

Peripheral primitive neuroectodermal tumor of the pleura in a 41-year-old female

Bader A. Alhariqi, MD, Nasser F. Alamri, MD.

ABSTRACT

يعد ورم الأديم العصبي الظاهر البدائي الطرفي من الأمراض النادرة وهو ورم عدواني جداً. ينتمي هذا الورم إلى مجموعة أورام الخلية الصغيرة المدورة وغالباً ما ينشأ من جدار الصدر. نستعرض في هذا المقال حالة مريضة أثبتت تشريحياً أنها مصابة بورم بدائي الأديم العصبي الظاهر المحيطي من الجنب والتي تمت معالجتها في مؤسستنا. لقد حضرت المريضة إلى المستشفى بأعراض آلام بالصدر وفقدان بالوزن. وأوضح التصوير المقطعي بالصبغة للصدر أورام ملونة متعددة للصدر اليساري الجنب مع توغل الورم في الحجاب الحاجز الأيسر. وبعدها خضعت المريضة للعلاج الكيميائي تليها عملية تصغير للورم. ولكن لسوء الحظ توفيت المريضة من تكرار رجوع الورم.

Peripheral primitive neuroectodermal tumor (pPNET) is a rare, very aggressive neoplasm that belongs to a small round cell tumor, and most often arises from the chest wall. Here, we report a female case with proven pPNET who was treated in our institution. She presented with a history of left side chest pain, cough, and significant weight loss. Contrast enhanced CT imaging of the chest showed multiple left pleural-based enhancing masses with left diaphragmatic involvement. She underwent chemotherapy followed by tumor debulking through thoracotomy. However, she died of rapid growth from recurrent local tumors 3 months thereafter.

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From the Department of Radiology, National Guard Hospital, King Abdulaziz Medical City, Riyadh, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Bader Alhariqi, King Abdulaziz Medical City, Mail Code 1222, PO Box 22490, Riyadh 11426, Kingdom of Saudi Arabia. Tél. +966 500013124. Fax. +966 (1) 80112222. E-mail: dr.bader25@gmail.com

Primitive neuroectodermal tumors (PNETs) are rare malignant tumor and belong to small round cell tumors of the central nervous system that mainly occur in children and adolescents. It occur outside the central nervous system in the chest wall, pelvis, and extremities that is called peripheral primitive neuroectodermal tumor (pPNET). Peripheral primitive neuroectodermal tumor of the chest wall belongs to the Ewing's sarcoma family due to their genotypic and phenotypic appearance.¹ Primitive neuroectodermal tumors of the chest wall were first reported by Askin in 1979.² Since then, the tumor involving thoracopulmonary region was termed as Askin tumor. Askin tumors are highly aggressive and metastasize rapidly with poor prognosis.¹⁻³ The pPNET at pleura is very rare, we present this case to emphasize the location of this aggressive tumor.

Case Reports. A 41-year-old lady presented with history of left side chest pain, cough, and significant weight loss. Physical examination revealed decreased breath sound in the left lung with dull percussion of the left hemithorax. Laboratory result was within normal limits. Chest x-ray was initially carried out and revealed a total opacification of the left hemithorax with midline shift to the right side. Initial contrast enhanced computerized tomography (CECT) of the chest showed multiple pleural-based variable-sized enhancing lesions at the left hemithorax with diaphragmatic involvement and large pleural effusion (Figures 1A - 1D). The largest mass measured approximately 4.5 x 4.8 cm. A chest drain was inserted in the left hemithorax, which drained hemorrhagic fluid. Computerized tomography guided biopsies were taken from pleural masses. The immunohistochemical of the biopsies shows positive stain for cluster of differentiation 99 (CD99), CD56, and vimentin. The diagnosis of pPNET of the pleura was then established. After she received chemotherapy, a 3-months follow up was carried out by CECT of the chest (Figures 2A & 2B) that demonstrate the significant tumor regression, and only mild pleural thickening

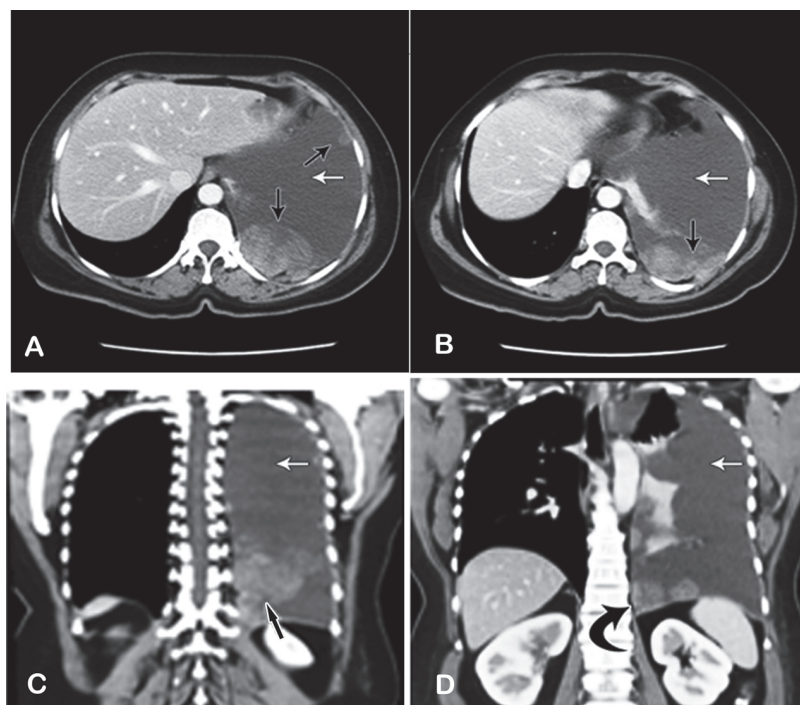


Figure 1 - Axial CT scan of the chest (A, B) and coronal reformat (C, D) shows multiple left pleural-based enhancing masses (black straight arrows) with pleural effusion (white arrows) and diaphragmatic masses (curved arrow).

