

Askin Tumor: A Case Report with Emphasis on Diagnostic Approach

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Abstract

Askin tumor is a rare, extremely malignant round cell tumor of the chest wall. The exact incidence is not known due to its rarity. This neoplasm is currently grouped under Ewing's family of tumors (EFT's). Metastasis to the sympathetic chain is a unique feature of Askin's tumor.

Keywords: Tumor, PNET, Computed tomography (CT).

Introduction

Askin's tumor is a rare, malignant undifferentiated small-round-cell sarcoma of the soft tissues of the chest wall, occasionally arising in the bone and rarely in the periphery of the lung.¹ This tumor probably develops from embryonal migrating cells of the neural crest² and is now recognized as a type of primitive neuroectodermal tumor (PNET). Askin's tumor is currently grouped under the Ewing's family of tumors. Though the frequency of Ewing's sarcoma and PNET among childhood tumors is 2%, the exact incidence of Askin's tumor is not clear because of the rarity of the disease.

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Case Report

A 24 years old male presented with a 5 month history of pluritic chest pain, cough with dyspnea, chest wall swelling, recurrent pleural effusion and weight loss.



Fig. 1a: Frontal chest radiograph showing left hemi thorax homogenous opacity, partial destruction of 6th rib and shift of trachea and mediastinum to opposite side.

The frontal chest radiograph on admission revealed a homogenous opacity in the left hemithorax with shift of the mediastinum and trachea to the opposite side. Erosion of the left 6th rib in its lateral aspect was noted (Fig.1a).

Computed Tomography (CT) scan of the chest showed a large heterogeneously enhancing mass in the left chest wall causing collapse and consolidation of the underlying lung. Destruction of the adjacent ribs, involvement of the mediastinal pleura and pericardium of the left atrium was noted (Fig.1b).



Fig. 1b: Contrast enhanced CT of chest shows large heterogeneously enhancing left chest wall mass involving mediastinal pleura causing collapse consolidation of underlying lung with rib destruction (arrow), and mediastinal shift to right.

A well defined rounded peripherally enhancing lesion in the left para-vertebral region suggestive of metastasis to the sympathetic chain was also noted (Fig. 1c). Ultrasound guided biopsy was done and histopathology of the tumor revealed solid nests and sheets of monotonous, primitive, small necrobiotic round cells (Fig.2). Both radiological and histopathological findings

together suggested the diagnosis of Askin's tumor (pnet).



Fig. 1c: Contrast enhanced CT of chest shows well defined rounded peripherally enhancing lesion in the left paravertebral region suggestive of sympathetic chain metastasis (arrow).

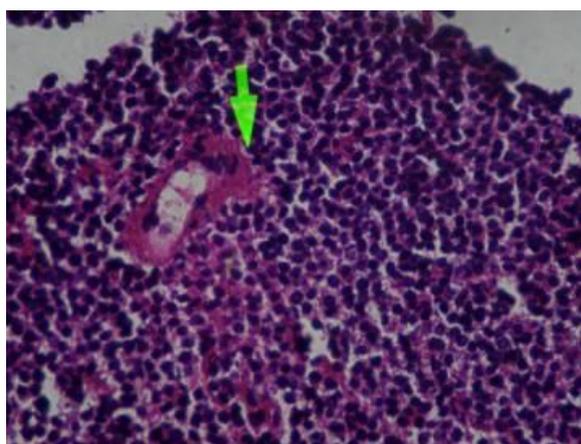


Fig. 2: Pleural biopsy histopathological examination shows diffuse proliferation of small necrobiotic round cells: HE stain, x 400.

Discussion

Askin's tumor is seen predominantly in children and young adults.¹ This tumor commonly arises from the soft tissue with variable rib destruction. Clinical presentation of these tumors varies depending on their location. Interestingly these tumors show propensity to metastasize to the nervous

tissue, which may be related to their neuroectodermal origin.

The CT features of Askin's tumor include a heterogeneously enhancing large soft tissue mass lesion of the chest wall with variable rib destruction. Local extension into the mediastinum and lung is common. More than 90% of the cases show pleural effusion. The combination of a thick, almost circumferentially based pleural mass with rapidly accumulating loculated pleural collections is unique in that very few non-infectious processes can do this.⁴ The swift growth of a pleural-based mass, with multiple sterile pseudotumors of pleural fluid unilaterally in a young patient, may help to suggest the diagnosis of Askin's tumor radiologically. This tumor usually metastasizes to the lung, bone and mediastinal lymph nodes with unique metastases to the sympathetic chain. MR findings include a heterogeneous mass lesion which shows predominantly high signal intensity on T1 and T2WI and which enhances with gadolinium. Histological features include small blue cell tumors with hyperchromatic nuclei and scanty cytoplasm which is seen in many other tumors including Ewing's sarcoma, rhabdomyosarcoma, neuroblastoma and lymphoma, all of which are included under the category of malignant small cell tumor. Immunohistochemistry or electron microscopy helps in differentiation of PNET from other tumors in malignant small cell group.⁵

The pathological distinction of Askin's tumor [PNET] from Ewing's sarcoma had previously been important, as the prognosis of these lesions were reported to be significantly different from Ewing's sarcoma. More recent studies however, have failed to

demonstrate any significant differences in outcome among these tumors,⁶ most likely as a result of the recent development of intensive chemotherapy. Recent studies have revealed that the pathognomonic translocation between the *EWS* gene on chromosome 22 and an *ETS* type gene, most commonly the *Fli 1* gene on chromosome 11, is implicated in more than 95% of Ewing's sarcomas, PNETs and Askin's tumors. Therefore, these lesions have currently been grouped under the same entity, dubbed the Ewing's family of tumors (EFTs).³

These tumors have similar response to therapy and all contain the protein product of MIC2 gene on their surface. All these tumors also have the same balanced translocation between chromosome 11 and 22.³ Treatment strategies include surgical resection when possible, chemotherapy, radiotherapy or bone marrow transplantation. The prognosis is variable.

Conclusion

Askin's tumor is an extremely malignant neoplasm of the chest. The rapid growth of a chest wall mass, with multiple sterile pseudotumors of pleural fluid unilaterally and typical metastatic nodules in the sympathetic chain in a young patient suggests the diagnosis of Askin's tumor.

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