-E). At the end of the surgery lateral midbrain, basilar bifurcation and third nerve were identified.

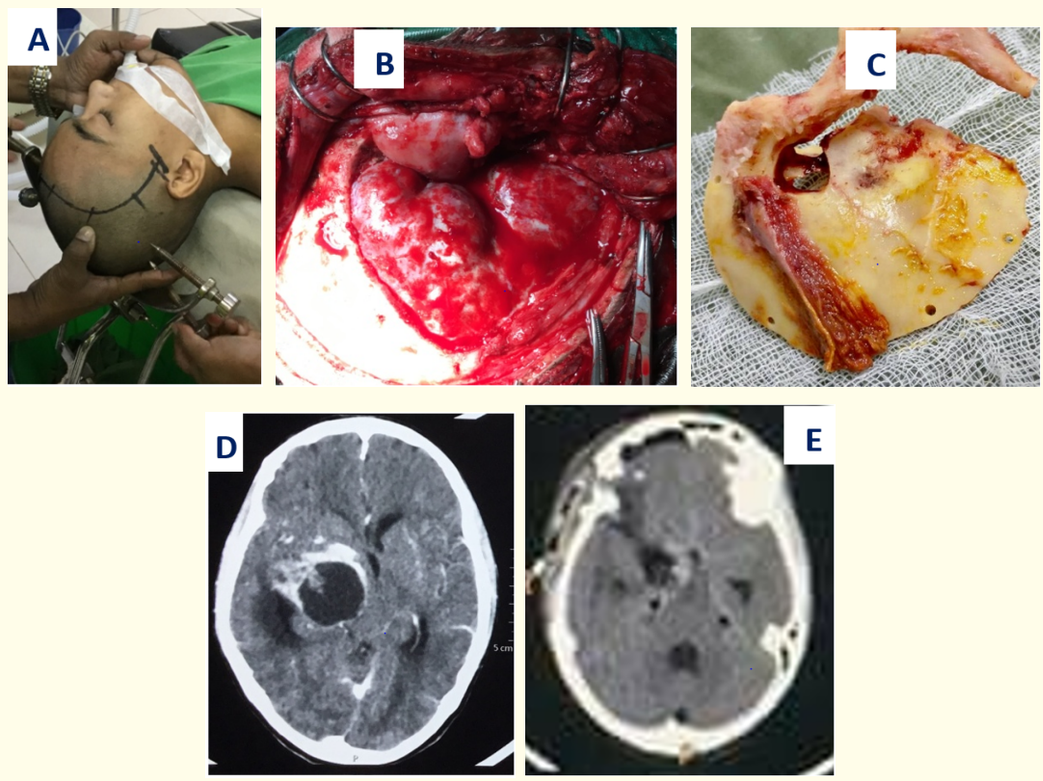

Figure 3: *(A) Per-operative image of second surgery: supine position, head extension and malar prominence in highest point(picture was published with patients kind permission); (B) and (C) Orbito- zygomatic one piece craniotomy for better exposure of 'looking up' extension of tumor; (D) and (E) Pre and post op image showing complete resection of intracranial extension of tumor.*

Histopathology (Figure 4: H&E stain 20X) revealed osteoclast like giant cells, spindle cells, many histiocytes and small number of lymphocytes in a fibrovascular stroma. A few mitoses were noted. Osteoid formations were present. Post operatively his headache and weakness were improved. Vision didn't improved. Ptosis was present.

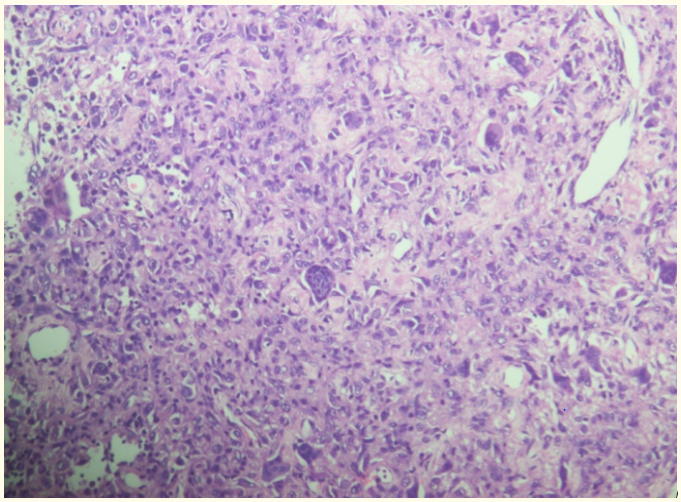

Figure 4: *H&E staining (H&E stain 20X): Section showing osteoclast like giant cell, spindly cell, histiocytes and small number of lymphocytes in a fibrovascular stroma.*

Discussion

Giant cell tumors comprise 3 - 7% of bone tumors and 90% of these involve the end of long bones such as the distal femur, proximal tibia, fibula and distal radius or ulna. They are seldom found in the skull [2-5]. Most reported cases of giant cell tumors of the skull involve the sphenoid and temporal bones of the middle cranial fossa [2]. Embryologically these are generated through endochondral bone formation. The relative infrequency of this tumor in other bones of the skull may be related to their membranous bone formation [6,7]. Giant cell tumors are histologically variable but usually benign tumor. It has some features of local aggressiveness [3]. Metastases appear in 2%of cases [5].

Giant cell tumors most commonly occur in the third or fourth decade with a female preponderance. Signs and symptoms depend on the site of the lesion. The most common are headache, visual disturbances such as diplopia, and cranial nerve palsies [2,8]. Plain skull radiographs most commonly show bony erosion, destruction of the sella turcica and the body of the sphenoid with or without a soft-tissue mass in the sphenoid sinus [10]. CT with thin (1 - 3 mm) slices is the best way to show the bone destruction. A combination of axial and coronal sections appears to be ideal for demonstrating bone erosion and extension into the sphenoid sinus [9]. The tumor is a lytic, homogeneous soft-tissue mass which enhances with intravenous contrast medium [7]. Calcification is generally absent [2,11].

