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Primitive Neuroectodermal Tumor of Mediastinum in an Adult: A Case Report[1]

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A peripheral primitive neuroectodermal tumor (PNET) is a rare and aggressive malignant tumor, which most frequently occurs in children and young adults. The most well described site of origin is the chest wall. We report the case of a primitive neuroectodermal tumor in the anterior mediastinum with unusual CT findings.

Index words : Neuroectodermal tumors, primitive, peripheral
Mediastinal neoplasms
Adult
Tomography, X-ray computed

A peripheral primitive neuroectodermal tumor (PNET) is a rare, malignant small round cell tumor of presumed neural crest origin that arises outside the central and sympathetic nervous system (1). PNETs occur most often in soft tissue of the thoracopulmonary region, pelvis, and lower extremities of children and young adults (1, 2). To our knowledge, PNET of the anterior mediastinum was reported in one case (3). The radiologic appearance of PNET is nonspecific. They usually appear as a large, non-calcified mass with a heterogeneous appearance and variable enhancement (4). Most tumors tend to displace rather than encase adjacent structures such as vessels, the trachea and bronchi, as well as the mediastinum or solid abdominal organs (5). We herein report the case of a PNET of the anterior mediastinum with unusual CT findings in an adult patient.

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Case Report

A 45-year-old man presented with a 5-day history of chest discomfort and worsening dyspnea. He suffered from chest discomfort when performing exercise for the last 20 years (Chest discomfort on exercise of class II from the Canadian Cardiovascular Society). The man has worked as a goldsmith for the last 25 years.

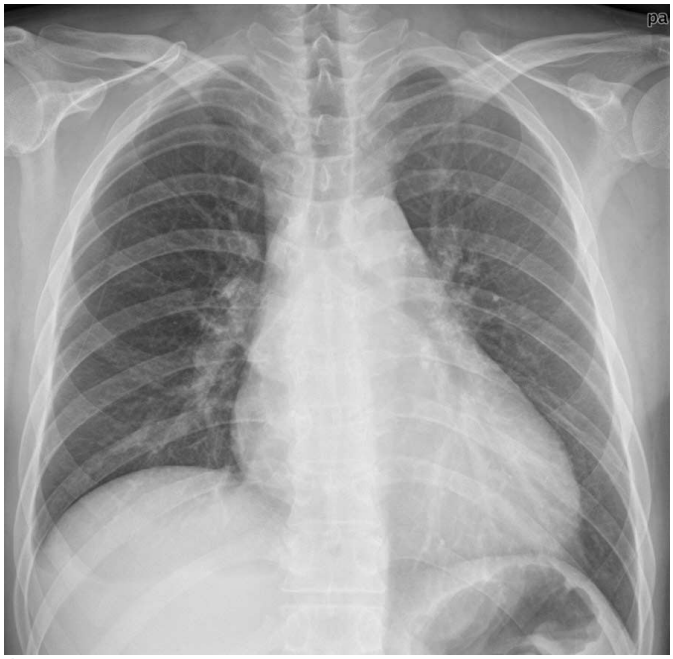
A posteroanterior chest radiograph showed cardiomegaly and lymph node calcifications in the mediastinum and both hilum. A left lateral chest radiograph showed an anterior mediastinal mass and tracheal displacement (Fig. 1A, B). A non-enhanced CT scan of the chest revealed a large lobulated homogenous soft tissue mass without calcification in the anterior mediastinum. On an enhanced CT scan, the mass showed homogeneous enhancement without a low density necrotic portion. The mass extended along the mid-mediastinum and diffusely infiltrated the pericardium and encased vascular structures with diffuse narrowing. The mass did not invade the chest wall or destroy adjacent bony structures (Fig. 1C–G). These CT findings mimic those of lymphoma. Multiple calcified lymph nodes were seen in the mediastinum and both hilum. In addition, small

well-defined centrilobular nodules were observed in both upper lung zones, which was probably due to pneumoconiosis. The PET/CT showed an uneven conglomerating hypermetabolic mass (maximum standardized uptake value, maxSUV 8.4) in the anterior mediastinum that encircled the large vessel and extended along the whole pericardium. No significant hypermetabolic lesions were observed in the lung, abdomen, or pelvis (Fig. 1H). There are multiple mild hypermetabolic lymph nodes (maxSUV 1.2) in mediastinum and both hilum, which are probably inactive granulomatous lesions.

