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***Case report-Olgu sunumu***
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# Askin Tumour: A rare childhood thoraco - pulmonary malignancy of Ewing's family of tumours

## *Askin Tümörü: Ewing tümör ailesinin nadir görülen çocukluk çağı torakopulmoner malignitesi*

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**Abstact**

Primitive neuro-ectodermal tumour of chest wall or the Askin tumour is a rare, malignant small cell neuroepithelioma that arises from the chest wall or lung. It is seen predominantly in children and adolescents. Although its appearance by light microscopy is similar to Ewing's sarcoma, rhabdomyosarcoma, neuroblastoma and lymphoma, which account for bulk of chest wall neoplasms in this age group, yet the Askin tumour is a distinct entity. We present a case of Askin tumour in a child, a rare malignant tumour in the thoraco-pulmonary area with highly aggressive clinical course.

**Keywords:** Askin Tumour, primitive neuro-ectodermal tumour, malignant small cell neuroepithelioma

**Özet**

Göğüs duvarının primitif nöro-ektodermal tümörü veya diğer bir deyişle Askin tümörü nadir görülen bir küçük hücreli nöroepitelyoma olup göğüs duvarı veya akciğer kökenlidir. Ağırlıklı olarak çocuk ve adolesanlarda saptanır. Her ne kadar ışık mikroskopisindeki görünümü Ewing sarkomu, rabdomyosarkom, nöroblastom ve lenfoma gibi bu yaş grubunda çoğunlukla görülen göğüs duvarı neoplasmlarına benzese de Askin tümörü farklı bir antitedir. Bu makalede yüksek derecede agresif klinik seyirli, torakopulmoner alan yerleşimli nadir bir malignite olan Askin tümörlü bir çocuk hasta sunulmuştur.

**Anahtar sözcükler:** Askin tümörü, primitif nöroektodermal tümör, malign küçük hücreli nöroepitelyoma

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## Introduction

Primitive neuro-ectodermal tumour (PNET) and Ewing's sarcoma are biologically related malignant small round cell tumours of the soft tissues and bones [1]. Both have been categorized into a group known as Ewing's family of tumours because of their immunohistochemical, ultrastuctural and molecular similarities [2]. PNET are malignant tumours of central nervous system (CNS) usually found in infants, children and young adults. But they can also occur sporadically outside the CNS (peripheral-PNET or peripheral neuroepithelioma) and the chest wall is most common of these sites [3]. PNETs of the chest wall were originally reported by Askin et al [4] in 1979 in 20 children and adolescents with a mean age of 14 years. All the patients had malignant small cell

tumours of thoraco-pulmonary region especially within thorax and involved chest wall and pleura. These tumours histologically resembled Ewing's tumours. Cytogenetic, ultrastructural and biologic studies with characteristic t (11;22) (q24;q12) translocation, confirm the commonality of Ewing's sarcoma, PNET and Askin tumour, which together are referred to as the PNET or Ewing's family of tumours [2].

## Case report

A seven year old female child presented to our hospital with the chief complaints of swelling and dull aching pain at the right side of chest wall for one month along with decreased appetite, low grade fever, and difficulty in walking for 7 days. There was no history of any trauma or cough with expectoration, palpitation or hemoptysis. The child was pale and moderately nourished. There was no evidence of any lymphadenopathy. Pulse was 120/min, regular, respiratory rate was 34/min and blood pressure was 100/70 mm of Hg. Air entry on right side of chest was reduced with stony dullness on percussion. A non compressible, fixed, non mobile swelling was evident on right side of chest wall. Liver was palpable 5 cms below right costal margin, with mildly palpable spleen. The patient had waddling gait. Plain chest radiograph showed homogenously opaque, large soft tissue mass in right hemi-thorax with local rib erosion and minimal effusion. Ultrasonography revealed large hypoechoic lesion with cystic areas seen in whole of right thoracic cavity with no flow on color doppler, pushing down the right dome of diaphragm and liver along with few hypoechoic lesions in the subcutaneous and muscle plane abutting the underlying rib (Figure 1a). Contrast Enhanced Computed Tomography scan (CECT scan) demonstrated destruction and sclerosis of the anterior end of 5th rib on right side with a large soft tissue mass, mainly intrathoracic with small extra-thoracic component. The mass was showing heterogenous enhancement with no calcification in it. There was expansion of intracostal spaces and mediastinal shift to left side with the diaphragm pushed downwards. Minimal effusion was also seen (Figure 1b). Fine needle aspiration cytology (FNAC) (Figure 2a) showed dyscohesive small round cells forming rosettes at places with scant cytoplasm, hyperchromatic nuclei, inconspicuous nucleoli and numerous mitotic figures. A diagnosis of non lymphomatous malignant small round cell tumour was made. Biopsy revealed sheets of small round hyperchromatic cells with evidence of crushing, scattered Homer Wright pseudorosettes, high mitotic index with many atypical mitosis and infiltrating fibrocollagenous tissue (Figure 2b). Differential diagnosis of PNET, Embryonal rhabdomyosarcoma and lymphoma was considered. On immunohistochemistry the tumour cells expressed vimentin, Neuron specific enolase, synaptophsin (Figure 2c), CD 99, (Figure 2d) , while LCA and myogenin were negative (lymphoma and embryonal rhabdomyosarcoma were ruled out) and hence conforming to the diagnosis of PNET. The patient received aggressive chemotherapy (Adriamycin, vincristine, cyclophosphamide). Subsequent chest radiograph showed marked reduction in tumour size.

