# Askin tumor- in 2 year old child- Case report

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**ABSTRACT:** Askin's tumor is a primitive neuroectodermal tumor developing from the soft tissues of the chest wall. Its diagnosis approach is complex and requires a multidisciplinary team. Given the rarity of this entity, no regimen has been validated in the literature. We report a case of Askin's tumor, treatment of Askin's tumor should be multimodal, requiring discussion in multidisciplinary tumor working groups.

**Key words:** Askin tumor, diagnostic imaging differences, immunohistochemical examination

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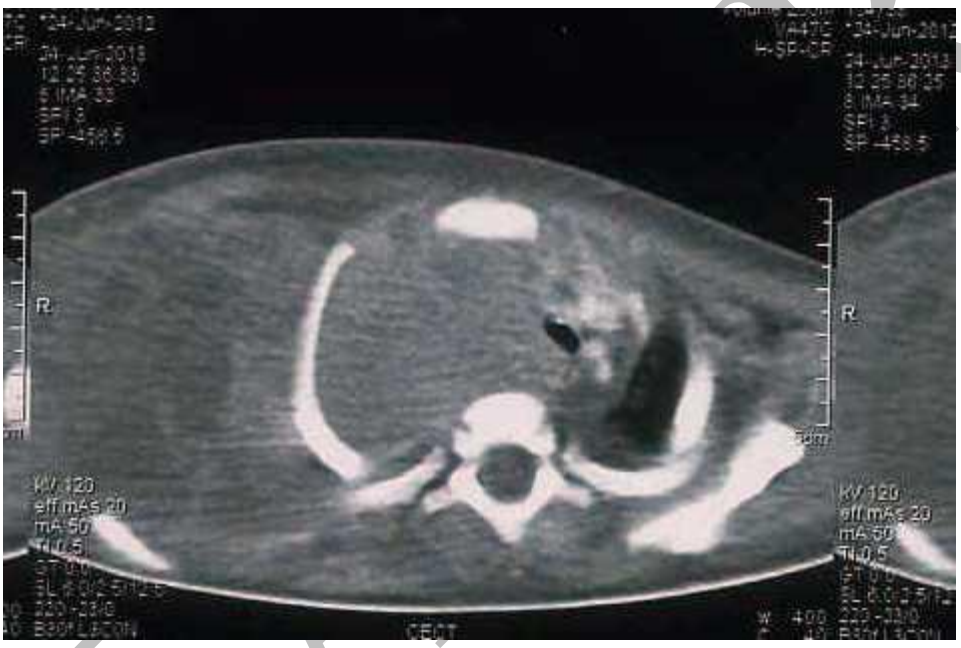
**INTRODUCTION:** Askin tumor has been defined by Askin and Rosai in 1979. This malignant round cell tumor, which originates from the soft tissue of the chest wall, is also called extra skeletal Ewing sarcoma or peripheral primitive neuro-ectodermal tumor (PNET) [1]. Although once accepted as distinct entity, today Ewing sarcoma, Askin tumor and PNET are all considered as members of the Ewing family of tumors and if localized in the thoraco-pulmonary region, they are defined as Askin tumor[2]. The frequency of Ewing sarcoma and PNETchildhood tumors is 2% and the incidence of Askin tumor is not clear as the disease is very rare. Very few clinical studies performed, after the report of Askin et al. in 1979, are available [3]. Surgery, radiotherapy and chemotherapy, isolated or in combination is used as treatment modality. Prognosis is generally poor. We are presenting a case of Askin tumor in a 2 year old child with a short duration of history.

**CASE REPORT:** A 2 year old male child presented with fever, cough and right axillary swelling since 3 days. Fever was on and off, moderate in intensity. Swelling was sudden in onset, gradually progressing in size and there was no history of trauma or previous hospitalisation. Child was highly irritable and constantly crying. Laboratory investigations were within normal limits except increased serum LDH. Heart rate was

116/min and respiratory rate was 62/ min. Chest examination revealed decreased breath sounds bilaterally. CECT scan revealed a large soft tissue enhancing mass with cystic area involving mediastinum, right axilla, right upper arm and right prescapular area along with right sided pleural effusion with underlying collapse of lung (Figure 1).

