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Case Report

Primary Extra Skeletal Ewing Sarcoma Presenting as Soft Tissue Mass Diagnosed on Fine Needle Aspiration Cytology

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ABSTRACT

Extra skeletal Ewing sarcoma (EES) is a rare malignant mesenchymal tumor. It is histologically similar to its primary counterpart. However, its cutaneous presentation is a rare occurrence.
Ewing sarcoma or Primitive Neuro Ectodermal Tumor (PNET) when present in soft tissues characterizes an unusual clinical presentation. Commonly affected are the upper & lower extremities, trunk, head, and neck.
We present a case of 9 years old boy presented with a single subcutaneous mass in upper thigh region diagnosed as Ewing sarcoma on cytology.
Pre-operative cytological diagnosis of ES or subcutaneous Ewing sarcoma (SES) is extremely rare with only few cases being reported based on fine needle aspiration cytology (FNAC) findings.

Key words: Extra skeletal Ewing's sarcoma, pediatric malignancy, small round blue cell tumour, FNAC.

INTRODUCTION

James Ewing [1] in 1921 first described, what is now known as Ewing sarcoma. Initially regarded as an undifferentiated type of bone sarcoma of children, Ewing sarcoma is now linked to Primitive Neuro Ectodermal Tumor (PNET). Owing to the recent advances in cytological, histological, molecular studies & genetics, the term Ewing sarcoma/ PNET is currently favoured for this tumor family. Extra skeletal Ewing sarcoma was recognised as a soft tissue tumor in 1975 by Angervall & Enzinger. [2] Till 2010, primary cutaneous Ewing's sarcoma was reported in only 82 cases. ES is usually diagnosed on histology, immunohistochemistry & chromosomal studies.

CASE REPORT

A 9 year old boy presented to the surgery OPD with complaints of swelling in the left thigh region since 3 months (Figure 1). Examination revealed a single, tender, subcutaneous, mobile, firm swelling not attached to deeper tissues and approximately measuring 2 x 2 cm was seen. The overlying skin was however unremarkable.

X ray lateral aspect of thigh revealed a subcutaneous 2.2 x1.8 x1 cm opaque mass (Figure 2). Plain CT scan described a subcutaneous 2 x 2 x 1.8 cm radio-dense

lesion in the lateral aspect of upper left thigh with a feeding vessel indicating increased vascularity (Figure 3).

FNAC was performed using aspiration& non aspiration techniques. Smears were obtained & studied under light microscopy. Microscopically, the cell showed 2 cell populations. Large, pale cells with fine granular chromatin &1-2 nucleoli with abundant cytoplasm along with numerous small dark cells with scanty cytoplasm in loosely cohesive sheets, clusters& scattered singly were noted (Figure 4). Also seen were numerous stripped nuclei. Occasional rossette like structures were also noted (Figure 5).

Based on above finding, diagnosis of soft tissue lesion suggestive of subcutaneous Ewing sarcoma / PNET was given.

According to few studies EES shows dissociated clusters of closed packed small uniform round cells with round bland nuclei. The tumour shows rich vascularity.

IHC was performed on the cell block preparation which revealed CD 99 membrane positivity. There by confirming the diagnosis.

Pre-operative cytological diagnosis of Ewing sarcoma or subcutaneous Ewing sarcoma is extremely rare with very few cases being reported based on fine needle cytology aspirate findings.

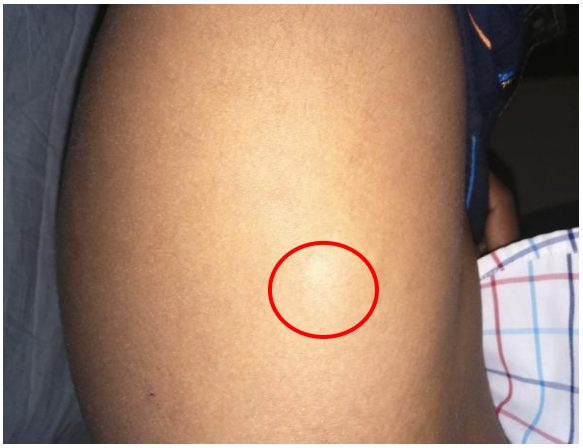

Figure 1: Photograph showing a small nodule over the left thigh(circled).

