Case Report

**Left orbital roof giant cell tumor of bone: A case report**

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**Abstract**

**Background:** Giant cell tumor of bone originating from the connective tissue within the bone marrow is benign but locally aggressive lesion. In all, 90% of the cases involve the epiphysis of long bones and less than 2% involve the skull. Giant cell tumors of the skull occur most frequently in the sphenoid and temporal bones, and very rarely in the ethmoid, frontal, parietal, and occipital bones. We would like to share a case of giant cell tumor of bone arising from the left orbital roof with involving ethmoid sinus, which was diagnosed to be a meningioma before surgery.

**Case Description:** A 32-year-old lady presented to us with the chief complain of left proptosis, diplopia, and left eye soreness without decline of visual acuity for about 2 months. Her orbital magnetic resonance imaging (MRI) disclosed a mass lesion located in the left frontal base, orbital roof, and upper medial orbital region with adjacent dural-tail sign favoring meningioma. She underwent a left supraorbital pterional craniotomy with the gross total removal of tumor and dura reconstruction. Histology examination of the tumor showed a picture of giant cell tumor of bone. Considering giant cell tumor of bone is locally aggressive, postoperative adjuvant therapy with Denosumab was introduced after full explanation.

**Conclusion:** Standard treatments of skull-base giant cell tumors have yet to be established due to small number of cases reported in the literature. The standard treatment of giant cell tumor of bone is complete resection of the tumor.

**Key Words:** Anti-RANKL monoclonal antibody, giant cell tumor of bone, orbital roof tumor

**INTRODUCTION**

Giant cell tumor of bone was described by Cooper and Travers in 1818, which is characterized histologically by multinucleated giant cells with a background of mononuclear stromal cells. Giant cell tumor of bone originating from the connective tissue within the bone marrow is benign but locally aggressive with high recurrent rate after treatment. Giant cell tumor of bone accounts for about 3% to 7% of primary bone tumors. In all, 90% of the cases involve the epiphysis of long bones and less than 2% involve the skull. Giant cell tumors of the skull occur most frequently in the sphenoid and temporal bones, and very rarely in the ethmoid, frontal, parietal, and occipital bones.

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the sphenoid and temporal bones, and very rarely in the ethmoid, frontal, parietal, and occipital bones.\(^\text{[1,4,6,8]}\) The first case of giant cell tumor of the orbit reported in the English literature was published in 1993.\(^\text{[12]}\) We would like to share a case of giant cell tumor of bone arising from the left orbital roof with involving ethmoid sinus, which was diagnosed to be a meningioma before surgery.

**CASE REPORT**

In March 2017, a 32-year-old woman without any systemic disease or ocular traumatic event presented to us with the chief complaint of left proptosis, left eye soreness, and diplopia on upper right gaze without decline of visual acuity since January 2017. Noncontrast computed tomography (CT) of orbit done at an outside hospital showed a left intra-orbital tumor with intracranial invasion. Orbital magnetic resonance imaging (MRI) was arranged and it disclosed a mass lesion about 3.8 cm × 3.7 cm × 3.3 cm in size, located in the left frontal base, orbital roof, and upper medial orbital region. This mass lesion showed relative intermediate intensity on T1-weighted image and T2-weighted image, and the postcontrast study showed good enhancement with adjacent dural tail sign causing mass effect on the left eyeball as well as the left frontal brain parenchyma, which favored meningioma [Figure 1]. On admission, her neurological examination showed impairment of the left eye ball movement to upward gaze and medial gaze, otherwise essentially negative finding. Under general anesthesia, she was put in the supine position and underwent a left supraorbital pterional craniotomy with the gross total removal of tumor and dura reconstruction. Grossly, the tumor was hypervascular, relatively firm in consistency, which destructed the left orbital roof thoroughly and invaded the dura of the left frontal base causing the compression of the left frontal lobe and displacement of the left eye ball. This tumor also extended to ethmoid sinus. Histology examination showed that the tumor consisted of evenly distributed osteoclast-like giant cells in a background of round or spindle-shaped mononuclear cells. By immunohistochemistry, the tumor cells showed CD68(+), GFAP(−), EMA(−), p63(−), S100(+scattered), CD1a(+scattered), and p53(−) [Figure 2]. Proliferation index was about 6% by Ki-67 immunostain. Giant cell tumor of bone was diagnosed based on the morphology of the tumor cells and the result of immunohistochemical stains. The resected left orbital roof remnant and diseased dura showed focal involvement by the tumor. The patient’s postoperative course was uneventful. After surgery, her left proptosis and limitation of the left eye ball movement were resolved. The postoperative orbital MRI showed some postoperative change without definite residual tumor [Figure 3]. Considering giant cell tumor of bone is locally aggressive with high recurrent rate, postoperative adjuvant therapy with Denosumab was introduced after full explanation. She is doing well and is undergoing regular follow-up at our outpatient department.

