Giant cell tumour of the sphenoid bone – Lefort 2 approach for surgery

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Citation

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Abstract

Giant cell tumour is one of the rare tumors of the sphenoid bone .The tumour usually has delayed presentation due to late onset of symptoms .The excision is relatively difficult due to narrow corridors available for surgery. We have described here our experience with use of Le Fort 2 osteotomy in removal of the tumour .Despite having adequate corridor, we could not achieve total excision and had to administer adjuvant therapy for control of disease.

INTRODUCTION

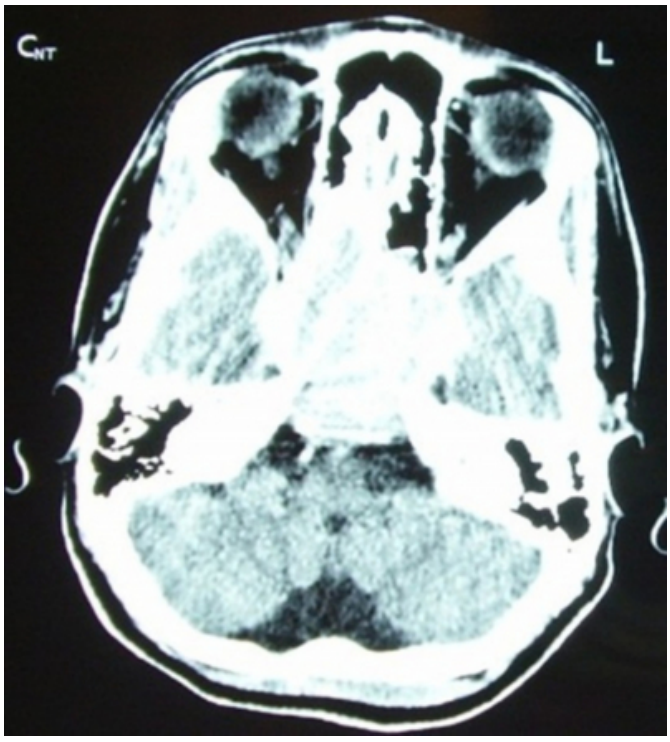
Giant cell tumour (GTC) is primarily a tumor of long bones of uncommon variety, accounting for less than 5% of all bony tumors. [1,6,9,18] They are rarely located in cranium, where it usually affects sphenoid and temporal bones. [18] GCT of the sphenoid bone accounts for 1-2 % of all Para nasal sinus tumors. [7,5] GCT of sphenoid may present initially with non-specific symptoms like headache. The patient can have cranial nerve palsies, endocrinopathy due to involvement of pituitary. [16,12,14,15] The tumor may spread to involve orbit or laterally into sub- temporal region or may enlarge posteriorly with compression of brain stem .The removal of such tumors is difficult because of its location and limited access. Various skull base approaches have been described to remove these tumors. [15]

CASE REPORT

A 24-year-old female presented in our hospital with history of headache for and diplopia on seeing towards left side for past two months. She had left sixth cranial nerve palsy. There was no other neurological deficit. CT scan revealed a large tumour measuring 5.4x6.2x5.5 cm involving sphenoid bone, extending to clival-basiocciput region, and eroding the sella. (Figure 1)

Figure 1

Figure 1: CT scan showing tumour-involving sphenoid bone, extending to clival-basiocciput region, and eroding the sella



MRI defined the extent of tumour filling the sphenoid sinus and eroding the sellar floor and the mass was abutting the cavernous sinus, predominantly involving the left side, eroding the clinoids on left side, and inferiorly involving whole of clivus. (Figure 2a, b)

Figure 2

Figure2a,b: MRI showing tumour filling the sphenoid sinus and eroding the sellar floor extending to involve whole of clivus

