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Case Report
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Ewing's Sarcoma of Sphenotemporal Bone of Skull: A Rare Presentation

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Abstract

A 17-year-old girl presented with progressively increasing diplopia,projectile vomiting of 4-month duration with no neurological deficit. Local examination showed a hard swelling that seemed to be arising from temporal bone. General and systemic examination was normal. MRI revealed a osseous tumor in left sphenotemporal region with solid cystic component. The patient was operated upon and excision of tumor was done. Histopathological examination showed a monomorphic small round cell tumor of bone infiltrating into the subcutaneous tissue. Immunohistochemical stain showed diffuse immunopositivity for CD 99 (MIC-2) in tumor cells, thus final diagnosis of Ewing's sarcoma was made. The patient was kept for regular follow up.

Key-words: Ewings sarcoma,MIC2.

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Introduction

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 [1]. Primary Ewing's sarcoma of the cranial bone is rare and contributes about 1% of all Ewing's sarcoma [2]. Considering its unusual site we report a case of Primary Ewing's sarcoma of sphenotemporal bone with soft tissue extension.

Case History

A 17-year-old girl presented with progressively increasing diplopia,projectile vomiting of 4-month duration with no neurological deficit. Local examination showed a hard swelling that seemed to be arising from temporal bone. General and systemic examination was normal. MRI revealed a osseous tumor in left sphenotemporal region with solid cystic component. The patient was operated upon and excision of tumor was done. Intraoperative frozen section revealed monomorphic small round cells arranged in clusters and scattered singly. Diagnosis of a malignant round cell

tumor was made. The tumor was sent for histopathological examination. Grossly, the specimen consists of one large grayish brown soft tissue attached to a flat bony fragment measuring cms. External surface of the soft tissue was smooth and partially encapsulated. Cut surface of the soft tissue was gray white to yellowish gelatinous and showed few hemorrhagic areas also . On microscopic examination, section revealed a monomorphic round cell tumor arranged in lobular, trabecular, and micro- and macrofollicular pattern with eosinophilic secretion in the lumen (Figure2). Tumor cells had round nuclei with stippled chromatin, prominent nucleoli, and thick nuclear membrane. Cytoplasm was moderate in amount and vacuolated. Connective tissue septae with fine blood vessels were seen throughout the tumor. Mitosis was 0–2/hpf. There was also infiltration of tumor cells in the surrounding fibroadipose tissue. Section examined from bone showed bony trabeculae and bone marrow revealing marked fibrosis and infiltration by tumor cells. On Periodic Acid Schiff (PAS) stain, tumor cells were negative. On immunohistochemistry, tumor cells were immunopositive for MIC-2. Keeping in view the morphological and immunohistochemical profile, a diagnosis of Ewing's sarcoma of bone was made.FISH studies showed EWS/FLI1 translocation which further confirmed the diagnosis of Ewings sarcoma

