Case Report

Base of the skull giant cell tumor: Rare presenting features of a rare tumor

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ABSTRACT

An 18-years old girl presented with progressive diminution of vision and trismus for 3 months. On examination, light perception was absent in the right eye along with decreased vision and lateral ophthalmoplegia in the left eye and Grade 2 trismus. On magnetic resonance image, a large mass, isointense on T1 weighted image with contrast enhancement, hyperintense on T2 weighted image, was seen to be arising from the base of the skull with the surrounding bone and soft tissue destruction. She was initially treated with partial decompression of mass. Histopathological examination showed giant cell tumor. Patient was then treated with radiotherapy. The case is described along with a review of the literature.

Key words: Base of skull, giant cell tumor, radiotherapy

INTRODUCTION

Giant cell tumor (GCT) of bone is classified as benign primary bone neoplasm. It represents 4-9.5% of all bone tumors and 20% of benign bone tumors. GCT usually occurs in 20-40 years of age-group with a slightly higher predominance in woman. The tumor is most frequently identified in long bones. It rarely manifests in skull. GCT of the cranium represents only 1% of all GCT.[1] When these lesions are encountered in the skull, they preferentially involve sphenoid and temporal bones. The tumor is initially treated with surgery. But it may locally recur after surgery depending upon the status of margin. Hence, adjuvant radiotherapy is often given. Metastases to lungs have been reported. In rare instances, GCT may transform into sarcoma. We report a case of GCT in the base of the skull in an 18-years old girl with the loss of vision, ophthalmoplegia, trismus and atypical imaging features.

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CASE REPORT

An 18-years old girl presented with the complaint of headache for 3 months, progressive diminution of vision for the same duration and trismus for 1 month. There was no history of vomiting, convulsion. Physical examination revealed the absence of light perception in the right eye. She was able to count fingers at 1 m distance by the left eye. Lateral ophthalmoplegia was present in the left eye. Ophthalmoscopy did not show any abnormality. Higher mental function was normal. Motor, sensory, and autonomic functions were preserved. Mouth opening was up to 2 cm.

Magnetic resonance image (MRI) showed a large lobulated mass at the midline of base of the skull with destruction of sphenoid body, clivus, part of greater and lesser wing and pterygoid plate; encasement of bilateral cavernous sinus and internal carotid arteries. Pituitary and nasopharynx were normal. The lesion was isointense on T1 with contrast enhancement;[Figure 1]; it was hyperintense on T2. Radiologically, it was suggestive of chordoma. The patient was initially treated with fronto-temporal craniotomy and partial decompression of mass.

Histopathological examination revealed large multinucleated giant cells that were uniformly dispersed among oval or spindle-shaped stromal cells. Giant cells displayed nucleoli that were similar to the surrounding spindle cells, suggestive of osteoclastic type, which was

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consistent with GCT. Histological diagnosis was GCT. [Figure 2].

Post-operative computed tomography (CT) scan of the brain showed progressive disease. The patient's vision in the left eye further worsened; she ultimately loose vision in both eyes. She also developed significant pain and headache. In view of residual disease, conformal external beam radiotherapy (54 Gy in 30 fractions) was delivered. The patient tolerated radiotherapy well, with good relief of pain and headache.

DISCUSSION

Giant cell tumor accounts for approximately 20% of benign bony tumors. This tumour develops by endochondral ossification. Only 1% of GCTs occur in the skull, with the most common cranial sites being sphenoid and temporal bones. The fact that sphenoid and petromastoid portion of the temporal bone arise from endochondral ossification in the skull can explain why GCTs in the skull are mostly found in sphenoid and temporal bone.[2]

The tumors typically occur in third and fourth decade.[3] This is in contrast with our patient, who was 18 years of age. This case was locally very aggressive, which is also quite uncommon for a GCT.