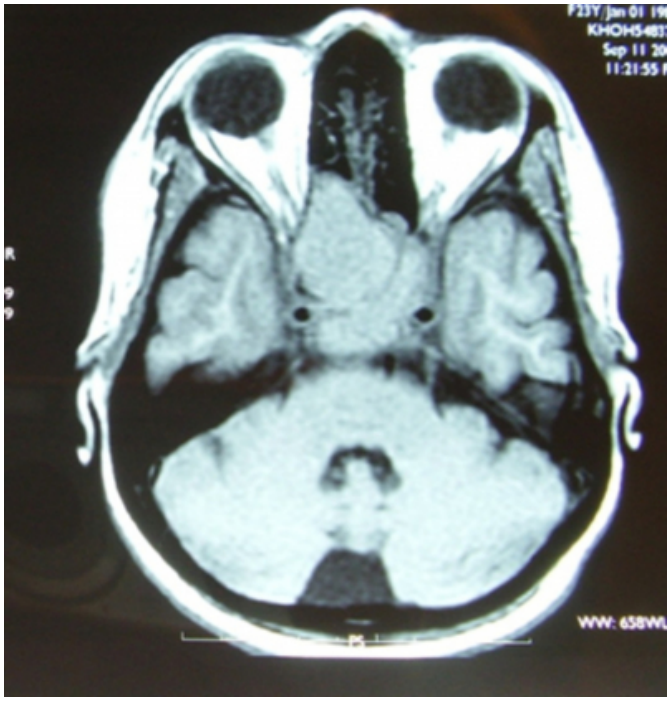
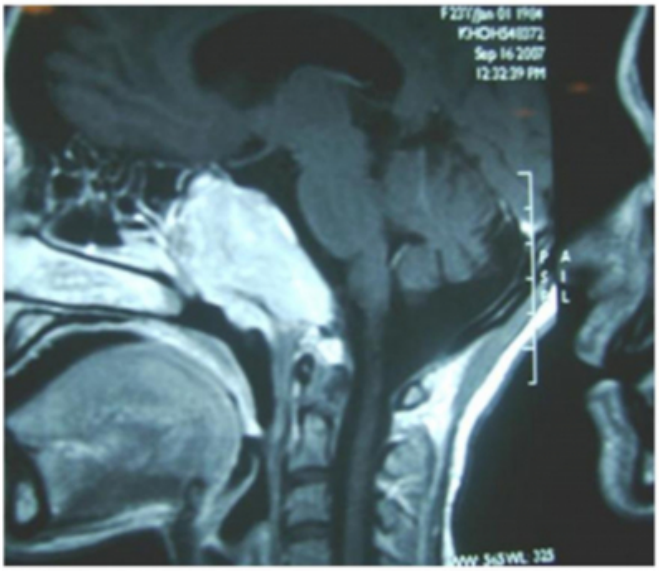


Figure 3

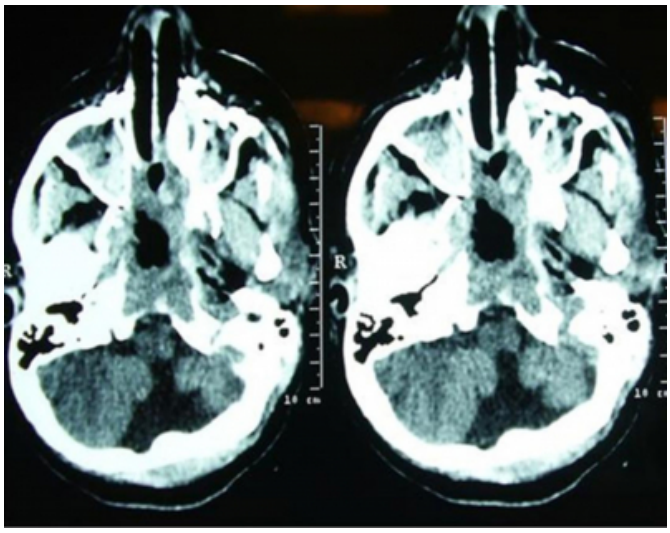


Tumour was encroaching carotid arteries bilaterally. .A pre op angiogram was done which showed vascular tumour with bilateral blood supply. Complete endocrinal profile was done which was normal. Serum electrolytes, calcium and phosphates were also normal. Before coming to this hospital, she had already undergone trans-nasal biopsy of the lesion, which was suggestive of Giant cell tumour.