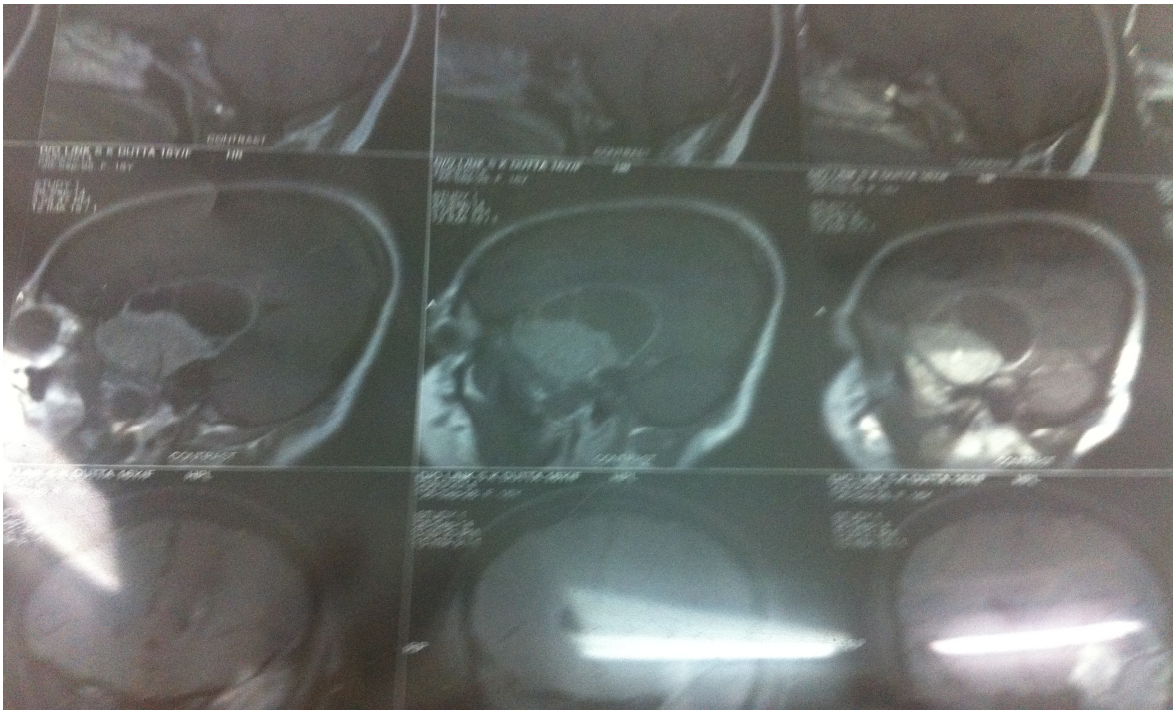

Figure 1: Postcontrast T1 weighted MRI showed intense, homogenously enhancing osseous tumor in left sphenoid and temporal bone causing destruction of bone.

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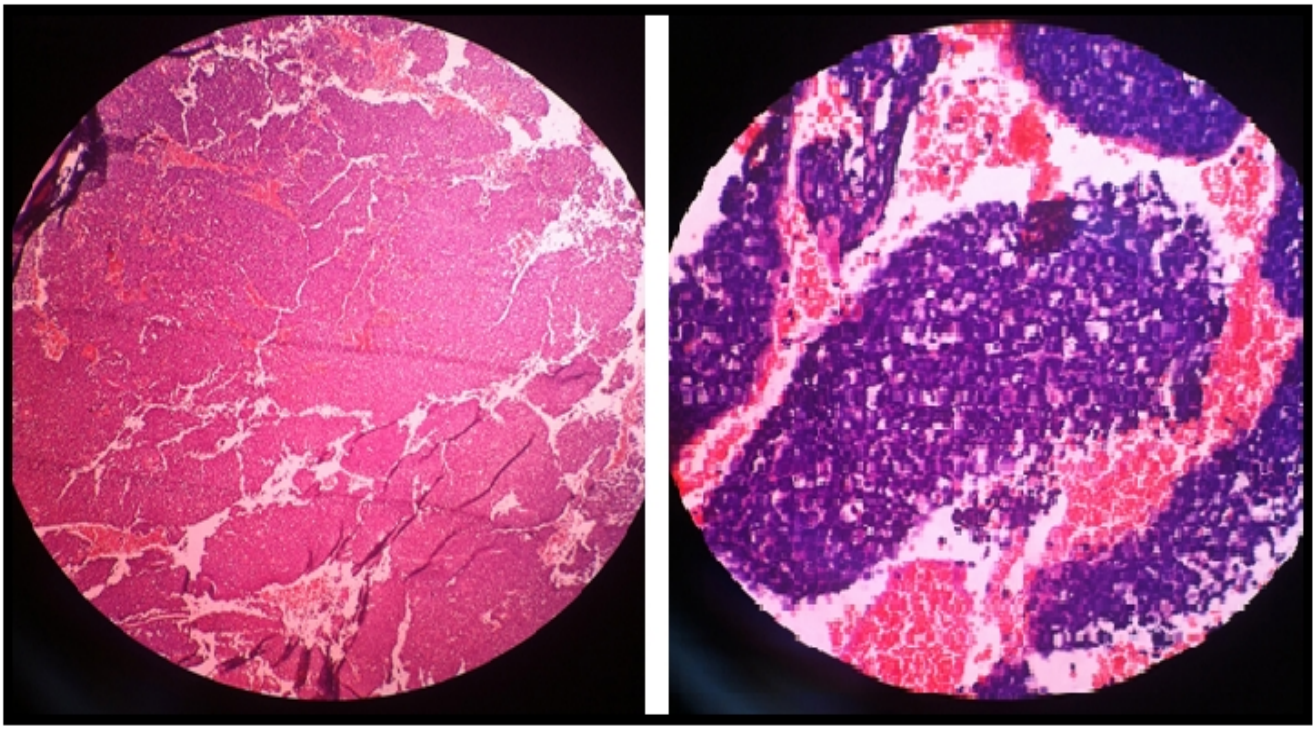

