Giant cell rich Osteosarcoma of the skull – Case report

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ABSTRACT

Giant cell rich osteosarcoma account for 3% of all osteosarcomas and often arise in the appendicular skeleton Giant cell rich Osteosarcoma of the skull is an extremely rare manifestation.

Case Presentation: Here we present a 15 year old girl with Giant cell rich osteosarcoma of the skull. Our patient presented with holocranial headache, diplopia and bilateral fundus atrophy. MRI brain revealed a heterogenous soft tissue density in sphenoid sinus with destruction of roof of sphenoid sinus and extending to posterior ethmoid sinus. Initial transnasal biopsy was suggestive of Giant cell tumor. Hence she underwent decompression of the sphenoclival mass. Subsequent histopathology confirmed Giant cell rich variant of Osteosarcoma. She received External beam Radiation followed by six cycles of chemotherapy with Cisplatin, Adriamycin and Ifosfamide. Currently she is asymptomatic and in clinical remission at 1 year of follow up.

A close differential diagnosis of this rare entity is Giant cell tumor of bone. Likewise in our patient the preoperative cytology resembled Giant cell tumor of bone. Hence she underwent upfront surgery. Post operative histopathology was in favour of Giant cell rich osteosarcoma. The standard approach to the treatment of Giant cell rich osteosarcoma is not yet well established. However the available literature suggests a multidisciplinary approach as that for high grade osteosarcoma. Nevertheless further studies are required to determine the appropriate management of this rare bone tumor.

Key words: Giant Cell, Osteosarcoma, Skull

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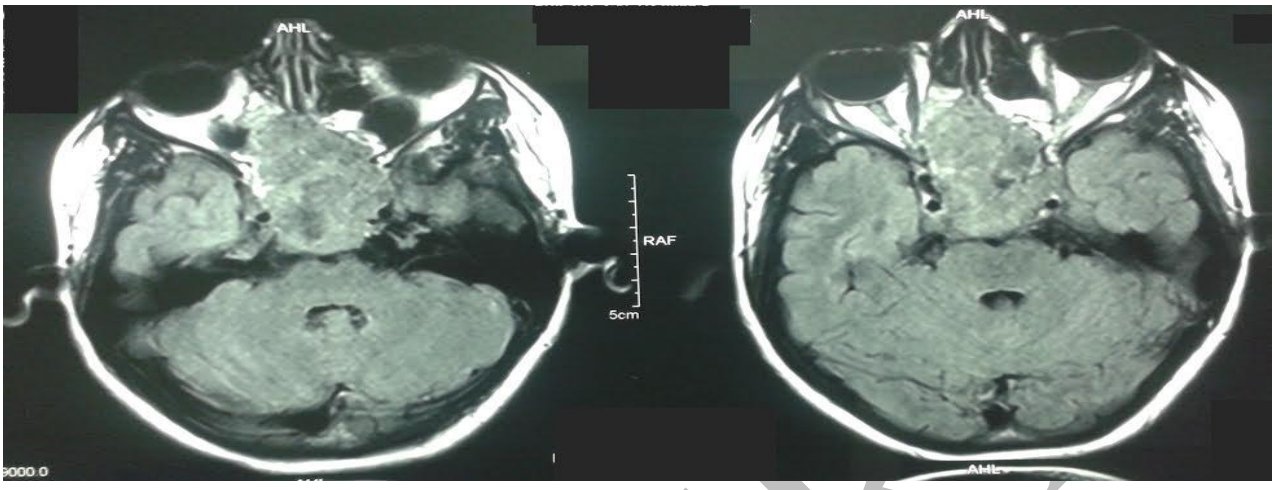
Conflict of interest: None

INTRODUCTION: Giant cell rich Osteosarcoma is a very rare entity and typically described in appendicular skeleton. [1] There are very few case reports of Giant cell rich Osteosarcoma described in craniofacial bone. They lack the characteristic clinical and radiological features of Conventional high grade osteosarcoma. The close pathological differential diagnosis is Giant cell tumor of bone and Malignant fibrous histiocytoma. Giant cell rich Osteosarcoma is considered as a high grade osteosarcoma. Taking into consideration the aggressiveness of this unique histology and different treatment strategy the right pathological diagnosis is crucial. Here we report a patient with Giant cell rich osteosarcoma involving the sphenoclival bone.

CASE HISTORY

A 15 year old girl presented with holocranial headache of 3 months duration and diplopia on looking towards right and for far objects of 1 month duration. Neurological examination was normal except for mild restriction of abduction of eye towards right. Fundus showed bilateral optic atrophy. Rest of the system examination was within normal limits.