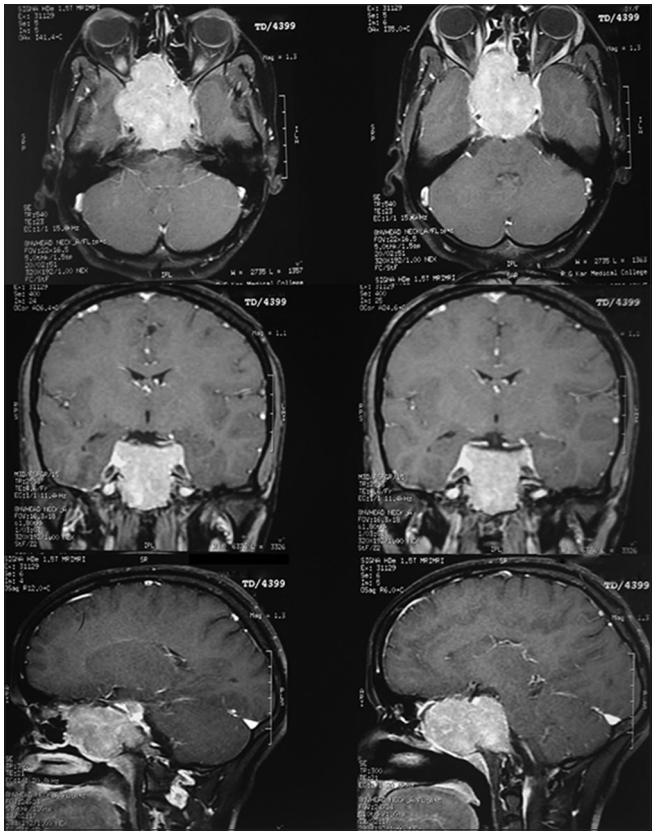

Figure 1: Magnetic resonance image

Compared to GCT of long bones, GCT of skull lacks the characteristic signs of expansion and "'soap-bubble"' appearance.[4] On CT scan, GCTs are typically seen as soft tissue specification with osteolytic lesions causing bone erosion and sharp margins. MRI of such tumors demonstrate isointensity on T1 weighted image and low signal intensity in T2 and in diffusion-weighted imaging. The mass may show mild contrast enhancement on T1. However in our case, the lesion shows contrast enhancement on gadolinium enhanced T1 weighted image [Figure 1] and is hyperintense on T2.

Histologically, GCT consists of three cell types; osteoclast-like multinucleated giant cells, round mononuclear cells and spindle-shaped, fibroblast-like mononuclear cells.[5]

Surgery should be performed as complete as possible in GCT.[6] But in cranial GCT, it is often difficult to perform complete excision. Hence, radiotherapy is often necessary after incomplete resection of the mass. A series of cases reported by Bertoni *et al*. used the treatment strategies involving surgical resection and radiotherapy.[7] However, the use of radiotherapy in completely resected tumor is not clear.

There is no well-defined accepted protocol for treatment with chemotherapy.[8] Though in some cases, chemotherapy has been given as adjuvant therapy with good result.[9]

Giant cell tumor generally recurs within first 3 years. But follow-up until 5 years is recommended, since late distant metastasis has been reported.

CONCLUSION

Giant cell tumor arising from the base of the skull may present with atypical symptoms, such as trismus and as well as with atypical radiological findings. Although most of the time it is slow growing in nature, but rare aggressive natural

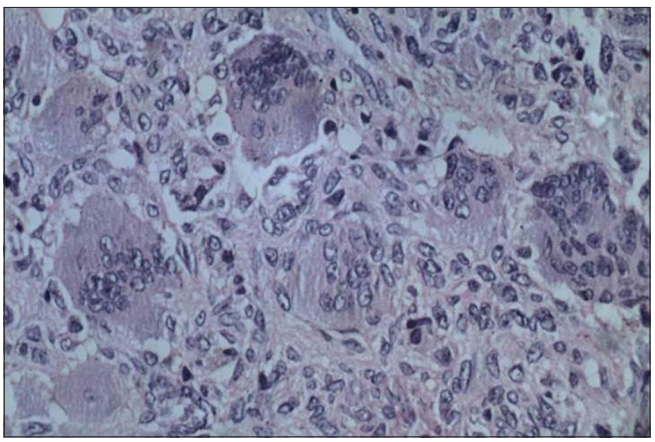

Figure 2: Microscopical image

history should also be kept in mind during treatment and follow-up.

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