**DISCUSSION**

Giant cell tumor of bone is an aggressive, bone lytic, osteoclastogenic stromal tumor.\(^\text{[6]}\) Lung metastasis and malignant transformation to high grade osteosarcoma have been reported although rare.\(^\text{[1,2,7,11]}\) The majority of

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**Figure 1:** Preoperative orbit MRI. Coronal T1 weighted image (a), coronal T1 weighted image post gadolinium enhancement (b), coronal T2 weighted image (c), and sagittal T1 weighted image post gadolinium enhancement (d) showing a well-circumscribed lesion isointensity on T1-weighted images and hypointensity on T2-weighted image with good enhancement and has small cystic/necrotic change noted in the left frontal base, orbital roof, and upper-medial orbital region. The black arrows point out the dural tail sign.

**Figure 2:** Histology of the specimen. Orbital roof HE stain 200X (a), Dura HE stain 100X (b), Tumor HE stain 200X (c) showing osteoclast-like giant cells in a background of round or spindle-shaped mononuclear cells. The resected orbital roof and dura had been involved by the tumor. Tumor CD68 stain 100X (d) showing positive staining. CD68 is particularly useful as a marker for giant cells, osteoclasts.
of giant cell tumor of bone by the anti-RANKL monoclonal antibody “Denosumab,” is an additional adjuvant therapeutic option.\textsuperscript{[1,2,10]} Recently, Denosumab has been reported to provide promising therapeutic effect on giant cell tumor of bone in cases of inoperable or locally advanced situation.\textsuperscript{[1,2,10]} Nevertheless, the long-term effect and the duration of treatment of Denosumab on giant cell tumor of bone and the safety of Denosumab need further clinical evaluation and basic research.\textsuperscript{[5,11]}

**CONCLUSION**

Orbitofrontal giant cell tumor of bone is very rare. To our best knowledge, only three cases had been reported in the English literature [Table 1]. Total surgical resection is the treatment of choice. The dural tail sign occurs as a result of thickening and enhancement of the dura and is most often seen adjacent to a meningioma, but interestingly, it was present in our patient who was confirmed to be a case of orbital roof giant cell tumor of bone.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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**Table 1: Cases of orbitofrontal giant cell tumor of bone reported in the English literature**

| Authors year of report | Age/sex of patient | Therapy | Tumor recurrence | Duration of follow-up |
|------------------------|--------------------|---------|-----------------|----------------------|
| Vernet et al. 1993\textsuperscript{[2]} | 10-year-old male | Gross total resection | Not reported | Not available |
| Kamoshima et al. 2011\textsuperscript{[8]} | 2-year-old female | Total resection | No recurrence | 18 months |
| Tang et al. 2017\textsuperscript{[8]} | 10-year-old male | Gross total resection | No recurrence | 4 months |

**Figure 3: Post-operative orbit MRI.** Coronal T1 weighted image (a), coronal T1 weighted image post gadolinium enhancement (b), coronal T2 weighted image (c) showing gross total removal of the tumor.
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