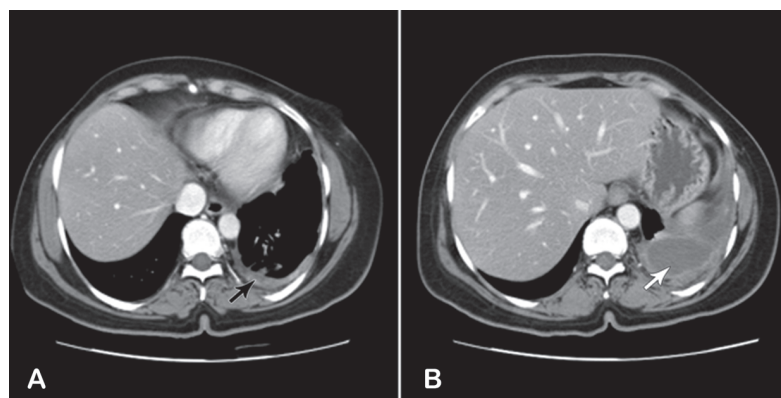


Figure 2 - Axial CT scan A) of the lower chest shows significant regression of the left pleural masses with residual mild pleural thickening (black arrow). B) There is mild loculated fluid (white arrow).

with loculated fluid are present. The next 6-month follow up by CECT of the chest shows aggressive tumor recurrence of the left pleura with areas of necrosis (Figures 3A & 3B). Surgical debulking of the tumor was performed via a left posterolateral thoracotomy. The masses adhering to the diaphragm and lower chest were excised, then, a diaphragmatic repair was performed.

Unfortunately, she died from rapid growth of recurrent local tumor 3 months thereafter.

Discussion. Patient with chest wall PNET usually present with weight loss, cough, fever, dyspnea, chest wall swelling, and hemorrhagic pleural effusion. Diagnostic work up usually include the chest x-ray as

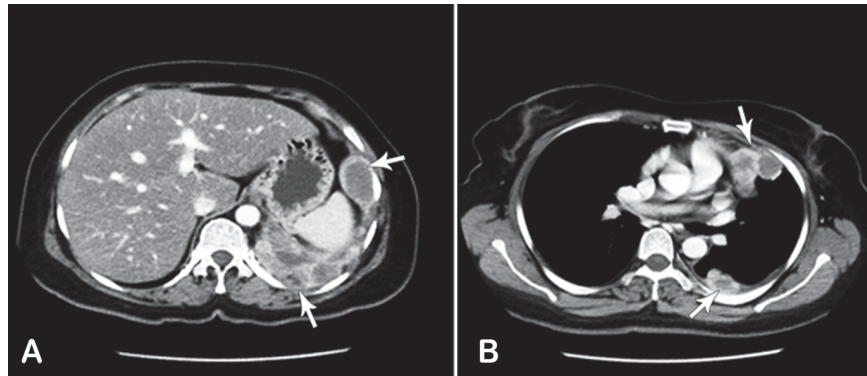


Figure 3 - Follow up axial CT scan (A & B) of the chest shows recurrence of the left pleural-based multiple enhancing masses with areas of necrosis (white arrows).

it will show unilateral opacification of the hemithorax. The CT findings show multiple enhancing masses with areas of necrosis, hemorrhage, and pleural effusion.¹ It is rare to have calcification or lymphadenopathy. On MRI, these tumors appear isointense to hyperintense compared with skeletal muscle on T1 weighted images. On T2 weighted images, it shows high signal intensity with internal heterogeneity. Neither MRI nor CT is reliable for evaluating invasion of the underlying lung, although MRI can predict chest wall invasion.^{1,4} Distant metastasis spreads to lungs, bones, bone marrow, liver, and brain, but it is rare.¹ The main differential diagnosis includes neuroblastoma, rhabdomyosarcoma, non-Hodgkin lymphoma and Langerhans cell histiocytosis. It is important to exclude non-Hodgkin's lymphoma from the differential as its treatment does not include surgery.^{1,2} The main diagnosis is usually carried out by histologic and immunohistochemical analysis. The tumor will show small round malignant cells that contain little cytoplasm, and are arranged in rows in cytologic smears. Using immunohistochemical examination, the tumor is positive for multiple neural markers, such as neuron-specific enolase (NSE), CD99, and vimentin.⁵ Also, there is a characteristic chromosomal translocation for the tumor at $t(11;22)(q24;q12)$.⁶

Treatment should be aggressive including preoperative and postoperative chemotherapy, surgical resection and postoperative radiotherapy.^{7,8} There is high risk for recurrence and metastases. The prognosis was poor with 2 and 6-year survival rates of 38% and 14%. The mean survival rate after recurrence is reduced to 11 months.² Reviewing the English literature, few cases were reported on plural PNET.¹⁻⁸

In conclusion, peripheral primitive neuroectodermal tumor of the pleura is a very aggressive tumor that could occur at any age. The diagnosis requires immunohistochemical workup supported by imaging investigations. The treatment includes chemotherapy, surgical resection, and radiotherapy. The prognosis of pPNET is poor with a high risk of recurrence.

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