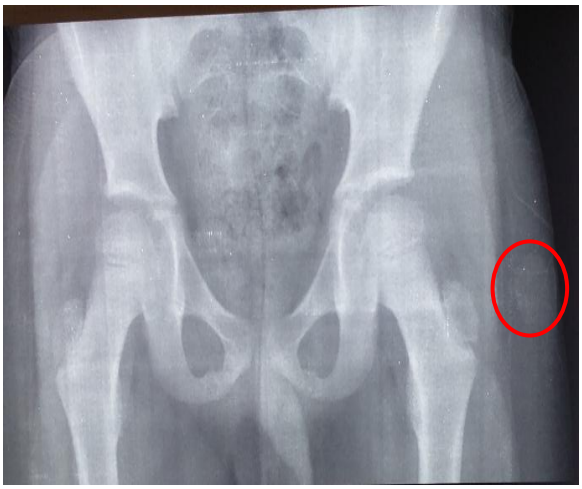

Figure 2: X-ray of the thigh region showing a small subcutaneous nodular mass (encircled) .

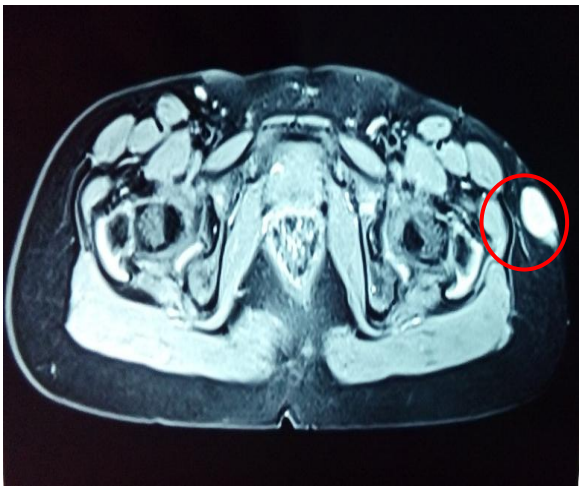

Figure 3: CT scan showing a subcutaneous mass in the thigh not attached to the underlying structures.

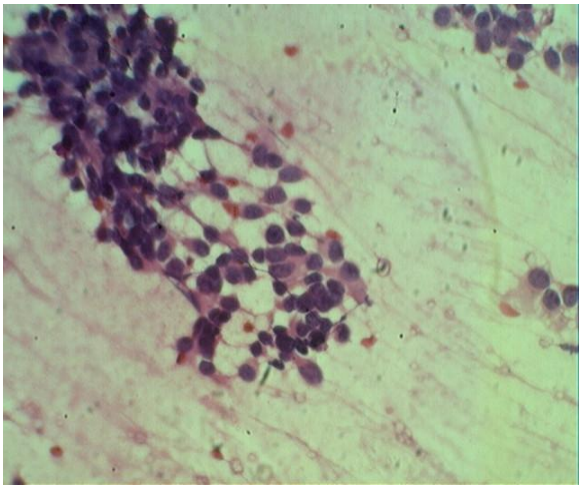

Figure 4: Photomicrograph showing hypercellular smear with small dark neoplastic cells with scant cytoplasm and large pale cells with fine granular chromatin and abundant cytoplasm. (H&E, 100x)

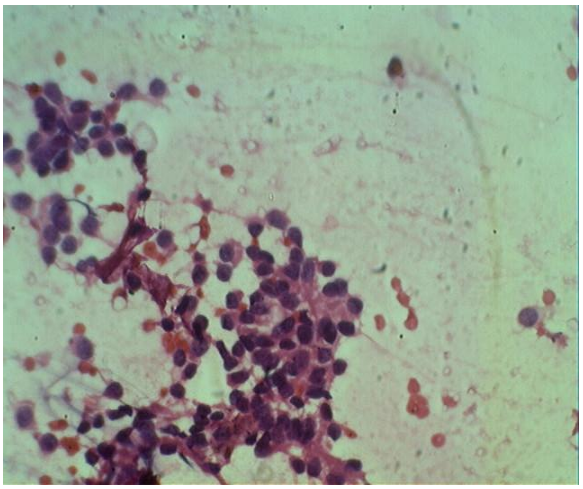

Figure 5: Photomicrograph showing hypercellular smear with small dark neoplastic cells with scant cytoplasm and large pale cells with fine granular chromatin and abundant cytoplasm alongwith few rosette like structures. (H&E, 100x)

DISCUSSION

Ewing sarcoma represents the prototype for small round blue cell tumors. It is now days referred as Ewing sarcoma/ PNET family.