Figure 1: Contrast-enhanced CT imaging displayed a large, heterogeneous pleural-based mass



Mediastinal vasulature appear compressed and displaced. Trachea and bronchi were lifted anteriorly. No mediastinal or axillary lymphadenopathy and distant metastasiswas found.An abdominal CT scan did not show any metastatic localization. USG guided FNA was done. FNAC smears showed malignant cells present in small groups and forming rosettes at places and dispersed singly alongwith some multinucleated tumor giant cells. These cells were small round showing nuclear pleomorphism having high N:C ratio and granular nuclear chromatin with small nucleoli. Cytoplam was scanty to moderate with vacuolation at places (Figure 2). Numerous mitotic figures were also seen (Figure 3).

**Figure 2:** FNAC Smears showing malignant cells present in small groups and forming rosettes at places and dispersed singly (Giemsa, 400x)

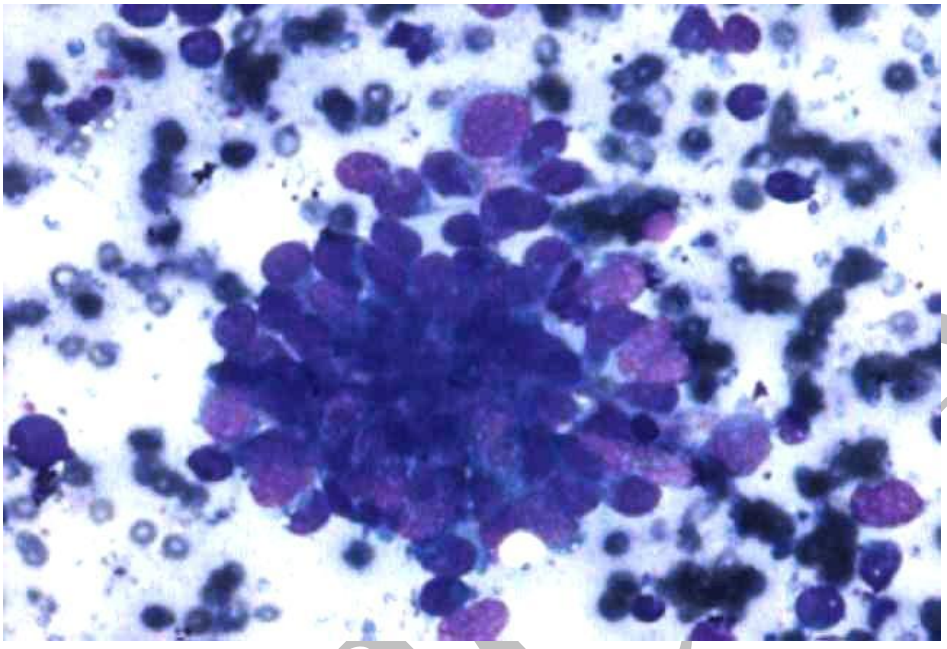
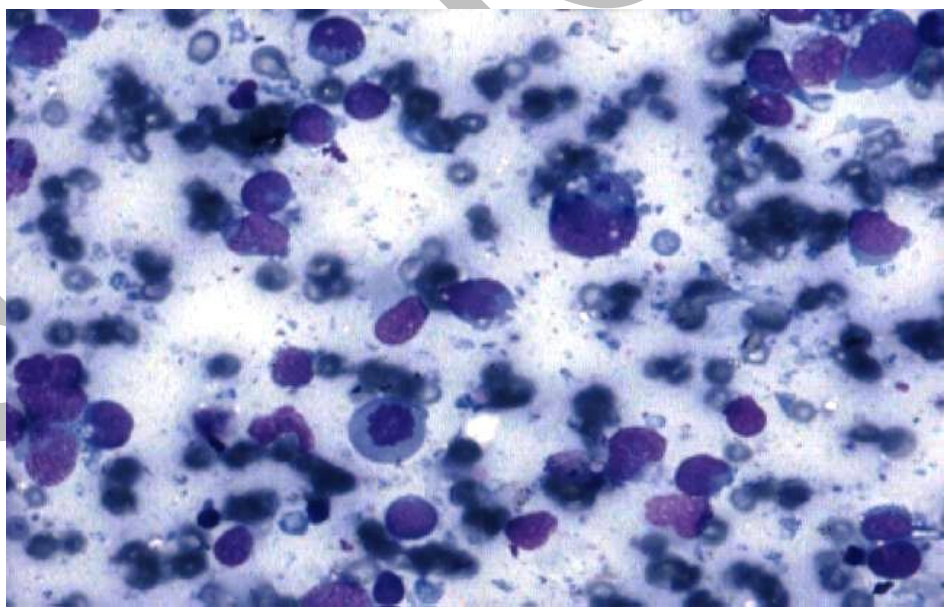


Figure 3: FNAC Smears reveal mitosis (Giemsa, 400x).



