International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Giant Cell Tumor of Temporal Bone: A Rare Case

Dr. C.S. Supreeth[1], Dr. Prakash. S. Mahantahshetti[2], Dr. S.S. Mahantahshetti[3]

[1]Resident, [2]Assistant Professor, [3]Professor and HOD,
Neurosugery, Jawaharlal Nehru Medical College, Belgavi.

Corresponding Author: Dr. C.S. Supreeth

Received: 12/12/2016 *Revised: 28/12/2016* *Accepted: 05/01/2017*

ABSTRACT

Giant cell tumor, (GCT) compromise 5% of skeletal tumor .GCTs involving the skull tend to arise in sphenoid at petrous part of temporal bone and account for less than 1%. Of their relative absence in cranium, temporal bone GCT is not surprising, as less than 25 case of Giant cell tumor of the temporal bone have been reported in world literature. It is a Benign Neoplasm but locally aggressive. Radical surgical removal is preferred modality of treatment. In this case report we had managed a similar case who presented with only pain and headache without any lateralizing deficits. MRI showed presence of large extra axial lesion in right medial temporal region which was confirmed as right temporal giant cell tumour and posted for surgical excision. Post surgery patient had good resolution of symptoms with no surgical complications. She is on regular follow up without any evidence of recurrence. We present this case in view of rarity of this tumour and good outcome following surgery.

Key words: Giant cell tumor, Temporal bone, Extra-axial, Radical excision, Radiation therapy

INTRODUCTION

Cooper in 1818 first described Giant cell tumors (GCT) of the bone. [(1)] Later Nelaton showed their local aggressiveness, and Virchow revealed their malignant potential. GCT represents approximately 5% of all primary bone tumors. [(2,3)] More than half of these lesions occur in the third and fourth decades of life. [(3)] GCTs are benign tumors with potential for aggressive behaviour and capacity to metastasize. Although rarely lethal, benign bone tumors may be associated with a substantial disturbance of the local bony architecture that can be particularly troublesome in peri-articular locations. There is no widely held consensus regarding the ideal treatment method selection. There are advocates of varying surgical techniques ranging from intra-lesional curettage to wide resection. GCTs involving the skull tend to arise in sphenoid at petrous part of temporal bone and account for less than 1%. [(4)] Of their relative absence in cranium, temporal bone GCT is not surprising, as less than 25 cases of Giant cell tumor of the temporal bone have been reported in world literature. Radical surgical removal is preferred modality of treatment. [(5,6)] We present this case in view of rarity of this tumour and good outcome following surgery.

CASE REPORT

A 45 Years female presented with history of pain in right ear which used to increase with jaw opening leading to restricted movement of mouth. There was also history of right sided throbbing headache with giddiness since 3 months which was associated with above complaints. No H/o hearing loss, seizures or trauma.

General and neurological examination revealed no focal deficits. MRI Brain with contrast showed- Ill defined enhancing extra axial lesion of size 3cm X 2.2cm X 3.5 cm with interspersed necrotic areas and extensive adjacent bony invasion causing mass effect on right medial temporal region. (Fig1) HRCT of temporal bone was done to confirm the extent.

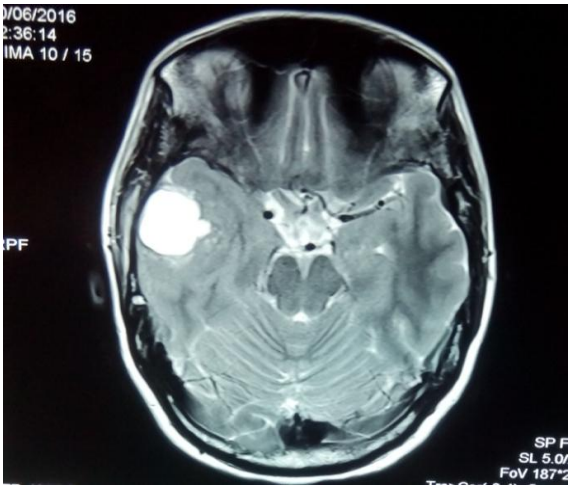

Fig1. MRI brain axial T2 weighted image showing extra axial mass 3.0 X 2.2 X 3.5 cm in right temporal region.

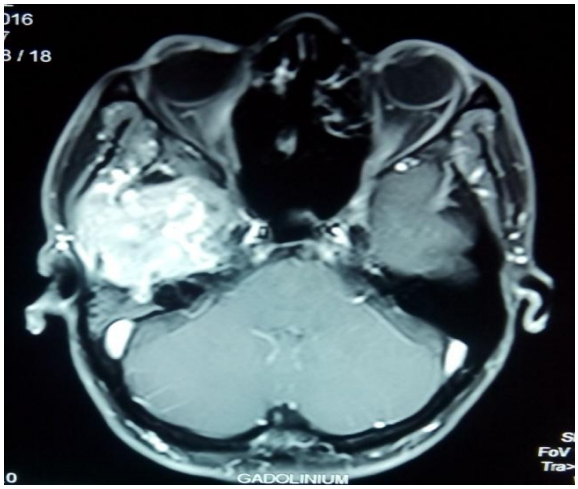

Fig.2. MRI brain contrast showing enhancement of right temporal extra-axial region with involvement of adjacent structures.

