A RARE CASE OF ATYPICAL PRIMARY EWING'S SARCOMA OF OCCIPITAL BONE

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ABSTRACT

BACKGROUND

Ewing's Sarcoma is an aggressive malignant neoplasm most frequently manifesting in the second decade of life and accounting for 4% of childhood and adolescent malignancies. These tumours were first described by James Ewing in 1921 as tumours that arise from bone. These osseous lesions have since become infamous for their highly aggressive course with 20% to 30% of patients having evidence of metastasis at the time of diagnosis and an estimated 10-year survival rate of 50%. Metastases to the CNS have most recently been estimated to occur in less than 5% of cases and are usually due to direct extension of an osseous lesion into the extradural space or more rarely through haematogenous spread.

CASE REPORT

In this article, we report a case of 17-year-old boy who presented to the radiology department with complaints of recurrent episodes of headache and vomiting for the past 3 days. On radiological investigation, there was a large well-defined, lobulated, extra-axial mass lesion measuring 3.6 X 5.7 X 5.9 cm noted in the supratentorial left occipital region which was fairly enhancing after contrast administration. The mass was causing permeative type of destruction of the left occipital bone and extending into extracranial soft tissue. Final diagnosis was done by biopsy and histopathology which showed "Atypical Ewing's Sarcoma" of the left occipital bone. Considering its unusual site and soft tissue extension, we report this case of Primary Atypical Ewing's sarcoma of occipital bone.

CONCLUSION

Primary cranial Ewing's sarcoma is to be considered in the differential diagnosis in children with a tumour involving the skull with destruction of the bone and presence of extra-axial soft tissue swelling. CT is the excellent modality for demonstration of bone destruction while MRI depicts soft tissue extension and metastasis if any.

KEYWORDS

Atypical Primary Ewing's Sarcoma, Occipital Bone, Metastatic Ewing's Sarcoma to the Brain, Radiology.

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INTRODUCTION: Ewing's sarcoma is the second most common malignant bone tumour occurring in children and young adults, and accounts for 10–15% of all primary bone tumours.[1] Ewing's sarcoma, along with other similar round cell tumours are called as "Tumours of the Ewing sarcoma family of tumours (ESFT)" and are characterised by morphologically similar round cell neoplasms as well as by the presence of common chromosomal translocation. These (ESFT) include Ewing sarcoma (ES) osseous and extra osseous/soft tissue type.[2,3] A "malignant small-cell tumour of the thoracopulmonary region" (i.e. Askin tumour).,[4] primitive neuroectodermal tumour (PNET).[5] of soft tissues and bone. Initially, ESFT were believed to be biologically distinct.

However, based on their wide spectrum of neural differentiation (PNET being the most differentiated); their immunohistochemical, cytogenetic, and molecular uniformity; and their identical response to Ewing-based chemotherapy regimens, it was determined that these sarcomas are related and that they originate from unique mesenchymal stem cells capable of multilineage differentiation.[6]

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CASE REPORT:

Patient: A 17-year-old boy referred from the department of neurosurgery for the evaluation of recurrent episodes of headache and vomiting since 3 days.

Clinical History: On obtaining detailed history, patient was complaining of recurrent headache and vomiting for the past 3 days. There is no history of any fever or neck rigidity. His general physical and systemic examination were

unremarkable. Routine haematological tests, ESR, chest X-Ray and USG abdomen were within normal limits.

On retinoscopy, patient was found to be emmetropic. Slit-lamp biomicroscopy and funduscopy of the anterior and posterior segments of the eye respectively shows no abnormality on both sides.

Plain Radiography: On plain radiograph of skull, there were areas of bone destruction with irregular and poorly defined margins of inner and outer tables of skull in the occipital region [Fig 1].