A transthoracic needle biopsy with a 22-guage aspirating needle and automated 18-guage cutting needle were performed at the anterior mediastinum under fluoroscopic guidance. The histopathological and immunohistochemical examination revealed a primitive neuroectodermal tumor. The tumor was a histologically small round cell tumor, which exhibited a positive immunoreactivity for the MIC-2 antibody and FLI1 antibody (Fig. 1I, J). The patient was treated with chemotherapy and radiation therapy for 5 months. Local recurrence of the tumor was found at 3 months after treatment.

Discussion

Peripheral primitive neuroectodermal tumors are small round cell tumors. PNET are classified together



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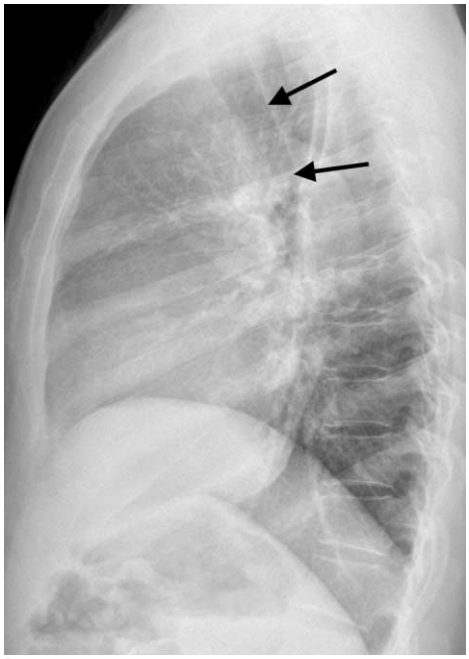

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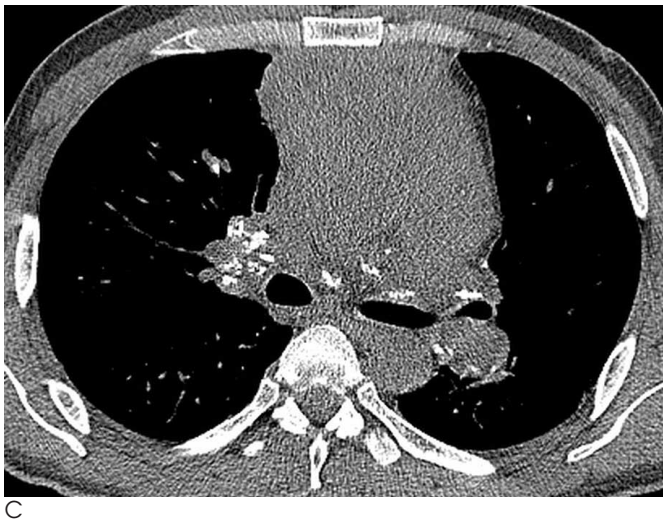

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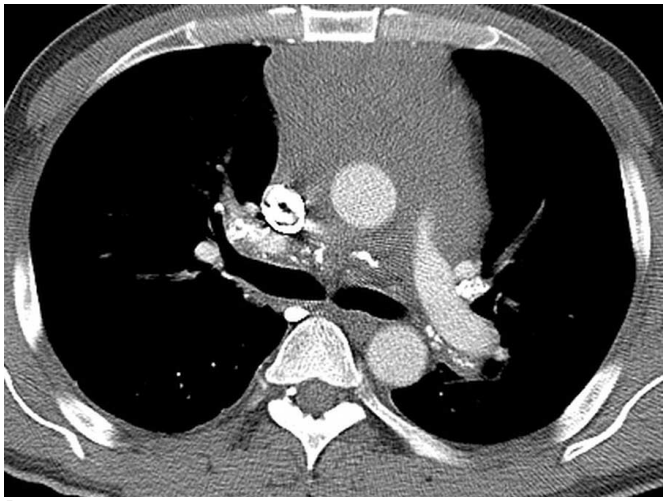
Fig. 1. A. Posteroanterior X-ray of the chest shows cardiomegaly and lymph node calcifications in mediastinum and both hilum.
B. Left lateral chest radiograph shows an anterior mediastinal mass and tracheal displacement (arrows).
C. Axial image of a non-enhanced CT scan shows the lobulated and homogeneous soft tissue density mass without calcification in the anterior mediastinum. Multiple calcified lymph nodes were seen in the mediastinum and both hilum.



