PRIMARY EXTRAOSSEOUS EWING SARCOMA / PNET AT AN EXTRAORDINARY SITE - THE ORBIT

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ABSTRACT

Ewing sarcoma/ PNET of the orbit is usually a metastatic tumor. Primary orbital Ewing sarcoma/ PNET is extremely rare with only 17 reported cases as per literature. One such rare case of a primary extraskeletal Ewing sarcoma/ PNET of the orbit, which is an extraordinary site for its occurrence in a 4year old boy, is presented here.

Keywords – Primary, Orbital, PNET, Ewing sarcoma

ABBREVIATIONS

PNET – Peripheral neuroectodermal tumor

SRCT- Small round cell tumor

INTRODUCTION

Ewing sarcoma/ PNETs constitute about 4% of head and neck tumors[1]. Orbital involvement in Ewing sarcoma/PNET is mostly through metastatic spread. Primary orbital Ewing sarcoma/ PNET is extremely rare and is reported only in 17cases till date as per extensive literature search. These tumors present in second and third decade of life. The common sites of occurrence are thoracic cavity and extremities. These malignant tumors can arise from the bone or soft tissues. Histologically they appear as small round cell tumors. The extraosseous Ewing sarcoma and PNETs share a unique and consistent genetic translocation t(11;22) (q24;q12).

CASE HISTORY

A 4year old boy developed proptosis and discomfort in his left eye over a period of two weeks. Ocular examination revealed decreased visual acuity, mild upper lid swelling and moderate (4mm) axial proptosis. Fundal examination showed retinal folds in the macular region. No other ophthalmic or systemic abnormalities were detected.

Orbital computed tomography showed a soft tissue mass in the superomedial aspect of the orbit in the retrobulbar region of both the intra and extraconal compartments. Hyperostosis of the orbital roof with expansion of the orbital apex was seen. The lesser wing of the sphenoid was eroded and the mass was seen bulging into the middle cranial fossa and posterior ethmoidal sinus. The clinicoradiological diagnoses considered were meningioma, lymphoma and rhabdomyosarcoma.

PATHOLOGICAL FINDINGS

The specimen consisted of multiple, friable, hemorrhagic, soft tissue fragments amidst small bony spicules.

Microscopic examination showed a highly cellular tumor with scattered nests and sheets of small round tumor cells having scanty cytoplasm, even nuclear chromatin and high nucleo-cytoplasmic ratio against a desmoplastic stromal background. Normal bony spicules were scattered amidst these tumor cells. Based on these features, a histological diagnosis of orbital Ewing sarcoma/ PNET was arrived at.

Immunohistochemistry showed diffuse membrane positivity for CD99.

Extensive systemic evaluation including computed tomography of the chest, abdomen and pelvis did not reveal any significant findings.

DISCUSSION

Ewing sarcoma/ PNETs are a group of soft tissue tumors of presumed neural crest origin arising outside the central and sympathetic nervous systems[2], occurring most commonly between 10 and 30years of age. These constitute approximately about 4% of all soft tissue tumors. They show a male predilection with a male to female ratio of 1.4:1. They arise in the deep soft tissue particularly of trunk and lower limbs.

Ewing sarcoma/ PNETs represent a cohesive family of tumors, which show varying degrees of neuronal differentiation with consistent EWS gene rearrangement[3]. The term Ewing sarcoma has been used for those tumors that lack evidence of neuroectodermal differentiation as assessed by light microscopy, immunohistochemistry and electron microscopy[4],

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whereas the term PNET has been employed for tumors that demonstrate neuroectodermal features as evaluated by one or more of these modalities.

Histologically, Ewing sarcoma/ PNETs show a predominantly lobular or sometimes trabecular growth pattern with a prominent ramifying capillary network. Ewing sarcoma lying at the poorly differentiated end of the spectrum shows cells with scanty cytoplasm and round to oval open nuclei with dusty chromatin. On the other hand, PNETs being at the better-differentiated end of the spectrum show cells with eosinophilic cytoplasm, coarser chromatin, more frequent nucleoli and numerous rosettes.

Primary orbital Ewing sarcoma/ PNET which was the final diagnosis in the present case, is extremely rare. But, secondary involvement of the orbit in Ewing sarcoma/ PNET is commoner. In the present case, the other small round cell tumors (SRCTs) considered in the differential diagnoses were, lymphoblastic lymphoma and alveolar rhabdomyosarcoma.

Lymphoblastic lymphoma is an important mimicker of Ewing sarcoma[5]. It was ruled out considering the nesting patterns and arrangement of the tumor cells in sheets. Alveolar rhabdomyosarcoma was ruled out considering the absence of abundant cytoplasm and prominent nucleoli in the tumor cells.

The primary nature of the neoplasm was confirmed by the lack of evidence of any other site of involvement. The soft tissue mass with tumor cells amidst normal bony spicules ruled out the possibility of primary sphenoidal involvement, thus confirming the primary nature of the tumor.

Now it is considered that the degree of neuroectodermal differentiation does not determine the prognosis. The survival depends on the type of genetic rearrangements associated with the tumor. The prognostic factors could not be assessed in the present case, since the patient was referred to a higher centre for management and was lost for follow up.

CONCLUSION

When a primary SRCT of the orbit is encountered, one may overlook the possibility of Ewing sarcoma/PNET, since the orbit is an extremely rare site for his tumor. The present case report highlights the fact that it is prudent to consider this tumor while arriving at the diagnosis since the prognosis and management depend on the precise category of the SRCT.

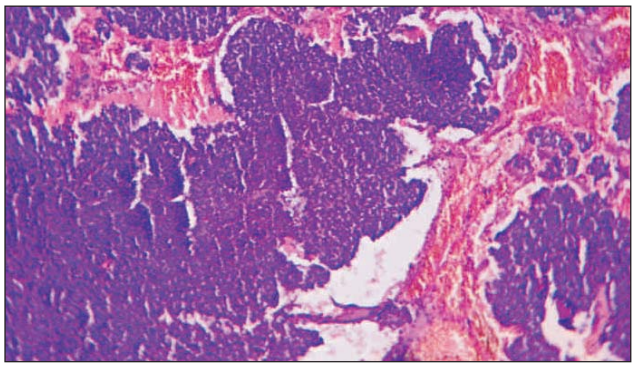

Fig.1 Small round tumor cells in sheets with intervening areas of hemorrhage (Hematoxylin and Eosin x 100)

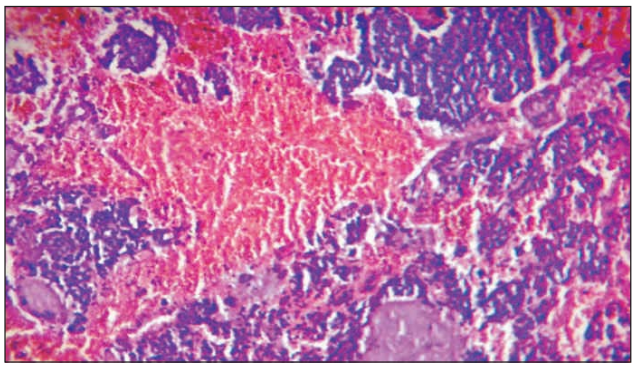

Fig.2 Small round tumor cell nests against desmoplastic stromal and hemorrhagic background. (Hematoxylin and Eosin x 100)

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