Probable diagnosis of PNET was made. Immunohistochemistry on cell block preparation showed positivity for CD99, NSE and synaptophysin (Figure 4, 5). On the basis of these findings

diagnosis of Askin tumor was made. Patient was put on chemotherapy but due to difficulty in respiration he died within 2 days after starting treatment.

Figure 4: Immunohistochemistry on cell block preparation showed positivity for CD99 (IHC, 400x).

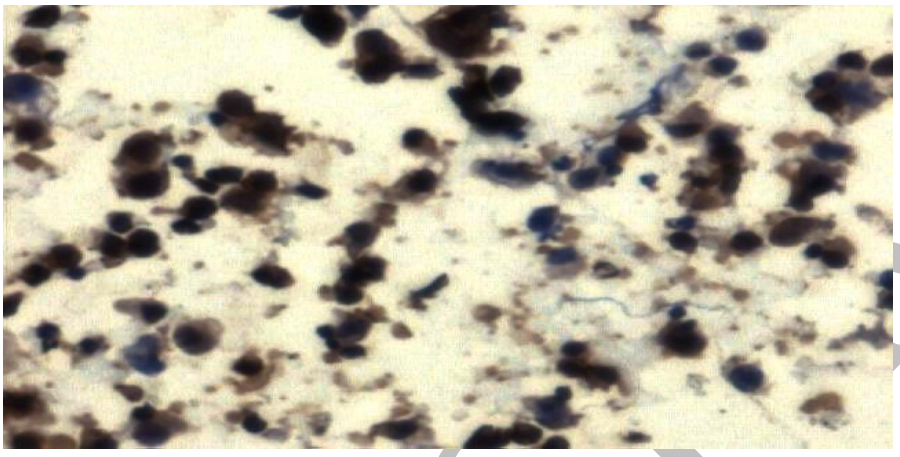
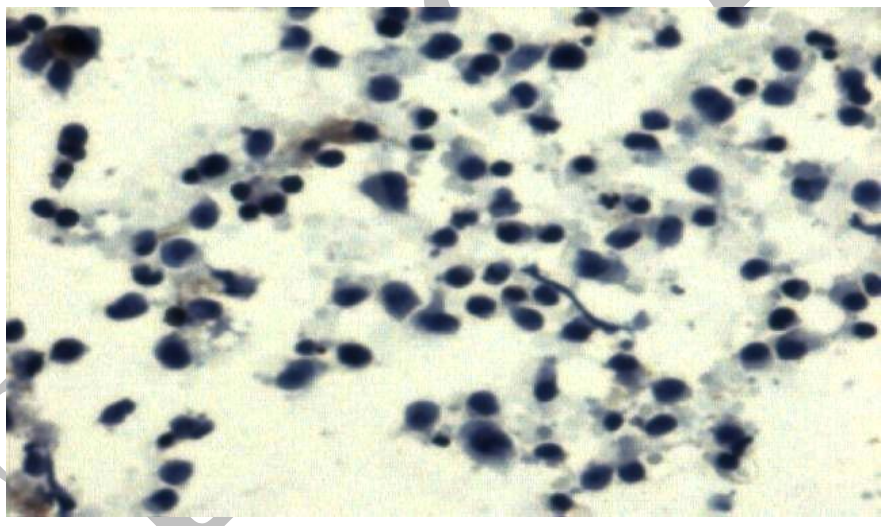


Figure 5 Immunohistochemistry on cell block preparation showed positivity for NSE (IHC, 400x).



**DISCUSSION:** Ewing's sarcoma (ES) and peripheral primitive neuroectodermal tumor (PNET) were originally described as distinct clinicopathologic entities. In 1918, Stout described a tumor of the ulnar nerve composed of small round cells focally arranged as rosettes; this entity was subsequently designated neuroepithelioma, and then PNET [4]. Later, in 1921, ES was described as an undifferentiated tumor involving the diaphysis of long bones and also reported to arise in soft tissue (extra osseous ES) [5].