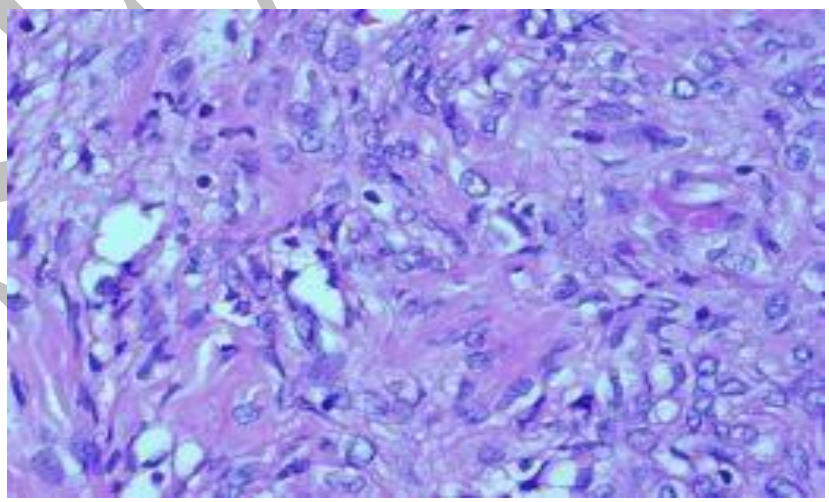
MRI brain revealed a well defined lesion 4.6 *3.5 *3 cm seen filling and expanding the sphenoid sinus with destruction of the roof and extending to posterior ethmoid sinus and also involving the upper margin of clivus. There was no calcification or periosteal reaction.



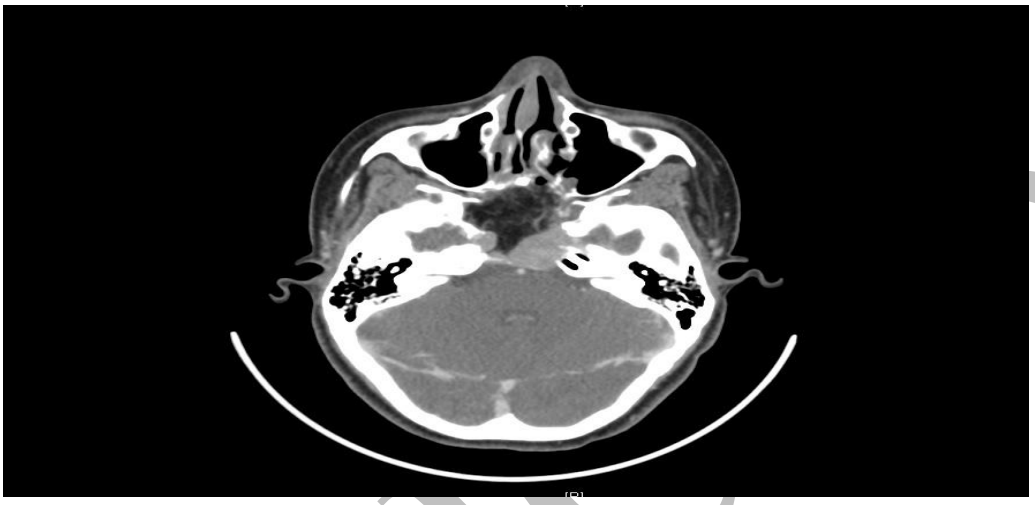
Computed Tomography of Chest and Bone scan were normal. Transnasal biopsy was done that suggested Giant cell tumor.

She underwent decompression of the sphenoclival mass through transbasal approach. Her postoperative period was uneventful. She remained afebrile, ambulant and did not develop any new neurological deficit.

Histopathology revealed a giant cell rich lesion with marked atypia and atypical mitosis suggestive of Giant cell rich Osteosarcoma. MIB 1 labelling index was high in few fragments and P63 negative.



Post operative CT imaging showed irregular heterogeneously enhancing soft tissue involving the residual superior, posterior, lateral and inferior walls of sphenoid sinus suggestive of residual tumor. Visualised brain and rest of the bones reveal no focal lesions.



She received Conventional External beam Radiotherapy with 50 Gy in 25 fractions. Subsequently systemic chemotherapy with 6 cycles of IFosfamide 1.4 g/m2, Cisplatin 100 mg/m2, and Adriamycin 60 mg/m2 every 21 days was administered with prophylactic G CSF support.

She is asymptomatic after a period of 1 year of completing treatment.