Figure 2 and fig 3 : Histopathology showing lobular arrangement of malignant round tumor cells with abundant vacuolated cytoplasm. Atypical mitosis also seen. Haematoxylin and Eosin (H&E) stain × 40x.

Discussion

Ewing's sarcoma involving the skull is rare and occurs in less than 1% of cases [2]. Sphenoid is an uncommon site for the tumor(3) as the commonest site of Primary Ewing's sarcoma is temporal bone followed by parietal and occipital bone. In our case there is no focal neurological deficit and the patient presented with diplopia and projectile vomiting.

On plain X-ray many patients may show areas of bone destruction as in our case, with irregular poorly defined margin. The most common CT finding is isodense lesion with marked heterogenous enhancement. Biopsy is essential for definitive diagnosis. In our case, the histological diagnosis is made by examining the morphology and immunohistochemical profile of tumor cells [5].

The main differential diagnosis of tumor of this particular morphology of small round blue cell tumor involving the skull with adjacent soft tissue extension in children would include metastatic neuroblastoma, PNET, chordoma, and lymphoma. rhabdomyosarcoma, osteosarcoma, meningioma, Langerhan's cell histiocytosis, desmoplastic small round cell tumor ans plasmacytoma. The differentiation between these may not be possible on light microscopy and require special stains and immunohistochemistry for final diagnosis. Although cytoplasmic glycogen content was considered to be important in differential diagnosis, in specimen fixed in formalin and embedded in paraffin, the glycogen may not be always demonstrated with PAS staining as in our case [6].

Primitive neuroectodermal tumor expresses neuronal marker such as synaptophysin, neurofilament protein, nonspecific enolase or S-100. Lymphoma cells express CD19, CD20,

and CD2,5,8. Chordoma express strong positivity for Pan CK and Epithelial membrane antigen [5]. CD99(MIC-2)is aspecific marker for Ewing's sarcoma and peripheral primitive neuroectodermal tumors [7]. Immunopositivity for MIC-2 confirmed the diagnosis in our case.

Early diagnosis and treatment prior to metastasis is essential for long-term survival in patient with Ewing sarcoma. The disease is treated through multidisciplinary approach that includes surgery, chemotherapy, and radiotherapy. This patient was kept for follow up and after five months this patient was seen to have a 30x30x15 mm residual mass left middle cranial fossa and temporal muscle. Chemotherapy and radiotherapy was then instituted in this case. In conclusion, primary cranial Ewing's sarcoma is to be considered in the differential diagnosis in children with a tumor involving the skull with destruction of bone and the presence of extra axial soft tissue involvement. Primary Ewing's sarcoma is reported to have a better prognosis as compared to Ewing's sarcoma elsewhere.

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