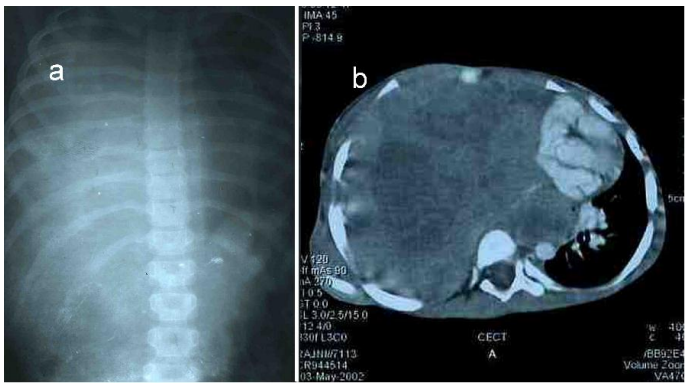


**Figure 1a. Chest X-ray revealing homogenous opacity on right side; b. CECT showing subcutaneous nodule along with right sided opacity.**

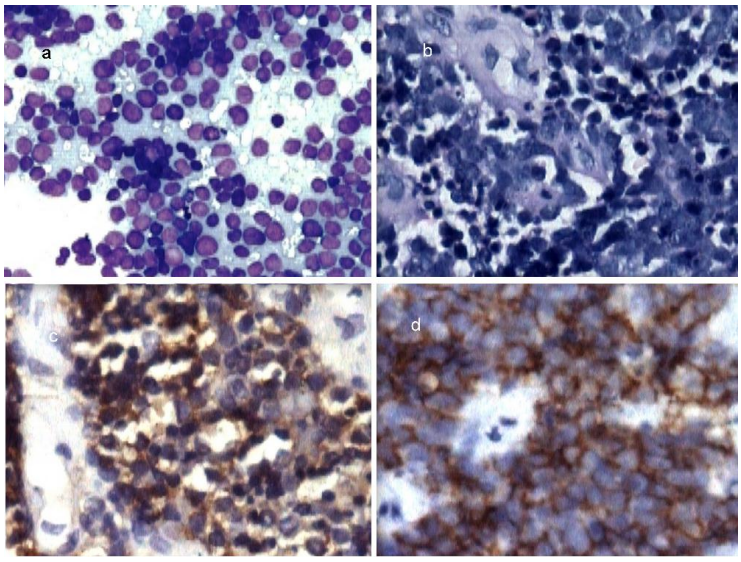


**Figure 2a. FNAC at 200x; b. Histopathology microsection revealing small round cells H& E, 400x; c. Synaptophysin positivity; d. CD99 positivity.**

## Discussion

The Askin tumour is a distinct clinical and pathological entity that is described as malignant neuro-ectodermal tumour of thoraco-pulmonary region (a type of PNET) arising from the soft tissue of chest wall, occasionally from the rib cage and rarely from the periphery of lung [5]. Immunohistochemistry and electron microscopic features can distinguish it from other small cell tumours of the childhood and young adulthood. However, histologic overlap between these neoplasms usually makes differentiation difficult, especially when light microscopy alone is used [6]. These tumours can occur at any age but are more frequent in children and young adults. The lesion has an insidious onset, leading to large size at presentation. Distant metastases are identified in 20-25% of newly diagnosed patients and are the most important adverse prognostic factors especially with bone or bone marrow metastasis [3]. Metastasis along the sympathetic chain has also been reported and this propensity to metastasize to nervous system may be related to its neuroectodermal origin [6]. Lymphadenopathy is uncommon [3]. The tumours have large soft tissue component with areas of necrosis and hemorrhage. The radiographic appearance of Askin tumour is non-specific. CT scan usually shows a heterogeneously enhancing thoraco-pulmonary mass arising from soft tissue of chest wall. Rarely areas of calcification may be seen. Magnetic resonance imaging is advocated for knowing the extent of tumour extension into the chest wall and the lung [5].Tissue biopsy remains the most important diagnostic tool, with imaging modalities playing a complimentary role to delineate the extent of the disease before planning the treatment and for eventual follow up. Treatment includes surgical resection (if possible) followed by aggressive chemotherapy which produces excellent results as in case of Ewing Sarcoma/ PNET elsewhere [7].

In our case the age of presentation, short duration of pain in thoracic region, chest radiograph and CECT scan were suggestive of an extrapleural thoracic wall tumour of non-osseous origin. FNAC and biopsy complimented by immunophenotyping conclusively proved the diagnosis. In recent times immunophenotyping has made distinction more objective with the demonstration of CD 99 positivity (while LCA and muscle markers are negative). A small proportion of rhabdomyoblasts may also be positive hence positivity for 2$^{nd}$ neural markers (S-100, synaptophysin), negativity for muscle markers helps to confirm the diagnosis [2]. In conclusion, Askin tumour should be considered in the differential diagnosis of small cell tumour located at thoracic wall regardless of age and patient should be aggressively treated.

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