DISCUSSION

Osteosarcomas are the most common primary bone tumor and are aggressive malignant neoplasms. Most commonly it affects the appendicular skeleton. Among all osteosarcomas 6 to 13% tend to affect the head and neck region. Among the head and neck tumors osteosarcomas constitute only 0.5%. [1]

Several histological variants of Osteosarcoma have been described. Giant cell rich osteosarcoma is an extremely rare variant of osteosarcoma first described by Bathurst and Sanekin. [2] It accounts for 3% of all osteosarcomas and typically affects the long bones. Giant cell rich osteosarcomas of the head and neck are extremely rare in

literature. To the best our knowledge only few case reports of Giant cell rich osteosaroma in the maxilla could be found. [3] Giant cell rich osteosarcoma affecting the cranial vault has not been reported.

The classical radiological features of Osteosarcoma are absent in case of Giant cell rich Osteosarcoma. They occur predominantly as lytic lesions in the appendicular skeleton. Usually the periosteal reaction is scanty with minimal soft tissue involvment. [2]

Histologically it is difficult to distinguish it from other giant cell rich tumors of bone such as Malignant Giant cell tumor bone and Malignant fibrous histiocytoma. [5] The characteristic microscopic appearance of giant cell rich osteosarcoma is non-uniform distribution of giant cells, atypical mitosis and presence of malignant osteoid. However malignant giant cell tumor microscopically shows uniform distribution of giant cells with variable mitosis and characteristically absent osteoid. Malignant fibrous histiocytoma contains small foci of osteoid and malignant giant cells. However the basic proliferative component is a fibrohistiocytic cell with storiform pattern that aids in diagnosis. [7]

Giant cell rich osteosarcoma and Malignant giant cell tumor show similar clinical presentation and radiological appearance. Giant cell tumor with malignant transformation occurs in 3rd to 4th decade of life. Osteosarcoma occurs in adolescent skeleton. Both these tumors are classically described in appendicular skeleton. The histological differentiation is crucial because of vast difference in prognosis and management. [6] Giant cell tumor with malignant transformation show increased propensity for local recurrence and lesser incidence of metastasis. It is managed with local curettage and cementing. While Giant cell rich osteosarcoma is treated like high grade osteosarcoma. [5] [6].

Gnathic osteosarcomas tend to occur in older age group and have a less aggressive clinical behaviour. They have a predilectation to involve maxilla and mandible. The treatment of choice is radical resection with clear margins. Bone sarcomas of head and neck are difficult to resect. Incomplete resection result in increased

incidence of local failure and poor outcome. [10]. The role of adjuvant radiotherapy and chemotherapy is doubtful. [4] Unlike craniofacial osteosarcoma, giant cell rich osteosarcoma behaves more aggressively.

There is no clear consensus regarding the management of this rare entity. Due to lack of specific data, the treatment of craniofacial osteosarcoma is extrapolated from that of Extragnathic OS. A retrospective analysis of 49 patients over a 28 year period in patients with craniofacial osteosarcoma showed that multidisciplinary treatment resulted in long-term survival in over two thirds of patients. Extragnathic sites and failure to achieve and maintain local surgical control emerged as strong negative prognostic factors. [11]. The role of neoadjuvant chemotherapy is controversial. In a retrospective analysis of 111 patients with Osteosarcoma of mandible neoadjuvant chemotherapy improved disease-free and metastatic-free survival and increased clear margins rates from 50% to 68%.[13]. Similarly the role of adjuvant radiotherapy and chemotherapy are also not well defined. Adjuvant radiotherapy is usually recommended for incomplete surgical resections. [14].Adjuvant chemotherapy was administered in studies to patients with poor prognostic factors including bulky tumor and marginal resection. In most of these studies patients receiving post operative chemotherapy achieved higher local control rates. However the ideal drug combination remains to be defined, although the most common seems to be doxorubicin and cisplatin-based protocols.[13]

CONCLUSION

Our patient has presented with a rare entity that has occurred at a rare site. In addition to the rare clinical presentation the challenges in establishing both radiological and histological diagnosis and controversies in treatment and prognosis are the reasons for presenting this case report. It is important to recognize this variant, as its prognosis is worse as compared to Gnathic osteosarcomas. The close histologic resemblance of this entity to giant cell tumor of bone makes the diagnosis a challenge as they are managed entirely different. The role of adjuvant radiotherapy and chemotherapy in this rare variant remains to be defined.

However considering the aggressive behaviour and difficulty to attain clear margins with radical surgery in Giant cell rich Osteosarcoma of the cranial vault adjuvant radiotherapy and chemotherapy may have a role. [13] [14]. Overall the optimal management of high grade osteosarcomas of skull remains to be defined. Our patient was initially mistaken for giant cell tumor of bone and underwent upfront surgery. She had an incomplete resection and so received adjuvant chemotherapy and radiotherapy. She is alive at one year post completion of treatment.

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