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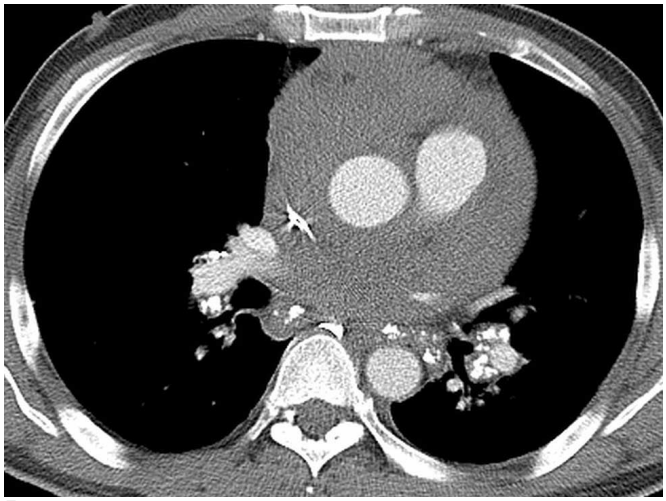

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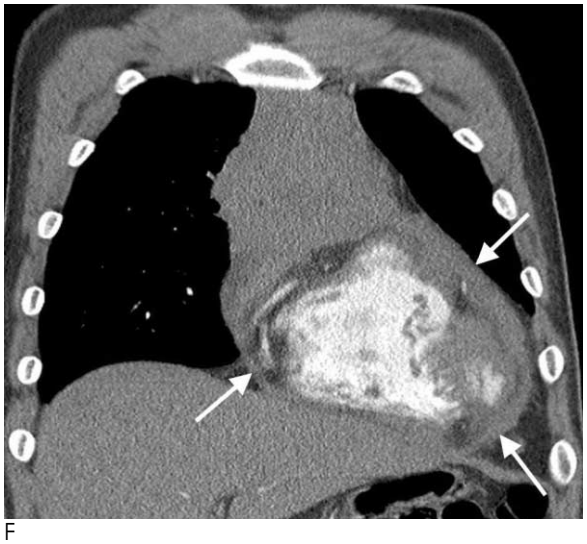

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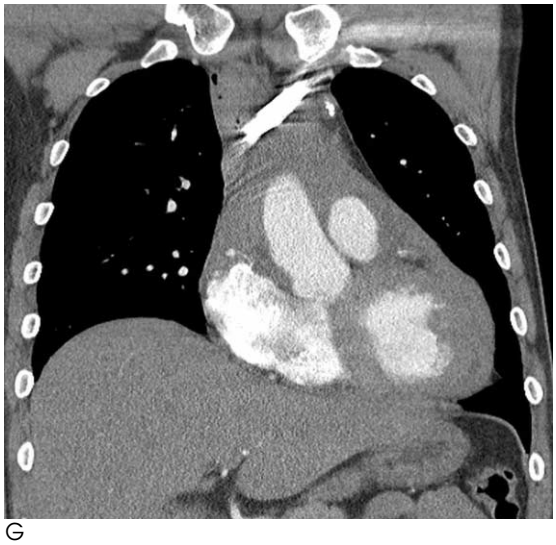

