A CASE REPORT OF OSTEOSARCOMA OF THE CRANIAL VAULT IN A PEDIATRIC PATIENT

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INTRODUCTION
Osteosarcoma is an aggressive malignancy of primitive mesenchymal origin. Although it is the most common primary bone tumor of all ages, it accounts for only 3-5% of all childhood malignancies. Pediatric osteosarcomas most commonly arise in the appendicular skeleton with 85% occurring in the metaphysis of long bones. In axial skeleton the pelvis is the most common site.

Craniofacial osteosarcomas are rare. They typically present in the 3rd or 4th decade of life, account for <5% of osteosarcomas in children and only 1%of all pediatric head and neck malignancies. The most craniofacial sites are the mandible and maxilla followed by calvaria and then the skull base.

Approximately 50% of all osteosarcomas occur in the context of underlying conditions which may account for the rarity of pediatric cases.

We report a case of cranial vault osteosarcoma in a 12 year old girl.

CASE REPORT
A 12 year old girl presented to our OPD with a history of a painless swelling over left fronto-temporal region since last 1 year which had started increasing since last 2 months. The patient denied any other symptoms and was neurologically intact. Patient had a 5x5 cm firm, well defined, non mobile, non tender swelling extending over the left fronto-temporal region of the cranial vault without any overlying skin changes.

CT scan with bony window was done which revealed a 51x49x32 mm sized well defined exophytic bone forming sclerotic lesion giving sun burst appearance in few areas arising from the outer table of squamous part of left temporal and frontal bone. Chest X Ray was done which was normal and did not show any lesions.

A left fronto-temporal craniectomy was performed with en bloc excision of mass and a 1cm of normal appearing bone circumferentially. Intraoperatively the tumor was encapsulated without visible invasion of the underlying dura or overlying scalp. A titanium mesh was placed to cover the bony defect. Postoperatively the patient was clinically stable.

Pathological evaluation of surgical specimen revealed high grade chondroblastic osteosarcoma with margins free of tumor. The patient was referred for further adjuvant therapy.

DISCUSSION
Primary osteosarcoma of the skull is rare with an incidence of approximately 1-2% of all skull tumors. Only fewer than 150 cases have been reported so far. The occurrence of osteosarcoma in the skull peaks in the 3rd decade.

Like osteosarcoma of the extremities, skull osteosarcoma frequently present as a slow growing mass or swelling. Unlike extremity tumors, however skull osteosarcomas are often painless or present with only mild pain. While osteosarcoma of extremities show early metastasis particularly to the lungs or brain, metastasis of skull osteosarcoma is uncommon. However relapse of skull osteosarcoma most often occurs locally.

Most common subtypes include osteoblastic, chondroblastic & fibroblastic. Grading of osteosarcomas can be low, intermediate or high based on degree of cellular atypia and architectural distortion. However there is no significant correlation between the histologic character and prognosis.

CT scan with bony window plays the key role in diagnosis in which bone growth with lytic regions and periosteal remodeling are the most common imaging feature. MRI with contrast is useful for assessing soft tissue involvement. A chest CT & bone scan can be done to look for lung and skeletal metastasis respectively.

Surgical resection is the mainstay of treatment. Complete surgical excision with wide surgical margins have been associated with improved survival. Adjuvant chemotherapy is given especially in cases with high grade tumor or when complete surgical resection is not possible. Adjuvant radiotherapy is given when subtotal or uncertain resection is achieved.

CONCLUSION
As osteosarcoma of skull is a rare disease, correct diagnosis and proper treatment plans are difficult. Therefore this uncommon entity needs to be considered when patients with skull lesions present. Complete surgical resection followed by combined chemoradiation is proved to be the most optimal treatment regimen which can be considered as a primary treatment plan.