SNI
SURGICAL NEUROLOGY INTERNATIONAL
SNI: Unique Case Observations
OPEN ACCESS
For entire Editorial Board visit : http://www.surgicalneurologyint.com
Editor: Ather Enam, M.D., Aga Khan University, Karachi, Sindh, Pakistan

Case Report

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

Chi-Man Yip[1], Huai-Pao Lee[2,3], Shu-Shong Hsu[1], Ying Tso Chen[1]

[1]Division of Neurosurgery, [2]Department of Pathology and Laboratory Medicine, Kaohsiung Veterans General Hospital, Kaohsiung, [3]Department of Nursing, Meiho University, Taiwan

E-mail: Chi-Man Yip - yip_chiman@yahoo.com; Huai-Pao Lee - hplee0627@vghks.gov.tw; Shu-Shong Hsu - sshsu@vghks.gov.tw;
*Ying Tso Chen - a7513366@yahoo.com.tw
*Corresponding author

Received: 06 December 17 Accepted: 23 May 18 Published: 26 June 18

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

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_467_17
Quick Response Code:

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.[4] 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.[1,3,8] 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.[1,6,8] Giant cell tumors of the skull occur most frequently in

How to cite this article: Yip CM, Lee HP, Hsu SS, Chen YT. Left orbital roof giant cell tumor of bone: A case report. Surg Neurol Int 2018;9:127.
http://surgicalneurologyint.com/Left-orbital-roof-giant-cell-tumor-of-bone:-A-case-report/

the sphenoid and temporal bones, and very rarely in the ethmoid, frontal, parietal, and occipital bones.[1,6,8] The first case of giant cell tumor of the orbit reported in the English literature was published in 1993.[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 complain 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.

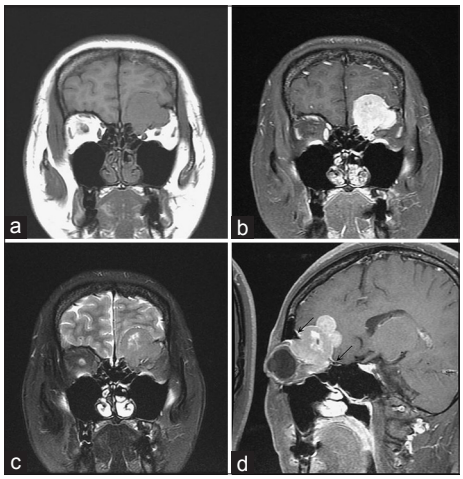

Figure 1: Preoperative orbit MRI. Coronal T1 weighted image (a), coronal T1 weighted image post gadolinium enhancement (b), coronal T2 weighted image (c), and sagittal T1 weighted image post gadolinium enhancement (d) showing a well-circumscribed lesion isointensity on T1-weighted images and hypointensity 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

DISCUSSION

Giant cell tumor of bone is an aggressive, bone lytic, osteoclastogenic stromal tumor.[6] Lung metastasis and malignant transformation to high grade osteosarcoma have been reported although rare.[1,2,7,8,11] The majority of

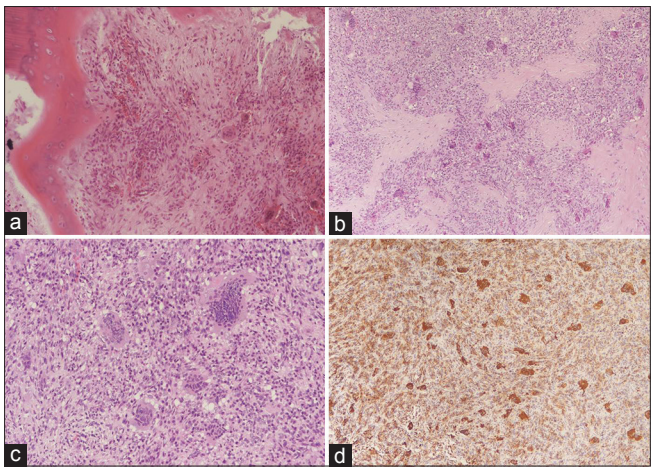

Figure 2: Histology of the specimen. Orbital roof HE stain 200X (a), Dura HE stain 100X (b), Tumor HE stain 200X (c) showing osteoclast-like giant cells in a background of round or spindle-shaped mononuclear cells. The resected orbital roof and dura had been involved by the tumor. Tumor CD68 stain 100X (d) showing positive staining. CD68 is particularly useful as a marker for giant cells, osteoclasts

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[12]	10-year-old male	Gross total resection	Not reported	Not available
Kamoshima *et al*. 2011[8]	2-year-old female	Total resection	No recurrence	18 months
Tang *et al*. 2017[9]	10-year-old male	Gross total resection	No recurrence	4 months