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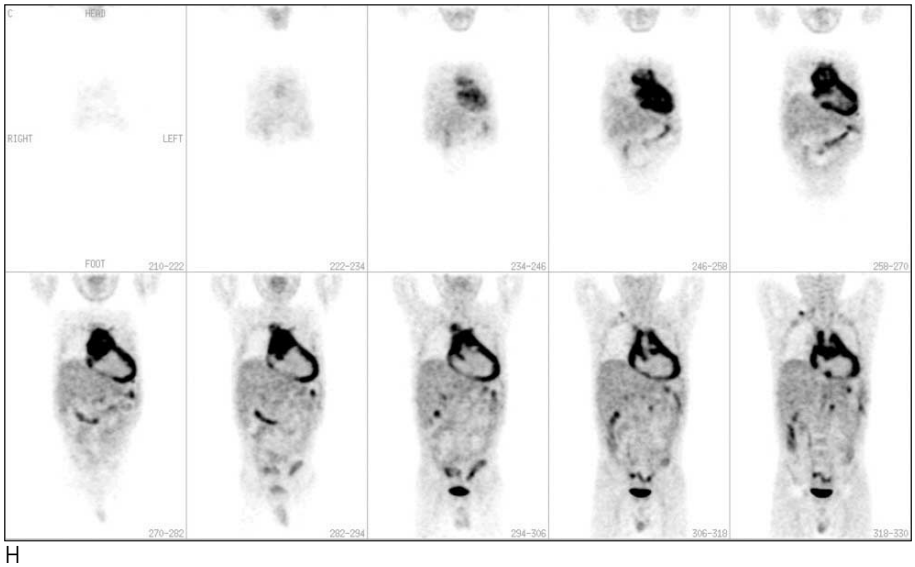
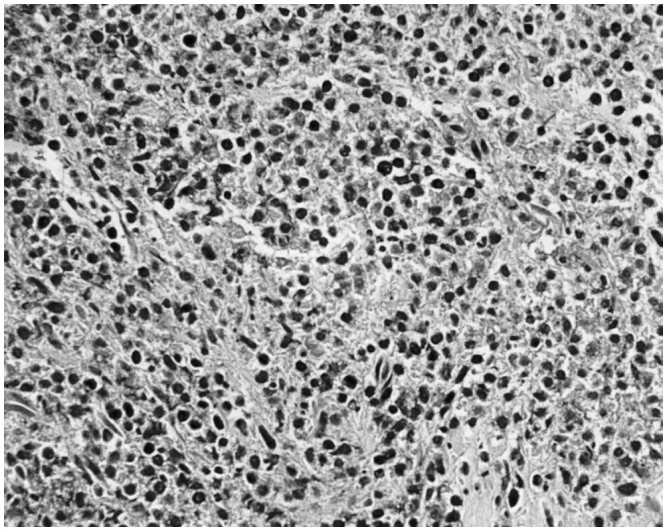


H

Fig. 1. D-G. Axial (D, E) and coronal (F, G) images of the enhanced CT scan show the soft tissue mass with homogeneous enhancement without low density necrotic portion. The mass extended along the mid-mediastinum and diffusely infiltrated the pericardium (arrows) and encased vascular structures with diffuse narrowing.
H. PET/CT shows an uneven conglomerating hypermetabolic mass (maxSUV = 8.4) in the anterior mediastinum that encircles large vessels and extends to the whole pericardium. There are no significant hypermetabolic lesions in the lung, abdomen, or pelvis.



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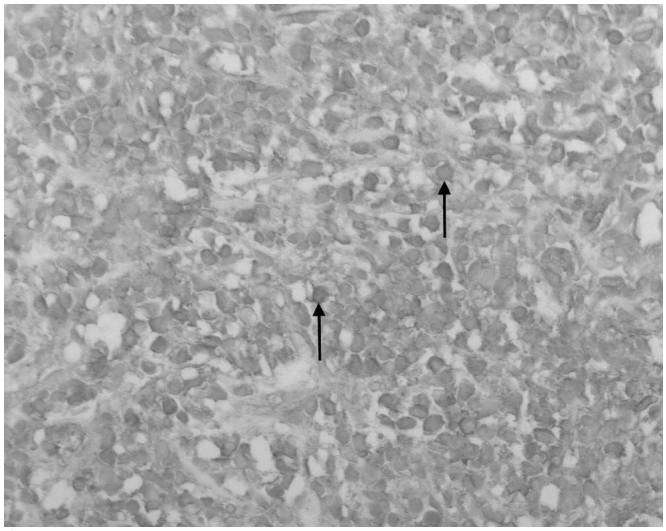