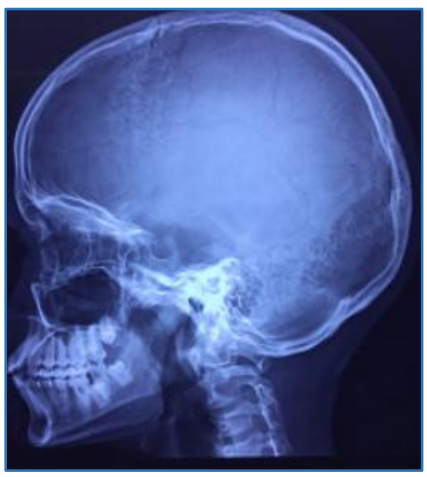

Fig. 1: Plain Radiograph of Skull, Lateral View Shows Mild Bone Destruction in the Occipital Region with Irregular Inner & Outer Tables

Computed Tomography: CT scan without contrast shows a well-defined hyperattenuating mass lesion which is broad based towards calvarium noted in the left occipital region. The mass is also noted extending into the extracranial soft tissue in the left occipital region [Fig 2]. On bone window, there is permeative type of destruction of left occipital bone noted with irregular inner and outer tables [Fig 3]. On post-contrast CT, there is intense homogenous enhancement of the mass noted including extracranial soft tissue part in the left occipital region [Fig 4A & 4B].

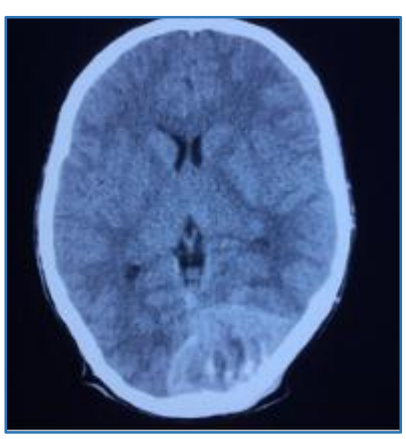

Fig. 2: NECT of Brain Showing Broad Based Hyperattenuating Mass in the Left Occipital Region which is Extending into the Extra Cranial Soft Tissue in Left Occipital Region

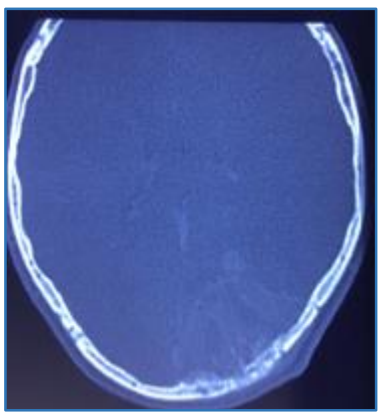

Fig. 3: CT scan Bone Window of Brain Showing Destruction of Left Occipital Bone with Irregular Inner and Outer Tables

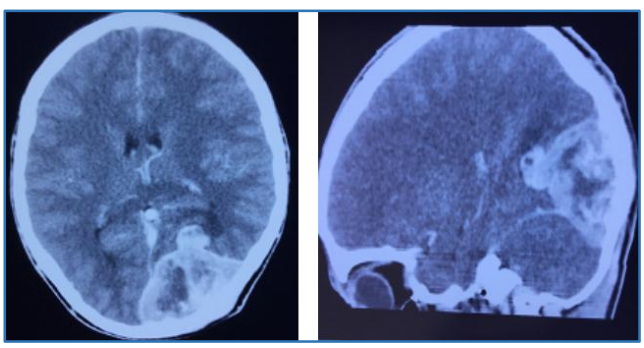

Fig 4A & 4B: Contrast Enhanced CT Axial (4A) and Sagittal (4B) Images of Brain Shows Intense Enhancement of the Mass Lesion as well as Extracranial Soft Tissue in the Region of Left Occipital Region

Magnetic Resonance Imaging: MRI brain axial and coronal sections showed a well-defined lobulated extraaxial mass lesion measuring 3.6 X 5.7 X 5.9 cm in the supratentorial left occipital region which was causing mild mass effect on midbrain from ipsilateral side. The mass was showing mixed intensity signals on both T1W & T2W Images [Fig 5 & 6]. There was no suppression of signal intensity on FLAIR sequence, but on DWI there was marked restriction of diffusion.

