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Peripheral Primitive Neuroectodermal Tumor of Chest Wall

KEYWORDS: askin tumor, Ewing's sarcoma, primitive neuroectodermal tumor, small round cell tumor

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ABSTRACT *We describe herein a rare case of a primary primitive neuroectodermal tumor (PNET) in the mediastinum of a 26-year-old man. Grossly, the tumor was located in the right anterolateral lower mediastinum. Transcutaneous fine-needle biopsy (TCNB) revealed small round-cell proliferation. The expression immunohistochemical analysis was confirmed the diagnosis of PNET. He was successfully treated with chemotherapy and surgical resection.*

INTRODUCTION

Peripheral primitive neuroectodermal tumor of the chest wall is a rare malignant tumor usually occurring in children and young adults. The tumor shares a similar histology, immunohistology and cytogenetics to Ewing's sarcoma.

Primitive neuroectodemal tumours (PNETs) are a rare group of tumours which carry identical chromal translocations t (11; 22) (q 24; q 12), to those seen in Ewing sarcoma . Askin originally described PNETs of the chest wall (so called Askin tumours) in 1978[1] , which are peripheral primitive neuroectodermal tumours (PPNETs) associated with the chest wall, ribs, and thoracic cavity. Patients may have pulmonary involvement, but usually in association with a chest wall mass. The condition of PNET is exceedingly rare in adults[2]

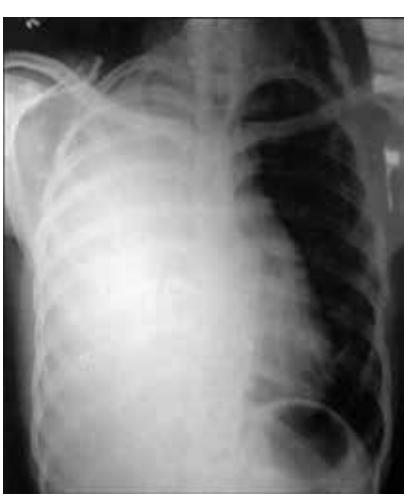
The location of the tumor in the chest wall poses a major challenge with respect to the diagnostic workup and treatment which involves multidisciplinary management. Neoadjuvant chemotherapy is given initially and is followed by complete surgical resectioning of the mass.The chemotherapy of choice for these tumors consists of combinations of doxorubicin, ifosfamide, cyclophosphamide, and vincristine[3].

We report a case of pPNET of the chest wall in a 26-year-old man.

CASE REPORT

A 26-year-old man presented with complaints of swelling in the right side of chest since 3 months and complains of dry cough since 1 month.

Chest X-ray showed homogenous density in right hemithorax with widening of lateral aspect of 8th rib.



Computed tomography (CT) of the chest demonstrated a cystic mass lesion measuring 21 x 15 x 17 cms arising from

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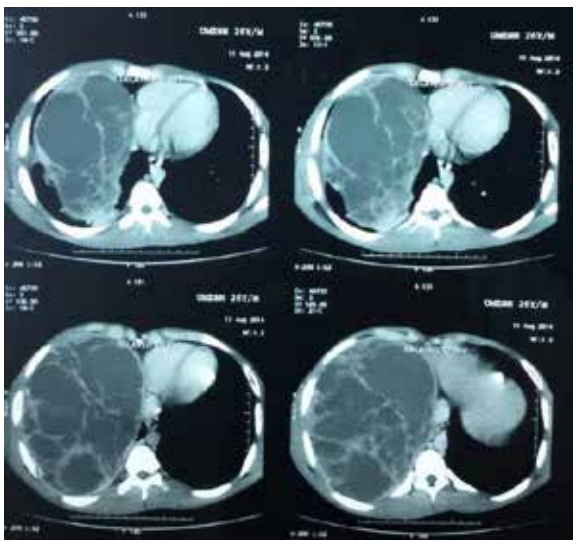
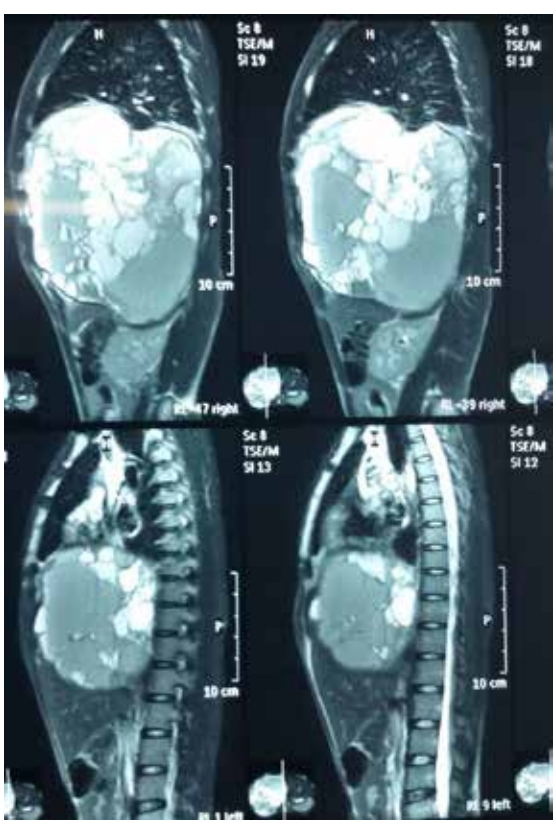
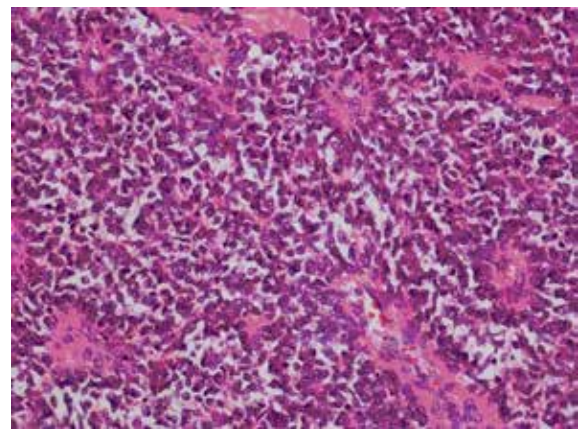
the right anterolateral lower chest and causing expansion, permeative destruction and adjacent periosteal reaction involving both medial and lateral cortices of the right 8th rib.