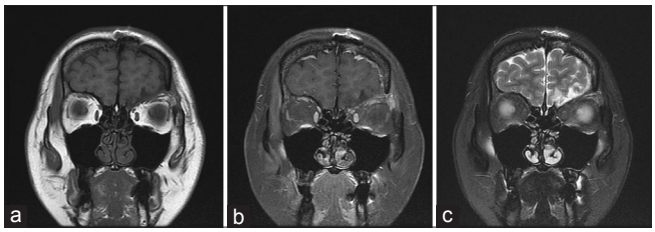

Figure 3: Post-operative orbit MRI. Coronal T1 weighted image (a), coronal T1 weighted image post gadolinium enhancement (b), coronal T2 weighted image (c) showing gross total removal of the tumor

cases develop at the epiphyses of long bones, but may occur in the sacrum, vertebral body and occasionally in the small bones of the hands and feet.[2,7] Less than 2% of the cases involve the skull.[8] The radiographic findings of giant cell tumor of bone on CT scan are osteolytic, radiolucent, and well-circumscribed.[8] The MRI features of giant cell tumor of bone are nonspecific, usually demonstrating a well-circumscribed lesion with low to intermediate signal intensity or isointense on T1-weighted images; intermediate to high intensity or hypointense on T2-weighted images; and homogenous enhancement after gadolinium injection.[8-11] However, there are no MRI features sufficiently characteristic to allow a preoperative diagnosis of giant cell tumor of bone especially those in the rare locations.[8,12] Due to the small number of skull-base giant cell tumors reported in the literature, standard treatments have yet to be established.[6] Total surgical resection is the treatment of choice of giant cell tumor of bone.[1,6,7,12] However, in some instances, radical resection is not feasible when the tumor involves the skull base. Chemotherapy may be beneficial for incompletely resected tumors.[8] Adjuvant radiation therapy is controversial because giant cell tumor of bone is not radiosensitive and irradiation may predispose the tumor to subsequent malignant transformation.[1,7,8,12] However, radiotherapy with the dosage around 40 to 60 Gy is still recommended as a postoperative adjuvant therapy in cases of incompletely resected skull base tumor.[6] Histologically, giant cell tumor of bone consists of reactive multinuclear osteoclast-like giant cells expressing receptor activator of nuclear factor kappa B (RANK) and neoplastic mononuclear stromal cells expressing receptor activator of nuclear factor kappa B ligand (RANKL).[1,2,10] Blocking of the receptor activator of NF-kappa B ligand (RANKL) signaling pathway, which plays a role in the pathogenesis of giant cell tumor of bone by the anti-RANKL monoclonal antibody "Denosumab," is an additional adjuvant therapeutic option.[1,2,5,10] Recently, Denosumab has been reported to provide promising therapeutic effect on giant cell tumor of bone in cases of inoperable or locally advanced situation.[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.[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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Bardakhchyan S, Kager L, Danielyan S, Avagyan A, Karamyan N, Vardevanyan H, *et al*. Denosumab treatment for progressive skull base giant cell tumor of bone in a 14 year old female- a case report and literature review. Ital J Pediatr 2017;43:32.
2. Borkowska A, Goryn T, Pienkowski A, Wagrodzki M, Jagiello-Wieczorek E, Rogala P, *et al*. Denosumab treatment of inoperable or locally advanced giant cell tumor of bone. Oncol Lett 2016;12:4312-8.
3. Chakarun CF, Forrester DM, Gottsegen CF, Patel DB, White EA, Matcuk GR. Giant Cell Tumor of Bone: Review, Mimics, and New Developments in Treatment. Radiographics 2013;33:197-211.

4. Cooper AS, Travers B. Surgical essays. London, England: Cox Longman and Co; 1818. pp 178-9.
5. Gaston CL, Grimer RJ, Parry M, Stacchiotti S, Tos APD, Gelderblom H, *et al*. Current status and unanswered questions on the use of Denosumab in giant cell tumor of bone. Clin Sarcoma Res 2016;6:15.
6. Goto Y, Furuno Y, Kawabe T, Ohwada K, Tatsuzawa K, Sasajima H, *et al*. Treatment of a skull-base giant cell tumor with endoscopic endonasal resection and denosumab: Case report. J Neurosurg 2017;126:431-4.
7. Harris AE, Beckner ME, Barnes L, Kassam A, Horowitz M. Giant cell tumor of the skull: A case report and review of the literature. Surg Neurol 2004;61:274-7.
8. Kamoshima Y, Sawamura Y, Imai T, Furukawa H, Kubota K, Houkin K. Giant Cell Tumor of the Frontal Bone in a Girl- Case Report. Neurol Med Chir (Tokyo) 2011;51:798-800.
9. Tang PH, Mettu P, Maltry AC, Harrison AR, Mokhtarzadeh A. Giant Cell Tumor of the Frontal Bone Presenting as an Orbital Mass. Ophthalmol Ther 2017;6:215-20.
10. Van der Heijden L, Dijkstra S, Blay JY, Gelderblom H. Giant cell tumour of bone in the denosumab era. Eur J Cancer 2017;77:75-83.
11. Van der Heijden L, Dijkstra S, Van de Sande MAJ, Kroep JR, Nout RA, Van Rijswijk CSP, *et al*. The Clinical Approach Toward Giant Cell Tumor of Bone. Oncologist 2014;19:550-61.
12. Vernet O, Ducrey N, Deruaz JP, Tribolet ND. Giant Cell Tumor of the Orbit. Neurosurgery 1993;32:848-51.