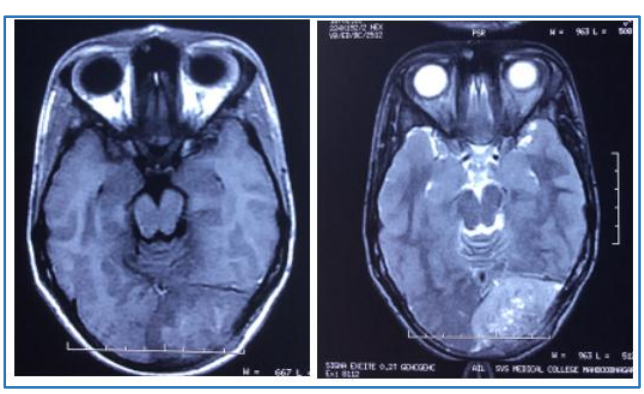

Fig. 5 & 6: MRI Axial Sections of Brain both T1W &T2W Images Shows Mixed Intensity Signal Mass Lesion in the Left Occipital Region with Suspicious Discontinuity of Left Occipital Bone and Extension into Extra Cranial Soft Tissue

Left parietooccipital craniotomy was done and a large highly vascular tumour, principally arising from the left occipital bone and involving the dura was found which was totally excised and subjected to histopathological examination.

On histopathology, the tumour showed malignant round cells arranged in sheets and few pseudorosettes were discernable. Immunohistochemistry and special stains were suggestive of Primary Atypical Ewing's sarcoma. The patient was subjected to local radiotherapy. The patient also received 9 cycles of combination chemotherapy of VAC protocol comprising Vincristine, Adriamycin and Cyclophosphamide. After 12 months, followup MRI of brain was done which showed no residual or recurrent tumour in the left occipital region.

DISCUSSION: Ewing's sarcoma is most commonly seen in children and young adults with a peak incidence in the second decade of life. It most commonly arises in long bones of the extremities (predominantly femur) and pelvis.[7] Primary Ewing's sarcoma of the cranial bone is rare and contributes about 1% of all Ewing's sarcomas.[8] Temporal bone is most commonly affected followed by frontal, parietal bone and occipital bone. Sphenoid and ethmoid bones are less commonly involved.[9]

Pathogenesis: A common chromosomal translocation that results in EWS-ETS fusion {Between the EWS gene on chromosome 22 and an ETS-type gene, most commonly the FLI1 gene on chromosome 11} has been implicated in 80% to 95% of cases of Ewing's sarcoma, PNET, and Askin tumour.[6,10] Thus, these lesions have been grouped as the same entity, called ESFT (Ewing sarcoma family of tumours).

Diagnosis: Accurate diagnosis of Ewing's sarcoma is crucial for the most appropriate clinical management of patients. Adequate clinical information and the recognition of the morphology by CT & MRI, immunocytochemical, and sometimes ultrastructural features of Ewing's sarcoma are all required for its differential diagnosis from other small round cell tumours of childhood.

Differential Diagnoses: The main differential diagnoses of tumour involving the skull with adjacent soft tissue extension in children would include metastatic neuroblastoma, PNET, chordoma, and lymphoma. Less common differential diagnoses include rhabdomyosarcoma, osteosarcoma, meningioma, Langerhans' cell histiocytosis, desmoplastic small round cell tumour, plasmacytoma, and solitary metastasis.

Treatment: Early diagnosis and treatment prior to metastasis is essential for long-term survival in patients with Ewing sarcoma.

The disease is treated through multidisciplinary approach that includes surgery, chemotherapy, and radiotherapy. Followup is necessary for the detection of recurrence.

CONCLUSION: In conclusion, primary cranial Ewing's sarcoma is to be considered in the differential diagnosis in children with a tumour involving the skull with destruction of bone and the presence of extracranial soft tissue involvement. Primary Ewing's sarcoma is reported to have a better prognosis as compared to Ewing's sarcoma elsewhere in the body.

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