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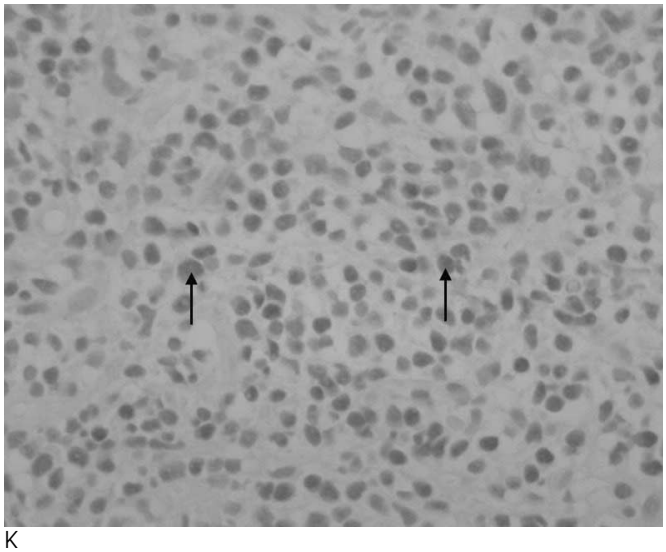

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Fig. 1. I. A photomicrograph shows diffuse proliferation of small round cells with a sparse clear cytoplasm (H & E staining, × 200).
J. A photomicrograph of an immunohistochemistry for detecting MIC2 protein (CD99) shows intense cell membranous immunoreactivity (arrows) (×400).
K. A photomicrograph of an immunohistochemistry for detecting FLI-1 protein shows intense immunoreactivity, which is mainly localized in the nuclei of tumor cells (arrows) (×400).

with Ewing's sarcoma (ES) into the ES/PNET family of tumors. These tumors have in common a poorly differentiated small round cell morphology, high expression of the cell surface glycoprotein MIC2 (CD99), and characteristic t (11; 22) (q24; q12) chromosomal translocation, which produce EWS/FLI1 fusion protein (6). Antibodies against the MIC2 and FLI1 protein are commonly used in the diagnosis of the ES/PNET family of tumors.

PNET makes up approximately 1–4% of all sarcomas (6, 7). Patients are usually children or young adults, although PNET can occur at any age. PNET occurs most often in soft tissue of the thoracopulmonary region, pelvis, thoracic paraspinal region, and lower extremities (1, 2). The radiologic feature of PNET is nonspecific and typically manifests the mass of heterogeneous soft tissue density on CT. Larger tumors (over 5 cm in diameter) contain some low attenuating cystic or necrotic areas. Calcification of the mass is rare. Rib destruction is common in chest wall tumors. On enhanced CT, there is uniform or heterogeneous enhancement of the lesions, with the larger tumors demonstrating more heterogeneous enhancement (3, 4, 8). Most tumors tend to displace rather than encase adjacent structures such as vessels, the trachea and bronchi, the mediastinum, or solid abdominal organs (5). A regional lymphadenopathy is rarely seen at diagnosis (5). Moreover, the MRI signal of PNET is typically intermediate, being isointense or slightly greater than skeletal muscle on T1WI and heterogeneous, but generally hyperintense on T2WI (3, 4, 8).

Treatment consists of resection, augmented by chemotherapy and radiation therapy. Patients with PNET have a poor prognosis with 2- and 6-year survival

rates of 38% and 14%, respectively (9). Poor prognostic factors include metastases at diagnosis and incomplete resection of the mass (10).

In summary, we report here on a rare case of PNET of the anterior mediastinum with unusual CT findings that mimic those of lymphoma in an adult. The lesion was depicted on an enhanced CT scan as a lobulated soft tissue mass without low density necrotic or cystic portion in the anterior mediastinum. As well, it encased vascular structures rather than displacing them.

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성인의 전종격동에 발생한 원시신경외배엽종양: 증례 보고[1]

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성 영 제 · 김 정 숙

원시신경외배엽종양은 대부분 소아와 젊은 성인에서 발생하는 드물고 공격적인 성향의 악성 종양이다. 가장 많이 기술된 원발 병소는 흉벽이다. 저자들은 비전형적인 CT 소견을 보이는 전종격동에서 발생한 원시신경외배엽종양의 증례를 경험하였기에 이를 보고하고자 한다.