

CASE REPORT

A CASE OF EXTRASKELETAL EWING'S SARCOMA/PNET PRESENTING AS A MEDIASTINAL MASS

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ABSTRACT: The Ewing's sarcoma/PNET family of tumors comes under small round blue cell tumors (SRBCT's). These are high grade relatively rare malignant neoplasms affecting children and young adults predominantly in 2nd decade. Most of these are seen affecting major long bones like femur, pelvis and ribs. Cases of extraskeletal Ewing's sarcoma (EES) involving chest wall, pleura, mediastinum, retroperitoneum, paravertebral region, lower extremities and even very few cases of isolated maxillary sinus Ewing's sarcoma have been reported.^[1] These malignancies are characterized by a chromosome 22 rearrangement.^[2] They are highly aggressive tumors with a high incidence of local recurrence and distant metastases.^[3] Recent studies suggest that this tumour is likely to originate from primitive stem cells and degree of malignancy depends upon the stage of stem cell arrest during differentiation. PNET shows variable degree of neuroectodermal differentiation.^[4]

CASE REPORT: We report a very rare case of primary mediastinal Ewing's sarcoma. An 18 year old, previously healthy, female presented in the OPD with left sided constant dull-aching chest pain, dysphagia and generalised weakness for two months. A CT scan was done which revealed a large increased attenuating space occupying lesion in left upper and middle zones having solid as well as cystic components, pulmonary deposits, and displacement of mediastinum and raised left lobe of diaphragm. CT-guided FNAC was performed from the left mediastinal mass and it revealed a dimorphic population of cells in the prepared smears. One population of cells showed clusters of uniform round cells with single hyperchromatic nuclei, dense chromatin and scanty basophilic cytoplasmic rim.

The other population showed larger round-to-oval light staining cells with fine chromatin and one or two small nucleoli having a moderate amount of vacuolated cytoplasm. These cells were tending to form rosettes at places suggesting a possibility of PNET/ Ewing's family of tumors. Pneumonectomy of the patient was done on the basis of CT findings and CT-guided FNAC and after the histopathological processing of the specimen, it revealed similar bimodal population of cells, one of them being small darkly staining blue cells and the other being large light cells with moderate amount of cytoplasm tending to form rosettes at places. It was further confirmed by immunohistochemistry. Tumour cells displayed diffuse cytoplasmic CD99 positivity.

DISCUSSION: Primary mediastinal tumors are a rare entity. They are mostly located in posterior mediastinum. Involvement of the anterior mediastinum is still rarer.^[5] The site and clinical presentation in our patient suggested a possibility of small round cell tumors like Ewing's sarcoma/PNET, Rhabdomyosarcoma, Neuroblastoma, Non-Hodgkin's lymphoma. Then, we went for a panel of immunohistochemical markers which revealed diffuse cytoplasmic CD99 positivity, while showing negativity for desmin, myogenin, NSE, CD56, TdT, CD45, CD3, C20 and ALK, thus ruling out the other possibilities.

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Rhabdomyosarcoma can be positive for CD99 but myogenin and desmin negativity ruled it out. Neuroblastomas are usually negative for CD99 and positive for neuron specific enolase (NSE) and CD56. Hence, it was also ruled out.

The most common types of non-hodgkin's lymphomas seen in children are T and B cell lymphoblastic, Burkitt and Anaplastic large cell lymphoma. This presents like SRBCT'S as the lymphoblastic lymphomas also presents in young teenage boys, very commonly, as a mediastinal mass. Most of these are positive for Terminal deoxynucleotidyl transferase (TdT). TdT negativity in our case did not support this diagnosis. Also, lymphomas usually present as showing singly dispersed monotonous population of lymphoblasts while in our case, there was seen a tendency of rosette-formation. Negativity for CD3, CD20, CD45 and ALK also ruled out lymphomas.^[6]

Thus, with the use of a variety of IHC markers and MIC-2 gene product (CD99) positivity, we could establish the diagnosis of Ewing's Sarcoma/PNET. It also shows FLI-1 positivity.^[7]

CONCLUSION: Although a rare entity, extraskeletal ES/PNET should be considered in the differential diagnosis of primary mediastinal neoplasms, especially in children and young adults. Further, if cytology and histology reveals small-round-cell-tumour-like picture, we should go in for immunocytochemistry, immunohistochemistry, cytogenetic analysis and other molecular tests to identify chromosomal translocations. 90% of the molecular analyses results show t (11; 22) (q24; q12) translocation. All these investigations are invaluable to establish a specific diagnosis.

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Fig. 1: Gross image of the pneumonectomy specimen received showing gray white friable areas representing the mass.



Fig. 1

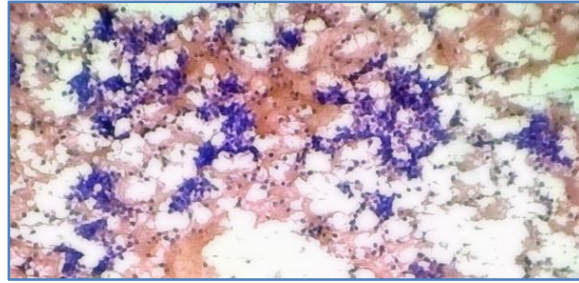


Fig. 2

Fig. 3: Histopathology showing bimodal population of small blue round cells and lighter staining larger cells. (400x H&E).

Fig. 4: Immunohistochemistry showing diffuse cytoplasmic CD 99 positivity.

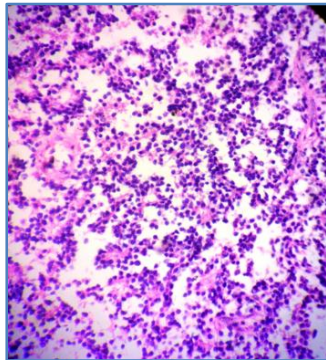


Fig. 3

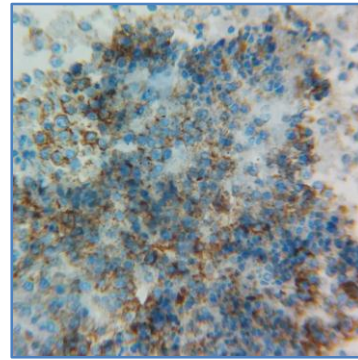


Fig. 4

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