Owing to the advent of IHC, cytogenetic & molecular genetics techniques, these tumors together represent a spectrum known as Ewing family of tumors [2] which includes Ewing's sarcoma of bone, extra skeletal Ewing sarcoma, peripheral primitive neuroectodermal & Askintumor. t (11: 22) (q 24; q12) is the common cytogenetic abnormality found in both Ewing's sarcoma as well as PNET.

Extra-skeletal Ewing sarcoma (EES) is a rare, rapidly growing, aggressive soft tissue tumor having a wide range of occurrence from infancy to old age. It can affect practically any location, with most common sites being paravertebral, intercostal regions followed by extremities.

EES arises in the soft tissue of the trunks. Other sites such as larynx, nasal fossa, neck, lungs, retroperitoneum, perineum & mediastinum are also affected. [3,4]

EES has a low incidence rate of around 1.1 % of all malignant soft tissue tumors& a wide range of occurrence ranging in age from 22 months to 81 years & more commonly found in males.

The clinical manifestations of EES are nonspecific. Patients may present with painless mass or swelling anywhere in body.

It shares histological, IHC & molecular features with its osseous counterparts. [5] Due to better compliance & reliable findings, FNAC is increasingly being used for the diagnosis of such cases.

Though, histopathology is a gold standard in the diagnosis of EES, aspiration cytology followed by IHC or cell blocks, increasingly has become a reliable mode of diagnosis. EES is a highly malignant round cell tumor, with an aggressive clinical behavior. [4]

Distant metastasis & local recurrence are very common. [4]

According to Xie et al clinical diagnosis of EES is hampered due to low incidence of tumor, due to nonspecific symptoms, signs of imaging & no distinct histological features.

Due to lack of characteristic morphological features, the differential diagnosis includes other round blue cell tumors such as RMS, lymphoma, synovial sarcoma, neuroblastoma. [4,6,7]

EWS FL 1 gene rearrangement is highly specific for ES/ PNET and is seen in more than 90 % of tumors. [8]

These tumors express prolific CD99/ MIC 2 membrane positivity. This is helpful in differentiating these tumors from neuroblastoma & other round cell tumors. [8]

The 5 year survival rate for Skeletal Ewing sarcoma is around 75%. EES has a worse prognosis having a 5 year survival rate of 38%. However, survival rates upto 61% by obtaining a wide tumor free resection margin following multi agent chemotherapy have also been documented. [4, 7]

Newer advances in investigation treatment protocols such as multidrug chemotherapy have indicated better prognosis in cases of EES.

FNAC avoids an invasive procedure by rendering reliable diagnosis & hence it is proving to be a rapid& reliable, patient compliant diagnostic modality.

REFERENCES

1. Ewing J. Diffuse endothelioma of bone. Proc N Y Pathol Soc. 1921; 21: 17
2. Malignant Soft tissue tumors of Uncertain type. Editors: John R Goldblum, Andrew L Folpe, Sharon W. Weise. In: Enzinger& Weiss Soft tissue tumors, 6ed. Philadelphia, Elsevier; 2014: p1028-1112.
3. Ahmad R., Mayol B.R., Davis M. &Rougraff B.T. Extraskeletal Ewing's sarcoma. Cancer, 85(3), 1999, 725-731.
4. Buch A.C., Panicker N. K., Sarawagi S., Anwekar S., Kharat A.T. Fine needle aspiration cytology diagnosis of paravertebral extraosseous Ewing's sarcoma. Journal of cytology/ Indian Acedemy of Cytologists, 27(4), 2010,146.
5. GuiterGE., Gamboni MM., Zakowski MF." The cytology of extraskeletal Ewing sarcoma." Cancer Cytopatholgy 87.3.1999, 141-148.
6. X Chun- Fang. M-Z Liu, and M Xi. "Extraskeletal Ewing's sarcoma: a report of 18 cases and literature review." Chinese journal of cancer 29.4.2010,420-424.
7. Manduch M., Dexter D.F., Ellis P.M., Reid K., Isotalo P.A. Extraskeletal Ewing's sarcoma/primitive neuroectodermal tumor of the posterior mediastinum with t(11;22)(q24;q12). Tumori, 94(6).2008,888.
8. Dr. Bharathi M , Dr. NeelimaPrabha C, Dr. Vani D, Dr.Sharath Kumar H.KExtraosseous Ewing Sarcoma – Cytological Diagnosis of a Rare Entity IOSR Journal of Dental and Medical Sciences, 2015; 14(2) 21–24.

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