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Case Report

Posterior Mediastinal Mass-Extraskeletal Ewings Sarcoma

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ABSTRACT

Ewings sarcoma arises from the bone. But sometimes they can arise from non osseous tissues which are labeled as Extraskeletal Ewings Sarcoma. Rarely they can arise from mediastinum. Posterior mediastinal mass is a common entity in clinical practice. Most of them are neurogenic in origin. However, rarely they can be of other histogical varieties. Proper histogical evaluation with immunohistochemical studies should be done to arrive at a proper diagnosis.

Key Words: Posterior Mediastinal Mass, Extraskeletal, Ewings Sarcoma, Small round cell.

INTRODUCTION

Ewings sarcoma is a highly malignant small round cell tumor and occurs most often in children and adolescents. [1] Extraskeletal Ewings sarcoma (ESS) is the Ewings sarcoma originating from non bone tissues. EES is a rare rapidly growing, round cell malignant tumor that can develop in soft tissues at any location. It was first described by Tefft et al in five young children all of whom presented with features of epidural spinal cord compression. [2] Later Angervall and Enzinger reported a larger series of these soft tissue tumors in adolescents and young adults. Involvement of mediastinum as the primary site of origin is extremely uncommon in literature, only four cases were reported in a large case series from tertiary hospitals in India over a span of twenty years. [3] Here, we are presenting a case of EES presenting as a posterior mediastinal mass.

CASE REPORT

A 17 year old girl presented in the hospital with dull aching backache and history of dysphagia and dyspnoea for last two weeks. It was insidious in onset and gradually progressive in nature. The pain was continuous in nature. Dysphagia was mainly for solids. Dyspnoea was gradually progressive in nature and at present it was at rest. There was associated history of dry intermittent cough. No history of change of voice. There was associated history of loss of weight and low grade fever.

On examination patient was poorly built, pale and dyspnoeic. There were

decreased breath sounds in right infrascapular area. Rest of the systemic examinations was within normal limits. Laboratory investigation revealed low hemoglobin. Chest roentograph revealed a lobulated posterior mediastinal mass measuring around 4x4 cm. CT scan thorax revealed a posterior mediastinal mass compressing the lower lobe of the right lung and with compression of the esophagus as shown in Fig: 1 Ultrasound guided FNAC of the mass was done which revealed small round cell tumor as shown in Fig: 2 Immunohistochemical studies were done on cell block which showed MIC 2 positivity and synaptophysin and chromogranin was negative as shown in Fig: 3. Cytogenetic study showed metaphases of t (11:22) translocation. A diagnosis of Extraskeletal Ewings Sarcoma (EES) was made. Bone scintigraphic studies showed extensive skeletal metastases with bone marrow examination was normal. Patient was started on palliative chemotherapy.

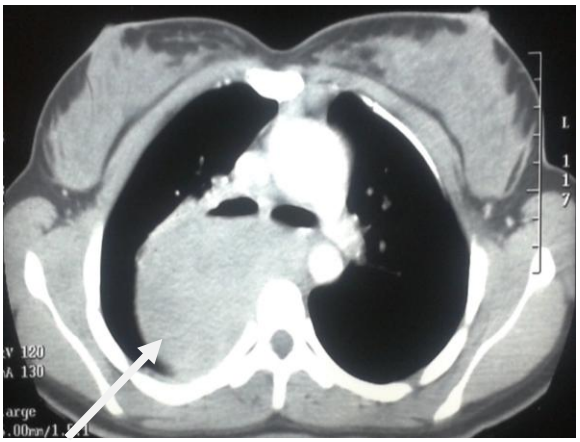

Fig. 1. Well defined smooth bordered homogenously enhancing soft tissue attenuating mass lesion is seen in right posterior mediastinum. Lesion causing compression and displacement of esophagus. Right hilum is partially encased and pushed anteriorly. Underlying rib and adjacent vertebral body is normal.

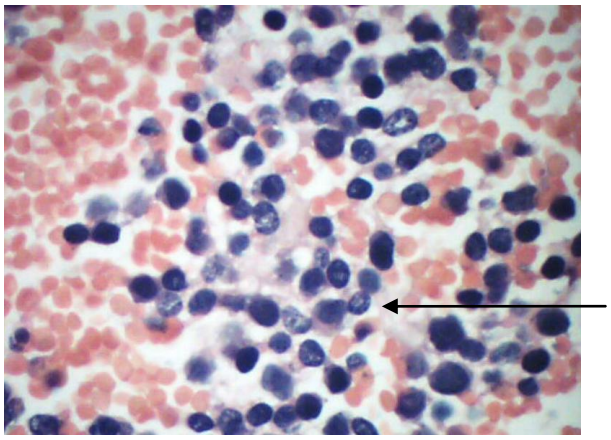

Fig. 2. PAP stained FNAC smear shows moderately cellular smear with round cells and scant cytoplasm and mild nuclear pleomorphism in a hemorrhagic background. Focal rosette formation is evident.

