Primitive neuroectodermal tumour of the chest wall
(Askin tumour): CT and roentgenographic findings in a
51-year-old male

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Introduction

Primitive neuroectodermal tumours of the chest wall (Askin tumours) are rare malignant neoplasms typically presenting as a large mass based in the chest wall in female children and adolescents (1,2). Askin tumours have been previously reported in a 35-year-old, a 38-year-old, and a 67-year-old man (2-4). We will describe a case of a primitive neuroectodermal tumour (PNET) of the chest wall occurring in a 51-year-old male, supporting the notion that Askin tumours, although uncommon, should be considered in the differential of an adult presenting with a large mass of the thoracic wall.

Case Report

A previously healthy, 51-year-old male presented to his private physician with a chief complaint of cough and chest pain of 1-week duration. Following a 2-week course of antibiotics, the patient returned with similar complaints with additional increasing dyspnoea. Physical examination revealed a palpable mass of the left lower hemithorax and decreased breath sounds at the left base on auscultation. Admission chest radiograph revealed a left pleural-based mass with a large unilateral pleural effusion (Plate 1a,b). Subsequent contrast-enhanced computed tomography (CT) of the chest displayed a large, heterogeneous pleural-based mass with internal regions of necrosis. The mass, measuring 17 × 12 cm in greatest axial dimension and 13 cm in height, extended from the posterior left lung base into the retrosternal pleural reflection (Plate 2a,b). A second smaller mass was noted in the peripheral aspect of the left upper lobe extending to the pleural surface. No mediastinal adenopathy or bony destruction was seen. Technecium-99m bone scan demonstrated increased uptake in the posterior aspect of the left tenth-twelfth ribs. Percutaneous biopsy was unsuccessful for diagnosis. Open surgical biopsy and pleurodesis demonstrated a neoplasm comprised of small, round, blue cells (Plate 3). Consultation with the Armed Forces Institute of Pathology in Washington, DC revealed the typical features of a PNET.

The patient was started on chemotherapy with four cycles of cyclophosphamide/adriamycin to initiate tumour shrinkage in preparation for more extensive resection. After debulking, the patient underwent three cycles of chemotherapy with cytoxan/MESNA followed by three cycles of high dose therapy with ifosfamide/VP16/MESNA. The patient has remained relatively asymptomatic, complaining only of left axillary numbness and intermittent nausea.

The most recent CT revealed post-surgical volume loss of the left hemithorax with persistent pleural thickening. No parenchymal or hepatic metastases were noted.

Discussion

Primitive neuroectodermal tumour of the chest wall was first described as a specific pathological entity by Askin et al. as occurring in children and young adults (5). The histologic characteristics which distinguish Askin tumours are the presence of neurosecretory granules on electron microscopy and neuron-specific enolase immunohistochemical markers (2,6). These neoplasms tend to achieve large sizes prior to presentation. A thoracic mass, often accompanied by pain, is the most common clinical manifestation. Additional symptoms include cough, fever, weight loss and dyspnoea (3).
Plate I  (a,b) A and lateral chest radiograph displaying smooth bordered mass occupying the left posterior lung base and adjacent thoracic wall. It is difficult to determine the precise anatomic tissue of origin by plain film criteria. Lobulated mass in the peripheral aspect of the left upper lobe is also seen, indicative of a pulmonary metastasis.

Plate 2  (a) Contrast-enhanced CT scan at the lung bases shows mass with heterogeneous regions of high and low attenuation, likely representing foci of tissue necrosis adjacent regions of haemorrhage and pronounced vascularity. (b) Smaller upper lobe mass displays more uniform density.

The radiologic characteristics of Askin tumours are non-specific. The initial radiograph generally reveals a large soft tissue density based in the chest wall, often accompanied by and ipsilateral pleural effusion. Rib destruction may also be present. However, bony involvement, if present, may only be identified following radionuclide examination (3). Typical CT findings, as reinforced by this case, include a large mass based in the chest wall with intrathoracic extension. The mass often has a heterogeneous appearance due to regions of central necrosis (1–3). Pulmonary metastases are common, as are
extra-pulmonary sites, such as mediastinal and retroperitoneal lymph nodes, extra-thoracic skeleton, liver, adrenal glands, and sympathetic nerve chain (2,7). Characteristic magnetic resonance imaging findings include heterogeneously bright signal intensity on T2-weighted studies and heterogeneous signal, predominantly greater than adjacent skeletal muscle, with regions of high and low signal intensity representing haemorrhage and necrosis on T1-weighted studies (1).

The Askin tumour has been described as a PNET of the chest wall occurring in children and adolescents. However, it is now apparent that the discussion of Askin tumours can no longer be limited to this population. This is the fourth reported case of an Askin tumour occurring in a patient above 35 years of age and the eldest case to date in the English literature. The eldest case is that of a 67-year-old man reported in the French literature (4).

Differential diagnosis of a large necrotic mass based in the chest wall in an adult has traditionally included lymphoma, sarcoma, mesothelioma and metastasis. The advanced age of this patient not only makes this case unique, but more importantly, reinforces the notion that the Askin tumour, although rare, must now be included in the above gamut to allow the most prompt, efficacious management to ensue.

References

