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Case Report

Primary Pulmonary Ewing's Sarcoma

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Abstract: Ewing sarcoma is a malignant small round blue cell peripheral primitive neuroectodermal tumor (PNET). Its most common location is flat bones including pelvic bones, ribs and clavicle. It does however occasionally occur in femur and humerus. Rarely Ewing's sarcoma occurs in soft tissues known as Extraskeletal Ewing's sarcoma. A large Ewing sarcoma arising in the chest from rib or vertebral column is known as a skin tumor! We report a case of a 10-year-old girl who presented with right sided lung mass. Radiology, histopathology and immunohistochemistry confirmed the diagnosis of Ewing's sarcoma. Our case highlights the fact that Ewings sarcoma/PNET should be considered in differential diagnosis of patients presenting with lung mass.

Keywords: Ewing's sarcoma, Ewing's sarcoma, Extraskeletal Ewing's sarcoma, small blue cell tumor, Primitive Neuroectodermal tumor, PNET

Case Report

A 10-year-old girl presented to local health facility with fever and right sided chest pain for last 2 weeks. She was given oral antibiotics as there was suspicion of right sided community acquired pneumonia on x-ray chest (fig-1). She took medications regularly without any improvement. This time she was advised empirical anti tuberculous therapy (ATT) as tuberculosis is common in Pakistan and there was no radiological resolution. She took ATT for 1 month, fever subsided but chest x-ray showed right sided pleural effusion that was transudative neutrophilic(Fig-2). Chest intubation was done. Some amount of fluid was drained. She was extubated and referred to Nishtar Hospital Multan.

There, her workup showed pleural fluid negative for AFB and Xpert MTB was also not detected, so, ATT stopped. USG chest was done that reported as moderately thick pleural effusion in right pleural cavity. Fluid was sent for examination that showed transudative predominantly neutrophilic picture. Chest intubation was done followed by CT scan chest with contrast that revealed large enhancing 7x8cm soft tissue mass in right hilar and perihilar region causing obliteration of right bronchus intermedium and right main bronchus with collapse of right middle and lower lobe, suggestive of hydropneumothorax/ neoplasm?." Thoracic surgeon reviewed the case and advised as" currently inoperable due to large size, need chest tube drainage, biopsy and oncology opinion."

She was discharged after extubation and called for follow up after getting opinion from oncologist. Baseline labs were normal except ESR that was 45.

She presented to pediatrics department PIMS Islamabad and got admitted. Workup done that showed mild microcytic hypochromic anemia with ESR 15. Rest of baseline labs were normal. On the basis of CXR (fig), previous CT Scan, USG guided lung biopsy was done. The histopathology examination was inconclusive.

She was referred to pulmonology department where physical examination showed pale looking young girls with decrease air entry in right side of chest and impaired percussion note. Her CT Scan Chest and abdomen was repeated and reported as "findings are of a large lobulated well defined heterogeneous soft tissue density mass in middle mediastinum crossing midline towards right and occupying the rest of right hemithorax causing encasement of right pulmonary artery with hepatic and nodal metastatic disease process, differentials include desmoplastic small round cell tumor or giant cell fibroblastoma" (Figures 1-5).

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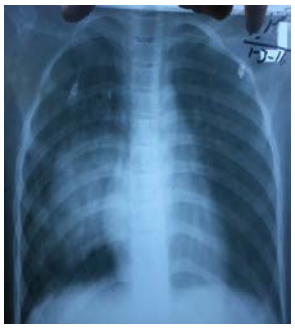

Fig-1

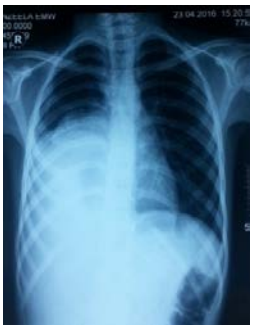

Fig-2

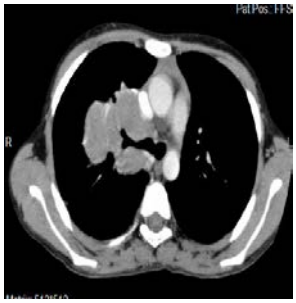

Fig-3

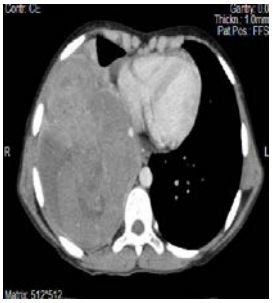

Fig-4

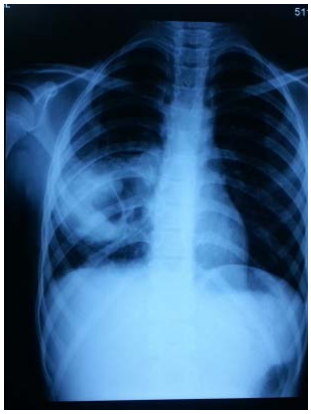

Fig-5

On reviewing her previous record and this CT scan report, trans-thoracic lung mass biopsy was done and sent for histopathological examination which revealed "small round blue cell tumor" positive for CD99+ve compatible with primitive neuroectodermal tumor PNET/Ewing sarcoma (Figure 6 & 7.).

