CASE REPORT

EXTRAOSSEOUS EWINGS/PNET-A CASE REPORT BY FNAC

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INTRODUCTION: Ewing sarcoma is one of the most common primary tumor of the bone, but extra-skeletal type is rare. Ewing's sarcoma (ES) and primitive neuroectodermal tumor (PNET) are small round cell tumors showing varying (usually minimal) neuroectodermal differentiation^[1]. These tumors occur predominately in adolescents and young adults, but rare below four years of age. The cytomorphologic features may be confused with other small round cell tumors.

Here we present a rare case occurring in one and half year male child presented with hughenape swelling, suggesting on FNAC cytomorphologic feature as EWS/PNET

CASE REPORT: A one and half year male child, presented with swelling in nape of neck since six months, measuring 13x10 cms, external surface was nodular and showed dilated veins. FNAC smears were highly cellular, showing sheets and dispersed population of malignant small round cells in a hemorrhagic background (Fig.1). The tumor cells had small round hyperchromatic nuclei, coarse chromatin, inconspicuous nucleoli, and scanty cytoplasm. Occasional rosette like formation and pseudopapillary patterns were noticed (Fig.2).

DISCUSSION: Extra-skeletal ES/PNET occurs predominantly in the deep soft tissue of the chest, paravertebral region, abdomen and pelvis, and extremities. EWS/PNET involving extra-skeletal tissue is also reported rarely in kidney, intestine, ovary, vagina, and skin^[2].

In most large series published to date EWS/PNET accounts 20% of soft tissue sarcomas, usually present in the second decade of life, with a slight male preponderance. Of note, these tumors are rare in African American children and children of Asian descent, with most worldwide cases occurring in white and Hispanic children and adolescents. They occur most often in children but rare in <4 years of age^[3].

The cytologic diagnosis in this case was primary soft tissue (extra-skeletal) Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET). This diagnosis was based on a combination of the clinical picture, cytology and the routine light microscopy. The differential diagnosis includes a number of other neoplasms which fall under the rubric of malignant small round cell tumors. As this was a primary soft tissue neoplasm, most of the other entities are even rarer than an extra-skeletal ES/PNET. Even though rosettes like structures were present neuroblastoma was ruled out due to site and absence of neurophil pigment inside rosettes. Alveolar rhabdomyosarcoma was ruled out due to absence of tumor giant cells and eosinophilic cytoplasm.

First sarcomas to be recognized having signature translocation, subsequently shown to be shared by PNET and Askin tumor. These show morphological overlap and have similar EWS

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gene rearrangements i.e.,(11,22)(q12,q24) involving the EWS gene at 22q12 and prognosis and response to chemotherapy. Hence these are considered as closely related members of the same family known as ES/PNET or Ewing family of tumors^[4].

Number of studies has addressed the diagnosis of EWS/PNET by fine-needle aspiration (FNA) biopsy based on cytologic features and the application of immunohistochemistry^[5].

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SRCTs form a formidable group and an overwhelming majority of childhood malignancies including non-Hodgkin lymphoma, neuroblastoma, retinoblastoma, hepatoblastoma, nephroblastoma, rhabdomyosarcoma, small cell anaplastic carcinoma, Ewings sarcoma, peripheral neuroectodermal tumor, and desmoplastic small round cell tumor. The patients may present in later (inoperable) stage with huge intra-thoracic and intra-abdominal mass, where chemotherapy and/or radiation therapy may be the first or only line of treatment^[6].

CONCLUSION: As a less invasive procedure fine needle aspiration (FNA) cytology has definite advantage over surgical excision biopsy to arrive at a tissue diagnosis before initiation of therapy^[6]. EWS/PNET/SRCTs are rare & morphologically very similar, but doesn't have specific antigens that could be demonstrated with immunocytochemistry or they lose them when poorly differentiated, cross-reactivity exists between some SRCTs, therefore it is difficult for cytopathologists to obtain experience for rendering reliable diagnoses.

The ancillary facilities may not be available in all laboratories in developing countries and even if available in limited form, hence a tissue diagnosis has to be offered often by FNA cytology based on cytomorphological features as a life saving measure in serious patients before the results of ancillary studies are finalized^[7].

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Fig.1. Microscopy, showing sheets and dispersed population of malignant small round cells in a hemorrhagic background.(H & E.10x)

Fig.2. Microscopy showing, tumor cells having small round hyperchromatic nuclei, coarse chromatin, inconspicuous nucleoli, and scanty cytoplasm, occasional rosette formation and pseudopapillary pattern (H & E.40x).