Right fronto-temporal craniotomy was done under general anaesthesia. Soft, brownish mass with variegated density with invasion of squamous part of temporal bone was noted. (Fig 3). Dura was not breached. Complete excision of tumor with adjacent bone was done. Skull base defect was reconstructed with pedicled temporalis muscle graft. Postoperatively patient was symptom free with no focal neurological deficits. Histopathological examination confirmed the diagnosis of giant cell tumour. CT scan after 3 months showed no recurrence and 6 month follow up patient was symptom free.

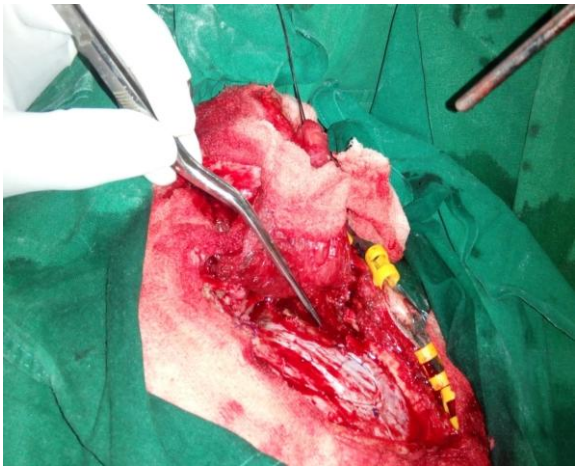

Fig.3. Intraoperative photogarph showing tumour.

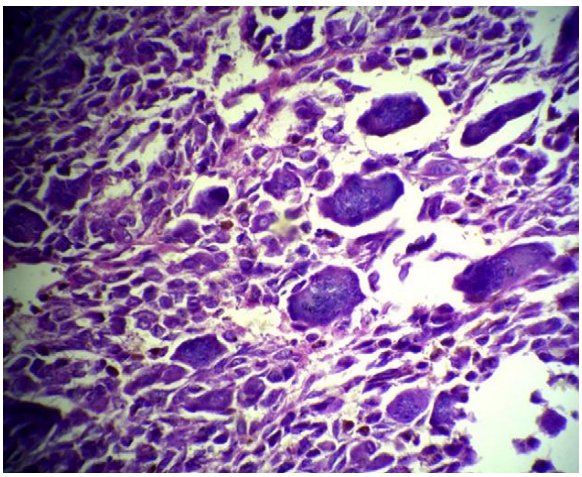

Fig. 4. Histolgy showing spatially dispersed multinucleated giant cells with eosinophilic cytoplasm with vesicular nuclei suggestive of giant cell tumour.

DISCUSSION

Giant cell tumors (also known as osteoclastomas) are thought to be derived from monocytic/histiocytic cells of the hematopoetic system. [(7)] They have often been confused with nonosseous fibroma, chondroblastoma, chondromyxoid fibroma, unicameral bone cyst with a cellular lining, giant cell (reparative) granuloma, aneurysmal bone cyst, brown tumor of hyperparathyroidism, and giant cell containing osteosarcoma. [8] Recent

experiments have characterized these lesions as consisting of three cell types: osteoclast-like multinucleated giant cells; round mononuclear cells resembling monocytes; and a spindle-shaped, fibroblast-like stromal cell. [9]

These experiments further suggested that the stromal cells secrete various monocyte chemoattractants that stimulate monocyte migration to bone and their subsequent fusion into osteoclast-like, multinucleated giant cells. Thus, the stromal cell component may actually be of neoplastic origin, with the multinucleated giant cells being a reactive component. [10] Giant cell tumor account for 5-9 % of all primary bone tumors. (11) The relative absence in cranial bone is not surprising as less than 25 cases of giant cell tumour of temporal bone has been reported in world literature. (10,11) Commonest sites of Giant cell tumors are epiphyses of long bones. The skull is rare location for Giant cell tumor. In cranium, sphenoid bone is commonest site followed by temporal bone. (11) Most commonly seen in 25 to 40 years of age with female preponderance.

They are locally aggressive and present with slowly progressive pain, hearing loss and cranial nerve paralysis.Treatment options and prognosis are mainly derived from the literature on tumors in long bones. Surgical excision is generally the treatment of choice, [12] with recurrence rates correlated to extent of surgical resection. [13] Prognosis is mainly related to extent of surgical excision, with little contribution from radiographic and histologic grading systems. [12,13] Chemical or physical agents such as phenol, liquid nitrogen, or methylmethacrylate have been used to augment the curettage or excision. [13] There is little evidence for a role for chemotherapy. [13] The treatment role of radiation therapy is controversial. While radiation has been used when complete surgical resection is impossible, there is evidence that irradiation predisposes the tumor to subsequent sarcomatous degeneration. [12,13] Some of the mononuclear cells and stormed cells in GCT express receptor activator of nuclear factor. K- β ligand (RANKL) inhibition of this eliminate giant cells. It may be useful for unresecteble tumor of skull. The rate of recurrence is about 40 to 60% after surgery. In 5 to 10 % of cases tumors are malignant. (12, 13)

CONCLUSION

Thus, to conclude surgical excision is the treatment of choice, and irradiation may predispose to sarcomatous degeneration. Radiographic and histologic grading systems do not predict clinical outcome, and extent of surgical resection has been shown to be predictive of prognosis.

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How to cite this article: Supreeth CS, Mahantahshetti PS, Mahantahshetti SS. Giant cell tumor of temporal bone: a rare case. Int J Health Sci Res. 2017; 7(2):327-330.

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