Echocardiography, ABGs were normal. USG abdomen/pelvis reported as right lung mass with hepatic metastasis. She was found to have a hairline fracture i n pelvic bone which was thought to be metastatic rather than primary disease. No erosion of chest bones was noted.

She was started on chemotherapy (vincristin, cyclophosphamide, adriablastin combination) and 6 cycles were done. Her bone scan shows active bone lesions in left hip and proximal end of femur suggesting metastatic spread. Radiotherapy was given on this report. Her latest CXR that was available showed marked improvement after two cycles (fig-5). But after last cycle of chemotherapy she developed chicken pox pneumonia and unfortunately not recovered and succumbed to her disease.

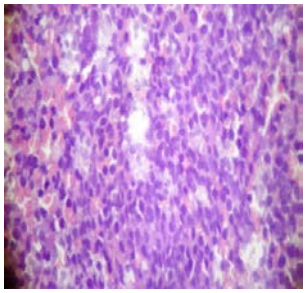

Figure 6: The tumor was comprised of small round blue cells (H&E X 100)

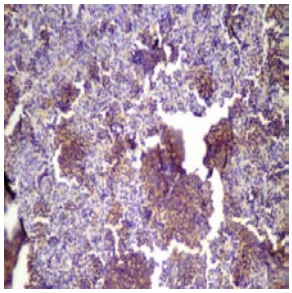

Figure 7: The tumor cells were strongly positive for CD-99 (X 100)

Discussion

Ewings sarcomas are relatively rare primitive neuroectodermal tumors (PNET) that arise primarily from the bone [1]. Extraskeletal Ewings sarcomas have been reported but are very rare. In 1921 James Ewing described it as an endothelioma of bone [2]. These are the second among primary bone tumors [1]. Translocation t 11, 22) (q24; q12) is pathognomonic for

this sarcoma, occurs in 85% of patients and it gives rise to the formation of the EWS-FLI 1 fusion gene [3].
Extraskeletal Ewings sarcoma is very rare. Review of the literature revealed only 17 cases and none from Pakistan. Hammer et al. reported the first case of primary pulmonary Ewing's sarcoma [4]. As per previous case studies, the patients were in the age group of 4- 67 yr and 10 out of these 17 were males [5].
The tumor cells are proliferated small round cells with clear and scanty cytoplasm, oval to round nuclei, finely granular chromatin, and inconspicuous nucleoli[5]. Due to the presence of cytoplasmic glycogen, it is usually PAS positive [5]. Histologic differential diagnoses include small cell carcinoma, neuroblastoma, malignant lymphoma and alveolar rhabdomyosarcoma [5]. This tumor has a strong reactivity to CD99/MIC-2 and with vimentin [5]. In some cases, these tumors may be positive for neural differentiation markers like S-100, NSE and positive for cytokeratins in 20% of cases [5]. Identification of translocation t (11, 22) (q24; q12) by FISH (fluorescent in-situ hybridization) and/or reverse transcription-polymerase chain reaction (RT-PCR) is used to support the diagnosis [1].
In the present case the tumor presented as large lung mass with metastases to the liver. The chest bones were normal showing no erosions and hence Askin tumor was ruled out. There was hair thin fracture of a pelvic bone which favored metastases rather than primary tumor. Due to rarity, of primary pulmonary Ewing's sarcoma, there are no agreed upon specific treatment guidelines. The treatment should be aggressive and consist of surgery followed by chemo and radiotherapy.

Conclusion

We have described an extremely rare case of primary pulmonary Ewing's sarcoma. Though rare, it should be considered in the differential diagnosis of patients presenting with large pulmonary mass without erosions of chest bones.

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KEY FOR CONTRIBUTION OF AUTHORS:

A. Conception/Study Designing/Planning
B. Experimentation/Study Conduction
C. Analysis/Interpretation/Discussion
D. Manuscript Writing
E. Critical Review
F. Facilitated for Reagents/Material/Analysis