MRI of the chest showed a multiloculated cystic lesion appearing to arise from the lateral aspect of 8th rib. Thereafter, a fine needle aspiration cytology was performed under MRI guidence.



Histopathologic examination of the cytology specimen indicated malignant, small, round-cell tumor. Some of the cells had irregularly vacuolated cytoplasm secondary to glycogen deposition, which was positive for Periodic Acid Schiff (PAS) stain. The morphologic characteristics and the immunohistochemistry (positive for CD99) were compatible with PNET.



Patient underwent 9 cycles of chemotherapy with vincristine followed by wide local excision of tumor with a part of the 8th rib excised. Postoperative recovery was uneventful. The histological examination of the mass showed tumor with sheets of small round cells and frequent Homer-Wright rosettes. The diagnosis of PNET of the chest wall was established.

DISCUSSION

Askin and Rosai in 1979 described a rare primitive neuroendocrine tumor involving the thoracopulmonary region. Primitive neuroendocrine tumors (PNET) and Ewing's sarcoma are small round cell tumors of neural origin. PNET are rare thoracic tumors found in adolescent or young adults and asymptomatic until advanced stage and hence have poor prognosis [(4)]. They are believed to develop from the peripheral nerves and highly aggressive neoplasms of the thorax with average survival of eight months, although occasional long term survival can be as long as 96 years. The PNET cells do not produce biologically active substances detectable in the blood and urine. They carry identical chromosomal translocations t(11; 22) (q 24; q 12) as seen in Ewing sarcoma. PNET is a rare undifferentiated sarcoma, which is believed to have its origin in embryonal migrating cells from the neural crest, and develops as solitary mass or multiple masses in thoracic area, with rapid growth that may involve pleura. Pain is the only or main symptom in 60% of cases. In the thoracic area, these tumors are invasive and prone to destroying bone, invading the retroperitoneal space, and spreading to lymph nodes, adrenals and liver.

Reports suggest that multimodality treatment is more beneficial, and surgery should be avoided in the first stage of therapeutic management, even if resection is obviously feasible. Surgery should be performed after chemotherapy, even if there is a complete response, because scattered malignant cells may still be found in the scar tissue [(6)]. After curative resection, postoperative chemoradiotherapy regimens are mandatory because of the particularly high risk of both local recurrence and distant metastases [(7,8)].

Survival rates for patients with these tumors have improved dramatically because of advances in chemotherapy and surgical treatment. A retrospective review of twelve patients with malignant chest wall tumors (four Ewing, three PNET, one Askin, two rhabdomyosarcoma, and two neuroblastoma) reported that, at three years, seven (58%) were disease-free and ten (83%) had survived [(5)]. All patients in that study had undergone surgical resection and chemotherapy. The average age at diagnosis was 8.9 years (range, six to sixteen years old).

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Surgery and adjuvant chemoradiotherapy are universally accepted steps in PNET treatment, but more data are needed to support the necessity of neoadjuvant chemotherapy [6,7,8]. Since it is a rare and extremely aggressive malignancy, multidisciplinary protocols should be used to achieve optimal results [9,10]. After completion of treatment, a thorough follow-up at short intervals is indispensable. In cases of recurrence, resection of the malignancy is feasible [11].

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