

CASE REPORT

ABNORMAL PRESENTATION OF PRIMITIVE NEUROECTODERMAL TUMOR (PNET) IN A YOUNG ADULT

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A rare case presenting with spontaneous hemothorax, severe dyspnoea and a cystic mass in the left hemithorax. Exploration revealed primitive neuroectodermal tumor (PNET).

INTRODUCTION

Askin and colleagues in 1979 were the first to describe a rare tumor of which they coined as malignant small cell tumor of the thoracopulmonary region in childhood.⁽¹⁾ The condition is extremely rare in adulthood.^(2,3) Thoracopulmonary Ewing's sarcoma is thought to overlap considerably with PNET, and the lesions can be grouped together as malignant small round-cell tumors.⁽⁴⁾ In this report we present abnormal presentation of malignant thoracic tumor in a young adult.

CASE REPORT

A 21 year old female presented with a history of trivial chest trauma 5 weeks from admission. She developed low grade fever over a week from the time of the mentioned trauma and progressive

shortness of breath. On examination she was feverish 39 °C, heart rate 125 beats / minute, regular and respiratory rate of 28 / minutes. Pallor was evident and diminished breath sounds over the left hemithorax together with an obvious shift of the trachea towards the right side could not be missed. Routine labs were normal apart from Hb of 7 gm % and TLC of 16000. Chest X ray and Computer tomography confirmed complete opacification of the left hemithorax with severe mediastinal shift as well a cystic well circumscribed swelling at the lower part of the left hemithorax (Fig 1). The patient was resuscitated and was prepared for exploration of her left hemithorax with a primary impression of evacuation of clotted hemithorax and probable coincidental benign cystic swelling for differential diagnosis.

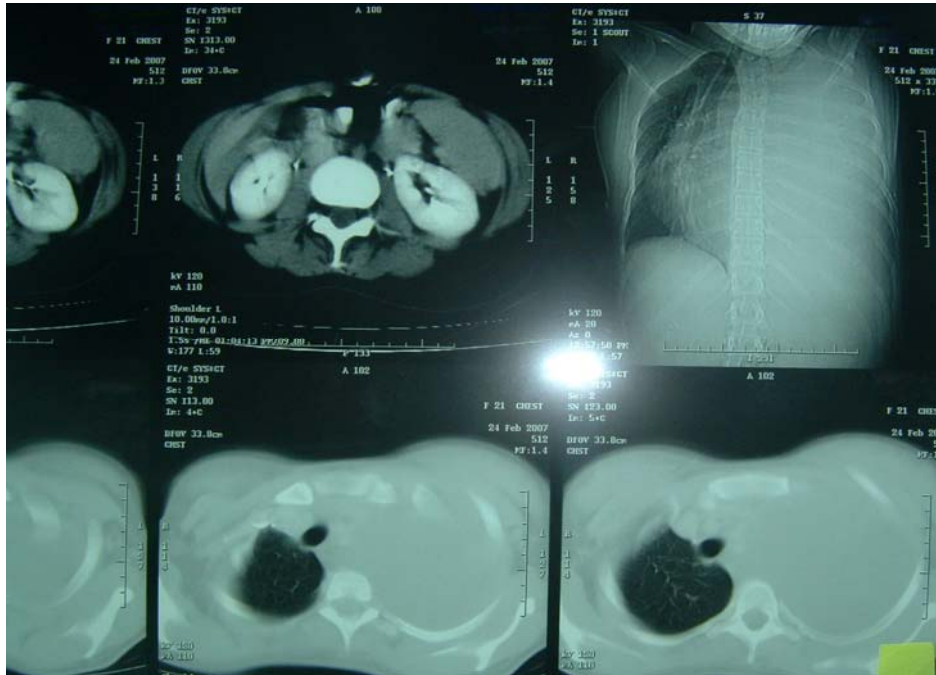


Fig 1

At Exploration a large rather firm mass was found arising from the diaphragm, ruptured and oozing blood (Fig 2). Blood clots were removed from the left hemithorax and the mass could be excised completely and primary closure of the diaphragm edges (Fig 3). Post operative irradiation was given and the patient was followed for one year without recurrence.

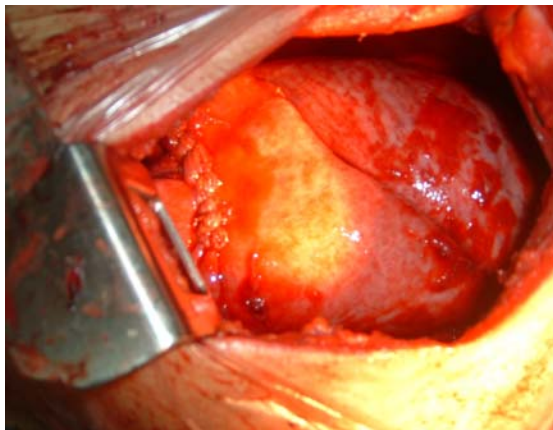
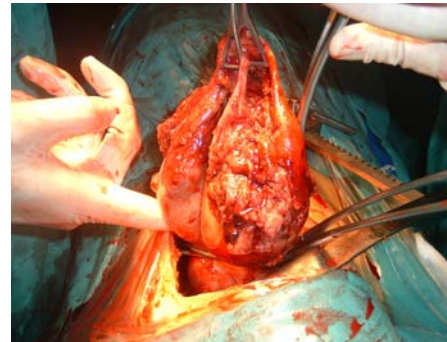


Fig 2



Fig 3

COMMENT

Primitive neuroectodermal tumor lesions are aggressive and usually lethal; they should be considered in the differential diagnosis of thoracic tumors. They were primary described in childhood but are considered rare in adults.

Lesions of PNET are typically painful, invasive thoracic tumors that may develop on and invade the chest wall, lung, or mediastinum. They are generally soft and fleshy, with areas of hemorrhage and necrosis. Although the tumors were classified by Askin and colleagues⁽¹⁾ with thoracic autonomic neurogenic tumors, the PNET cells do not produce biologically active substances detectable in the blood or urine. A similar chromosomal translocation occurring in both Ewing's sarcoma and PNET lesions suggests that these tumors are closely related. Characteristic PNET pathologic findings include Homer-Wright pseudorosettes and uptake of neuron-specific enolase stain.⁽⁵⁾

Although the estimated life expectancy is less than 1 year, aggressive surgery and multimodality regimen has led to disease free survivors.⁽⁴⁻⁶⁾

In our present case we report abnormal presentation of a rare malignant mass presenting in a young adult. The patient was aggressively managed with radical excision followed by radiotherapy. She was followed for the past year without any recurrence. PNET should be aggressively managed with multimodality therapy to improve survival.

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