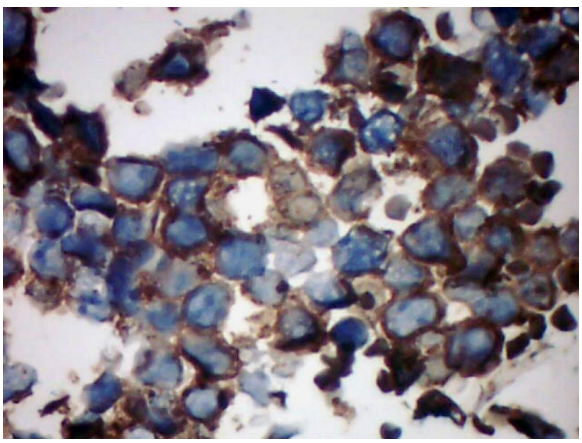

Fig. 3. Round cells showing cytoplasmic and membrane positivity for CD 99.

DISCUSSION

Hart and Earle first coined the term primitive neuroectodermal tumor (PNET) paying more attention to the primitive nature of the tumor rather than histogenesis. [4] ES/PNET is an uncommon malignancy of children and young adults. Primary osseous ES tends to involve the diaphysis or metaphyseal-diaphyseal regions of the long bones. Less frequently, they can have a primary soft tissue origin, with the neoplasm being classified as EES. Usually sites for EEs are supratentorial brain, spinal canal and soft tissue of thigh and primary involvement of extra cranial sites like mediastium is rare. [5] In a study of EEs by Ahmad et al, [6] only one of 24 patients presented with tumor involving the mediastinum. Classically, the most common neoplasms that arise in the posterior

mediastinum include neurogenic tumors like schwannoma,, neurofibroma, ganglio-neuroma, ganglioneuroblastoma, neuroblas-toma and para gangliomas.

ES is a small round cell tumor and it is difficult to distinguish histologically from other small round cell tumors like rhabdomyosarcoma, desmoplastic small round cell tumor, poorly differentiated synovial sarcoma, mesenchymal chrondrosarcoma, neuroblastoma and lymphoma, small cell osteosarcoma. Cytologic appearance of ES is distinctive. Smears are highly cellular and composed of both single cells and groups of loosely cohesive cells with a high nuclear/ cytoplasmic ratio, hyperchromatic nuclei with fine chromatin and one or two nucleoli. The cells always have perinuclear clear cytoplasm suggesting epithelial cell differentiation. Holmer Wright rosettes may be seen in differentiated one. In EES, the distinction of large and small cells is difficult and diagnosis is clinched by immunohistochemical studies. Confirmation of EES should rely on positive stainig for CD99 on IHC analysis. [7] However occasional cases of CD99 expression has been reported in poorly differentiated synovial sarcoma and mesenchymal chondrosarcoma. Ancillary techniques like cytogenetics, RT-PCR, and FISH is helpful. Detection of t (11; 22) (q24, q12) chromososme is highly specific and is found in more than 90% of cases.

Literature reviews of EES shows it to be an extremely rare neoplasm and for that reason it is not feasible to carry out comparative studies to determine the most effective modality of therapy. Early diagnosis is followed by extensive surgery combined with chemotherapy and high dose of radiotherapy for favorable outcome. Reports of neoadjuvant treatment with initial chemotherapy followed by surgery and then later chemotherapy and radiotherapy can also be given. [8] However in metastatic settings, only chemotherapy has shown benefit. Overall ES is an aggressive tumor with a five year survival of being 20% but prognosis of EES is found to be better with a retrospective showed five year survival of 61%.

CONCLUSION

In spite of being a rare entity, Extraskeletal Ewings sarcoma (EES) should be considered in differential diagnosis of primary posterior mediastinal neoplasms in children and young adults and if primitive small round cell tumor is encountered histologically. Additional investigations like immunohistochemistry, cytogenetics, FISH can help in confirmation of diagnosis.

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