The patient was operated through transfacial approach with Weber Fergusen incision with lip split with bilateral front maxillary swing and Le Fort 2 osteotomy. Midlpalatal split with sphenoidotomy was also done for exposure .The tumour excision was done but the tumour was vascular and once posterior part was reached, there was lot of bleeding and we decided not to remove the tumor further in this sitting. Her post op recovery was good with no new neurological deficits. Postopertive CT scan revealed presence of residual tumour. (Figure 3)

Figure 4

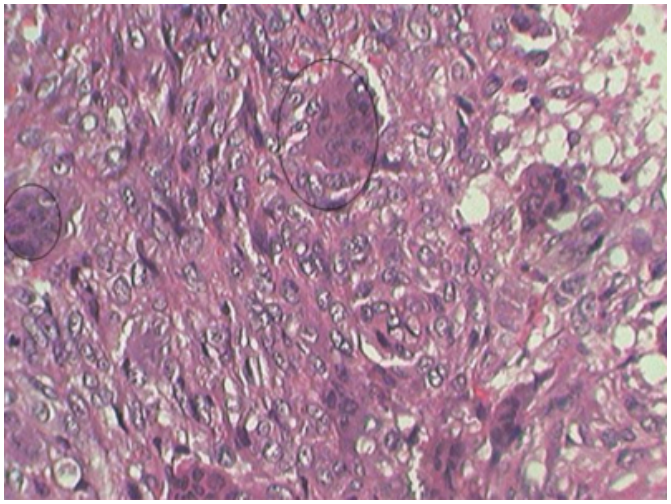
Figure 3: Postoperative CT scan showing residual tumour



She was advised re-surgery but she refused and hence she was referred for gamma knife surgery. The histopathogical examination revealed presence of tumor composed of two cell population. The mononuclear cells were oval to spindle shaped with eosinophilic cytoplasm arranged in sheet like pattern .The giant cells were distributed throughout the lesion with focal clustering at periphery. Giant cell nuclei were upto twenty in number. .There was no abnormal mitosis or nuclear atypia (Figure 4). Hence, diagnosis of giant cell tumor was made.

Figure 5

Figure 4: Microscopic appearance of the tumour showing multiple giant cells (having numerous nuclei) with interspersed spindle cells



DISCUSSION

The tumors are usually homogenous and hyper dense enhancing on contrast. [11] Bony tissue is destroyed by the tumour with local mass formation. The various approaches described in literature for this tumour involve midfacial approaches (Transoral, transpalatal, lateral rhinotomy and midfacial degloving) and lateral approaches include infratemporal approach. Each procedure has certain advantages and disadvantages. [15]

GCT involves sphenoid bone possibly because it is derived from endochondrium. [13] These are locally aggressive tumors with variable malignant potential, and are vascular tumors with multiple giant cells seen. They usually arise from epiphysis of long bones and rarely involve skull. . [9,6,1,18] .GCT is extradural tumour with a rare intradural involvement. [4] Le Fort osteotomy was first described by Langenbeck which was later on modified .[15] The combination of trans palatal and bilateral maxillary

swing with Le Fort 2 osteotomy allowed us an extensive exposure of the tumor which could not be provided by Le Fort1 osteotomy.

The diagnosis of GCT is based on histopathological examination .The tumour consists of multinucleate giant cells, which are evenly distributed amongst smaller spindle shaped cells .The extent of mitosis is variable. The main differential diagnoses are from tumors of hyperparathyroidism, giant cell reparative granulomas, aneurysmal bone cyst, and the bone invading tumors like chondrosarcoma, chordoma and fibrous dysplasia. [17,18]

The giant cell reparative granuloma is a reaction to trauma and intraosseous hemorrhage. They have clumping of giant cells around the areas of hemorrhage unlike in GCTs. Besides GCT is found in age group of 25-40 years, while reparative granuloma is usually seen in patients younger than 20 years of age. The present lesion did not have blood filled cavernous spaces; hence, aneurysmal bone cyst is excluded. Brown tumors of hyperparathyroidism are excluded as clinical features of hyperparathyroidism were not there and serum calcium was normal.

Fibrous dysplasia and cartilage forming tissues are also excluded here, as there was absence of foci of osteoid and bone formation and there were no mitotic figures seen. . [17,18,2]

The goal of surgery is total removal but this is not always possible due to involvement of adjoining structures especially carotids .The brisk bleeding we encountered was also one of the contributing factors that limited the extent of removal that we could achieve. The best option available for residual tumour is re-exploration; however, in certain cases radiotherapy has been given. The role of radiotherapy is highly controversial, as it has been

seen to induce malignant changes. [1783]Since our patient refused second surgery and there was significant residual tumour we performed gamma knife surgery to the residual tumour.

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