PNETs are malignant tumors of the central nervous system and usually found in infants, children, and young adults.

PNETs arise from the primitive nerve cells of the nervous system but they can also occur in outside the central nervous system (peripheral PNETs) including bone in the extremities, pelvis, and the chest wall. PNETs of the chest wall were originally reported by Askin et al in 1979 in 20 children and adolescents with a mean age of 14 years. Since that time, PNETs localized in thoracopulmonary region have been defined as Askin's tumors [6].

Askin tumor is a malignant small round cell tumor of neuroectodermal origin belonging to the Ewing tumor family due to their cytogenetic appearance; both demonstrate a characteristic chromosomal translocations t(11;22)(q24;q12) [7]. Askin tumors are extremely aggressive. They occur predominantly in children and adolescents, but can develop at any age [8]. Sex distribution is still unclear. The patients usually present with a swelling in the chest wall. Cough, fever, dyspnea, and weight loss are common accompanying symptoms. Local recurrence and remote metastases are common. The prognosis is usually poor with 2- and 6-year survival rates of 38% and 14%, respectively [1]. A chest wall soft-tissue density mass, sometimes associated with rib erosion and/or pleural effusion, is the commonest radiographic manifestation. The most important role of the CT scan is to confirm the presence of a solid chest wall tumor and to demonstrate their possible intrathoracic extension and/or direct lung invasion. Magnetic resonance imaging (MRI) findings of Askin tumors have been described as heterogeneous soft-tissue masses with a moderate to high signal intensity on T1-weighted images, greater than skeletal muscle, and high signal intensity on T2-weighted images [9]. The disease is diagnosed by histologic and immunohistochemical analysis. Cytologic smears of the tumor reveal small round malignant cells that contain little cytoplasm and are arranged in rows. The typical feature is the presence of Homer–Wright rosettes with various layers of cells with fibrillary material. Immunohistochemical examinations show positivity for several neural markers, such as NSE, CD99, and vimentin [10].

Differential diagnosis of imaging with chest wall mass occurring in children and adolescents should include neuroblastoma, rhabdomyosarcoma, non-Hodgkin lymphoma and Langerhans cell histiocytosis. Most neuroblastomas occur before 5 years of age, and characteristically

present invasion through neural foramina giving a dumbbell appearance due to their origin from sympathetic nervous tissue. Open biopsy is often needed to differentiate rhabdomyosarcoma and Askin tumor because both present as soft tissue mass with bone destruction and large pleural effusion. Isolated chest wall masses are rare in non-Hodgkin lymphoma at diagnosis, which instead usually presents with pervasive nodular thickening of the pleura. It is important to exclude non-Hodgkin lymphoma as its treatment does not include surgery. Imaging features of Langerhans cell histiocytosis are a bony lytic lesion with or without soft tissue masses, and multisystemic involvement such as brain, lung and abdominal organs can be found. An isotope scan should be performed as lesions may not show osteoblastic activity [11].

In a study of Sahu et al. on the diagnostic value of fine needle aspiration biopsy in tumors of Ewing sarcoma family, round tumor cells with narrow cytoplasm were seen in all except one of the needle aspiration biopsy specimens of 14 cases and PAS was found positive in all cases [12].

Given the rarity and the recent individualization of this disease, no regimen has been validated in the literature. In contrast to extraosseousosteogenic sarcomas when management follows the principles established for soft-tissue tumors rather than primary bone tumors, ES is treated in the same multidisciplinary manner regardless of whether it arises in bone or soft tissue. Treatment includes radical surgical resection, neoadjuvant and adjuvant chemotherapy and radiation [13].

**CONCLUSION:** Askin's Tumour (synonym: primitive neuro-ectodermal tumour PNET) is a rare neoplasm of the chest wall. As a consequence of aggressive growth of tumour, therapy should be performed in oncological centres in clinical studies. The treatment includes radical surgical resection, neo adjuvant and adjuvant chemotherapy plus radiation. It is difficult to establish an accurate preoperative diagnosis of Askin's tumour. Microscopy and immune histological stain of the specific marker--neuron-specific enclose--are essential. Multimodal treatment allows a long-term survival, but often the prognosis is infuse.

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