Case 8303

Primitive neuroectodermal tumor (PNET) of the pleura
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Section: Chest imaging
Case Type: Clinical Cases
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Patient: 43 years, female

Clinical History:
A 43-year-old woman presented with left side chest pain, slowly progressing shortness of breath, decrease of appetite and weight loss.

Imaging Findings:
A 43-year-old woman with a known history of hypertension and diabetes presented with left side chest pain, slowly progressing shortness of breath, decrease of appetite and weight loss. On physical examination the patient showed mild respiratory distress. Left side absent breath sound, stony dull on percussion and increase tactile focal fremitus.

Chest X-ray showed complete opacification of the left hemithorax with mild displacement of the trachea to the right side (Fig. 1). CT examination of the chest was performed and showed large left pleural effusion with complete collapse of the left lung. Pleural-based multiple variable-sized enhancing lesions in left hemithorax. The masses arised from the pleura without invasion of the chest wall or rib destruction (Fig. 2). The radiological differential diagnosis was:
- Pleural metastases.
- Pleural lymphoma
- Sarcomatoid mesothelioma.
- Infection such as TB.
- Spindle cell tumors.

The diagnosis was based on tissue analysis based on the positivity of certain markers in this case like V9, CD99 and CD56 and negativity of others like desmin (D33) and synaptophysin (SY38). Therefore on pathology and after immunochemistry the best possible diagnosis was Pleural PNET (Fig. 3).

Discussion:
In 1979, Askin et al. described a rare, malignant small-cell tumour (Askin tumour) that arises in the soft tissues of the chest wall, occasionally in bone, or rarely, in the periphery of the lung. This neoplasm is now recognized as a type of primitive neuroectodermal tumour (PNET). Askin tumours are seen predominantly in children and young adults and probably develop from embryonal migrating cells of the neural crest [1].

Peripheral PNET arises outside the central and sympathetic nervous systems and differs from central PNET in that peripheral PNET typically expresses high amounts of the MIC2 antigen (CD99) and exhibits highly characteristic chromosomal translocation.

Peripheral PNET is uncommon, and the overall incidence is 1% of all sarcomas. This tumour can occur at any age, although the peak age incidence is adolescence and young adulthood. The incidence in men is slightly higher than in women (ratio of 1.1:1). In general, PNET is a very aggressive neoplasm and it has a poor prognosis, with a 5-
year disease-free survival rate of 45–55%. The most common locations of peripheral PNETs have been the thoracopulmonary region, the retroperitoneal paravertebral soft tissues, the soft tissues of the head and neck, and the intraabdominal and intrapelvic soft tissues and extremities [3].

Traditionally, distinctions were made between classic Ewing sarcoma and PNET, it is now accepted that both are a spectrum of a single neoplastic entity.

Lesions of PNET are typically painful, invasive thoracic tumours that may develop on and invade the chest wall, lung, or mediastinum. They are generally soft and fleshy, with areas of haemorrhage and necrosis [2].

The diagnosis of PNET, which was earlier considered difficult, is now greatly facilitated by the availability of antibodies such as HBA-71 MIC2 (12E7) antigen and O13 that recognise the cell surface antigen defined by the clusters of CD99. Although not specific for PNET or Ewing sarcoma, CD99 is almost always present in these tumours. [2]

**Differential Diagnosis List:** Primitive neuroectodermal tumor (PNET) of the chest wall.

**Final Diagnosis:** Primitive neuroectodermal tumor (PNET) of the chest wall.

**References:**


Figure 1

Description: Complete opacification of the left hemithorax with mild displacement of the trachea to the right side. Origin:
Description: Large left pleural effusion with complete collapse of the left lung. Pleural-based multiple variable-sized enhancing lesions in left hemithorax. Origin:
Description: Large left pleural effusion with complete collapse of the left lung. Pleural-based multiple variable-sized enhancing lesions in left hemithorax. Origin:
Description: Large left pleural effusion with complete collapse of the left lung. Pleural-based multiple variable-sized enhancing lesions in left hemithorax. Origin:
Description: Large left pleural effusion with complete collapse of the left lung. Pleural-based multiple variable-sized enhancing lesions in left hemithorax. Origin:
Description: Malignant small round cell tumour. Neoplastic cells are positive for vimentin (V9) and CD99 (12E7). Origin:
**Description:** (CD99)
malignant small round cell tumou. Neoplastic cells are positive for vimentin (V9) and CD99 (12E7).

**Origin:**
**Description:** (V9)
malignant small round cell tumour. Neoplastic cells are positive for vimentin (V9) and CD99 (12E7).

**Origin:**
Description: (PAS)
malignant small round cell tumour. Neoplastic cells are positive for vimentin (V9) and CD99 (12E7).
Origin: