



## Case Report

# 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 in the scalp 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, FISH studies showed EWS/FLI1 translocation, thus final diagnosis of Ewing's sarcoma was made. The patient was kept for regular follow up.

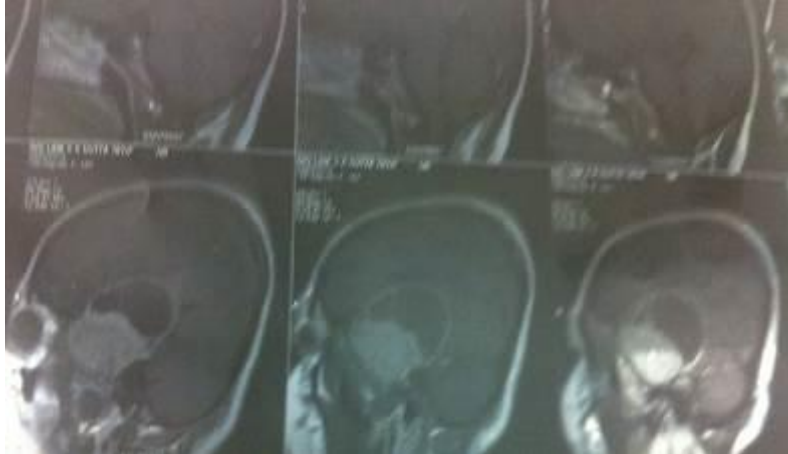
**Keywords:** Ewings sarcoma, MIC2, Sphenotemporal, EWS/FLI1.

## INTRODUCTION

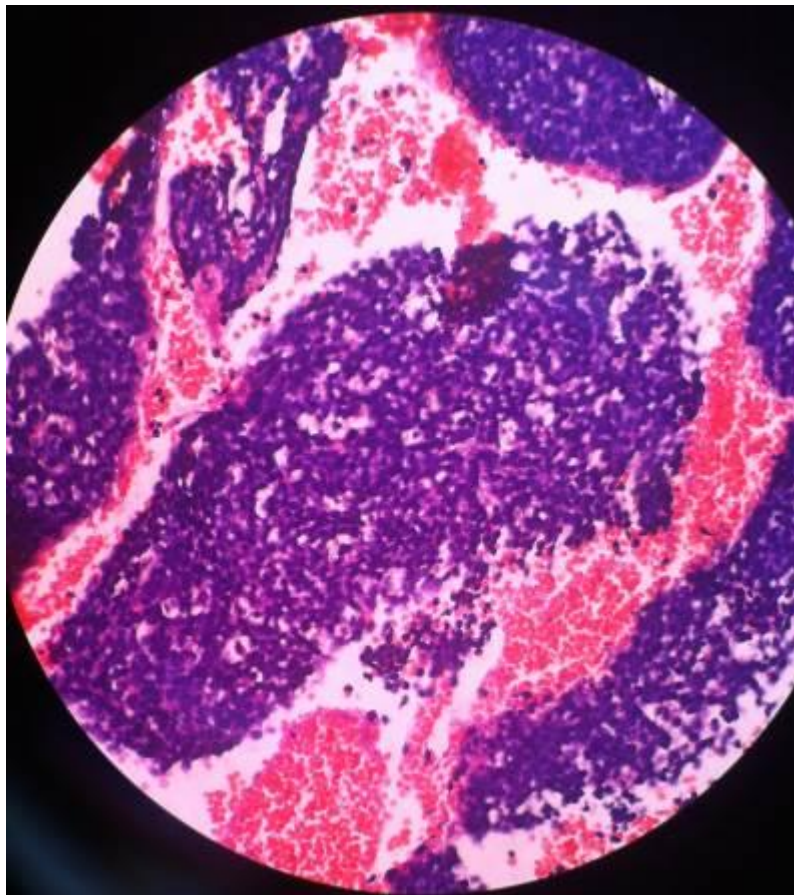
Ewing's sarcoma is a malignant, small, round cell tumor arising from bone and primarily affects children and adolescents. Ewing's sarcoma is an aggressive malignant tumor of childhood and the second most common primary bone cancer. Among pediatric patients, Ewing's sarcoma accounts for 6 to 9% of malignant bone neoplasms.<sup>1,2</sup> 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 (Schmidt Det al., 1987). Primary Ewing's sarcoma of the cranial bone is rare and contributes about 1% of all Ewing's sarcoma (Sim et al., 1979). Considering its unusual site we report a case of Primary Ewing's sarcoma of sphenotemporal bone which is very rare.

## 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 (Figure1). 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



**Fig 1.** MRI showing a tumor mass in the temporal region



**Fig 2.** Microphotograph showing monomorphic round cell tumor arranged in lobular, trabecular, and micro- and macrofollicular pattern with eosinophilic secretion in the lumen

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 Ewing's sarcoma. After surgery the patient defaulted follow up and after five months this patient was seen to have a 30x30x15 mm residual mass in left middle cranial fossa and temporal muscle. Chemotherapy and radiotherapy was then instituted in this case with good results. The girl is now under regular follow up.

## DISCUSSION

Ewing's sarcoma involving the skull is rare and occurs in less than 1% of cases (Schmidt Det al., 1987 and Sim et al., 1979). Sphenoid is an uncommon site for the tumor (Unni KK., 1996) 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 but the patient presented with diplopia and projectile vomiting.

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 (Unni KK., 1996; Steinbok et al., 1986 and Desai et al., 2000).

The main differential diagnosis of tumor of this particular morphology of small round blue cell tumor involving the skull in children includes metastatic neuroblastoma, PNET, chordoma, lymphoma, rhabdomyosarcoma, osteosarcoma, meningioma, Langerhan's cell histiocytosis, desmoplastic small round cell tumor and plasmacytoma. The differentiation between these may not be possible on light microscopy and require special stains and immunohistochemistry for final diagnosis.

The different tumor entities considered in the differential diagnosis are distinguished as the Primitive neuroectodermal tumor expresses neuronal marker such as synaptophysin, neurofilament protein, nonspecific enolase or S-100. Lymphoma cells express CD19, CD20. Chordoma express strong positivity for Pan CK, brachury and Epithelial membrane antigen. CD99(MIC-2) is a specific marker for Ewing's sarcoma and peripheral primitive neuroectodermal tumors (Ambros et al., 1991 and Dickman et al., 1982). Immunopositivity for MIC-2 confirmed the diagnosis in our case. (Miettinen et al.,

1982 and Navas-Palacios et al., 1984)

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 in left middle cranial fossa and temporal muscle. Chemotherapy and radiotherapy was then instituted in this case with good results. 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.

## CONCLUSION

This case report of a 17-year-old girl with progressive increasing diplopia adds to the five previously reported cases of this rare form of Ewing's sarcoma affecting the sphenoid bone. When this bone is affected, the tumor presents unique management considerations because of its intimate relationship with critical neurovascular structures in this region. Given its rarity for affecting the skull, particularly the sphenoid bone, our findings add to our global understanding of the treatment options and outcomes for this uncommon subset of patients affected by Ewing's sarcoma. 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 had no symptoms. 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 of sphenotemporal bone is reported to have a better prognosis as compared to Ewing's sarcoma elsewhere

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