MRI especially in the sagittal plane is useful to show the extension into the sella turcica and the displacement of the normal pituitary gland, thereby excluding a pituitary tumor which can otherwise have the same radiological features [8]. MRI is also of importance for showing extension into the cavernous sinuses. The signal from the tumor is nonspecific: low to intermediate signal on T1-weighted intermediate to high on T2-weighted images. There is rather homogeneous contrast enhancement [4,7]. A cystic component can be present with signal similar to that of cerebrospinal fluid [12]. In our case most part of tumor was isointense to gray mater. Small foci of hyper intense signal was noted in both T1W and T2W images which was probably due to vascular nature of tumor. Giant cell tumors sometimes have a slightly heterogeneous signal on T2-weighted images, with areas of decreased signal, which can be correlated with haemosiderin deposition secondary to hemorrhage [13].

The differential diagnosis of giant cell tumor includes eosinophil granuloma, giant-cell reparative granuloma, ectopic pituitary adenoma and the brown tumor of hyperparathyroidism [3,7,8,14]. If calcification is present chordoma, ectopic craniopharyngioma, meningioma, chondroma and chondrosarcoma enter into the differential [8,12]. Endocrine studies may be necessary to exclude an invasive prolactinoma, hypopituitarism secondary to the compression of the pituitary gland by the tumor, or hyperparathyroidism.

A complete surgical excision is generally the gold standard of treatment [15]. According to previous articles in the medical literature, the local recurrence rate varies widely from 7% to 60% and depends on the extent of surgery (e.g. en bloc resection or curettage only) [15-17]. For good local control, a wide local excision is preferred as the first step. However, en bloc resection for a cranial GCT is often difficult and may cause cosmetic or neurologic deficits. Radiotherapy had been used but the results were not promising [2,4,6,18]. The response of giant cell tumor to chemotherapy is disappointing [18]. Although it is virtually impossible to provide a specific radiological diagnosis, as the variety of the skull base lesions is very wide, modern imaging techniques have proved to be of value in localizing and determining the extent of skull base lesions.

Conclusion

Giant cell tumour may have intraorbital and intracranial extension and present with third nerve palsy, endocrine disturbances similar to pituitary tumor. Presence of cystic component may mimic the craniopharyngioma. MRI remains the diagnostic investigation of choice. Continuous improvement in skull base approaches and staged surgery have made these difficult tumor amenable to radical excision. Periodic follow-up is necessary to check the recurrence.

Bibliography

1. Gupta R., *et al*. "Giant cell tumour of the clivus". *British Journal of Neurosurgery* 22.3 (2008): 447-449.
2. Rock JP., *et al*. "Giant cell tumor of the skull base". *The American Journal of Otology* 15.2 (1994): 268-272.
3. Findlay JM., *et al*. "Giant-cell tumor of the middle cranial fossa: case report". *Journal of Neurosurgery* 66.6 (1987): 924-928.
4. Motomochi M., *et al*. "Giant cell tumor of the skull". *World Neurosurgery* 23.1 (1985): 25-30.
5. Henderson B and Whitwell H. "Giant- cell tumor of the skull: case report". *Neurosurgery* 23.1 (1988): 120-122.
6. Watkins L., *et al*. "Giant-cell tumors of the sphenoid bone". *Neurosurgery* 30.4 (1992): 576-580.
7. Kioumehr F., *et al*. "Giant-cell tumor of the sphenoid bone: case report and review of the literature". *Canadian Association of Radiologists Journal= Journal l'Association Canadienne des Radiologistes* 41.3 (1990): 155-157.
8. Wilbur AC., *et al*. "Giant cell tumor of the sphenoid bone mimicking a pituitary tumor". *American Journal of Neuroradiology* 7.2 (1986): 361-362.
9. Rimmelin A., *et al*. "Giant-cell tumour of the sphenoid bone: case report". *Neuroradiology* 38.7 (1996): 650-653.
10. Wolfe III JT., *et al*. "Giant-cell tumor of the sphenoid bone: review of 10 cases". *Journal of Neurosurgery* 59.2 (1983): 322-327.
11. Carmody RF., *et al*. "Giant cell tumor of the sphenoid bone". *Journal of Computer Assisted Tomography* 7.2 (1983): 370-373.
12. Rosen MP. "General diagnosis case of the day. Giant-cell tumor of the temporal bone". *American Journal of Roentgenology* 156.6 (1991): 1290-1292.
13. Aoki J., *et al*. "Giant cell tumors of bone containing large amounts of hemosiderin: MR-pathologic correlation". *Journal of Computer Assisted Tomography* 15.6 (1991): 1024-1027.

14. Harwood-Nash DC and Fitz CR. "Neuroradiology in infants and children". Mosby (1976).

15. Isaacson B., *et al*. "Giant cell tumor of the temporal bone: management strategies". *Skull Base* 19.4 (2009): 291-301.

16. Zorlu F., *et al*. "Malignant giant cell tumor of the skull base originating from clivus and sphenoid bone". *Journal of Neurooncology* 76.2 (2006): 149-152.

17. Lee HJ and Lum C. "Giant-cell tumor of the skull base". *Neuroradiology* 41.4 (1999): 305-307.

18. Manaster BJ and Doyle AJ. "Giant cell tumors of bone". *Radiologic Clinics of North America* 31.2 (1993): 299-323.

Volume 10 Issue 9 September 2018