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Research Article

ASKIN'S TUMOUR-RARE THORACOPULMONARY TUMOUR-A CASE REPORT

Balbir Malhotra and Nishanth PS

Chest and TB Department, Government Medical College, Amritsar

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ABSTRACT

Askin's tumour is a primitive neuroectodermal tumor developing from the soft tissues of the chest wall. Its diagnosis approach is complex and requires a multidisciplinary approach. We report a case of Askin's tumour in a 6 year old boy who was admitted in Department of Chest & TB, Government Medical College, Amritsar with prominence of whole of left hemithorax & on Fine needle aspiration diagnosis of neuroectodermal tumour (Askin's tumour) was made. Askin tumour is a primitive neuroectodermal tumour of thoracopulmonary region, develops from the soft tissue particularly in the paravertebral region. Neo-adjuvant chemotherapy followed by surgical excision of the tumour and post operative chemotherapy with or without radiotherapy is the treatment we intend to publish this case due to rarity of the case and often misdiagnosis of Askin's tumour.

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INTRODUCTION

Askin's tumour was described for the first time in 1979 in 20 children and adolescents with a mean age of presentation of 14.5 months, female preponderance, and median survival of 8 months¹. It presents with respiratory problems such as pain, dyspnea, and mass and weight loss. It is highly malignant with poor prognosis and short survival.² The reported overall survival is 60% at 5 years^{3,4}. They are rare malignant small round cell tumors that arise from the primitive nerve cells of the nervous system, but they can also occur outside the central nervous system in the chest wall, pelvis and extremities. Askin's tumour occurs in young Caucasian adults and is associated with poor prognosis. The frequency of Ewing's sarcoma and PNET among childhood tumours is 2%. Its occurrence in adults is rare.

Case History

Shekhar, 6 year old male child admitted with complaints of fever & dry cough for the last 3 months & breathlessness for the past 1 month. Fever was low grade in nature more during evening hours, not associated with chills or rigors. Breathlessness was gradually progressive & now he was dyspneic even on lying down. There was no previous history of any chest complaints or tuberculosis in the past. On general physical examination patient was irritable, poorly built and

poorly nourished, dyspneic at rest. Swelling on whole of the left side of chest with engorged veins over the chest wall.

On Respiratory system examination, shape of the chest was asymmetrical with bulge of left hemithorax with engorged veins over the left side of chest. Movements of chest absent over the left side of the chest, trachea shifted to right side, respiratory rate 34/min. On Palpation- Patient was febrile with tenderness over left side chest, Chest movement & expansion decreased left side of chest, Vocal fremitus -decreased on left side of chest. Percussion note was dull note on left side of chest as a whole. On Auscultation Breath sound absent left side of chest with no added sounds. Breathing on the right side is vesicular without any accompaniments.

Investigations

Hematology and blood chemistry were within normal limits. He was started on empirical antibiotics, bronchodilators and analgesics, HIV-non reactive

Chest xray shows (Fig 1) Homogenous opacity in the entire left side associated with gross mediastinal and tracheal shift towards the right.

*Corresponding author: **Balbir Malhotra**

Chest and TB Department, Government Medical College, Amritsar



Figure 1

CT scan (Fig 2) shows heterogeneously enhancing mass is seen occupying the left hemithorax completely with few cystic areas in it. Few tiny specs of calcification are seen in it. It is causing contralateral mediastinal shift, pushing the hemidiaphragm inferiorly and rib cage anteriorly.

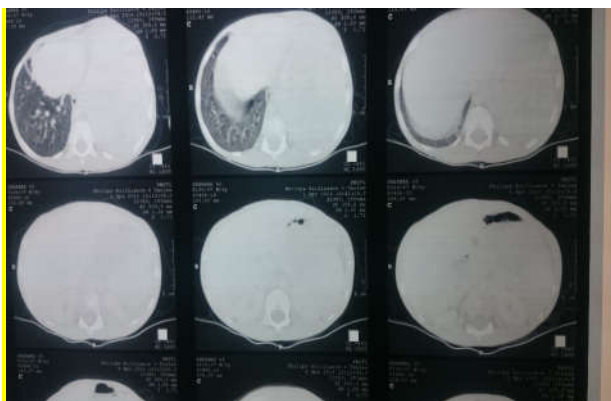


Figure 2

Few subcentimetric ipsilateral axillary lymph node are seen. Right lung is reduced in volume. FNAC - (Fig 3) Features suggestive of Extra osseous Ewing's sarcoma -Askins tumour.

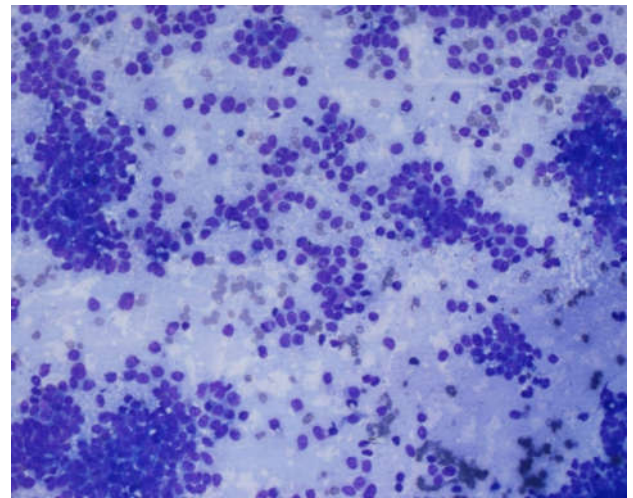


Figure 3

Other investigations were normal including USG Abdomen

DISCUSSION

Ewing's sarcoma (ES) was initially believed to be of perivascular endothelial origin. The Ewing's sarcoma family of tumors (EFT) includes Ewing's sarcoma of bone (ESB), extraosseous. Ewing's sarcoma (EES), peripheral primitive neuroectodermal tumor of bone (pPNET), and malignant small-cell tumor of thoracopulmonary region (Askin's tumor). All of these tumors are now known to be neoplasms of neuroectodermal origin⁵.

The prevalence of Askin's tumour was 0.2 cases per million in a 45 year database review of Dickinson *et al*⁶. Small round cell tumours encountered in children and young adults include Ewing's sarcoma, PNET, rhabdomyosarcoma, neuroblastoma and lymphoma.^{7,8} The malignant round cell tumour, that originates from the soft tissue of the chest wall, is also called extra-skeletal Ewing's sarcoma or peripheral PNET.^{9,10} Askin tumour, also named as 'extra-skeletal' Ewing's sarcoma' or 'soft tissue Ewing's sarcoma' arising from the soft tissue of the chest wall. Askin tumor is locally aggressive tumor and localized to thoracopulmonary region. It is a rare disorder mimicking common pediatric conditions such as tuberculosis, lymphoma, neuroblastoma, and rhabdomyosarcoma. So, early diagnosis and timely intervention is critical for favorable outcome of the patients. Typically, Askin's tumor develops as a solitary mass or multiple masses in the thoracic area (thoracic wall, lung, mediastinum, or pericardium). Pain is the only or main symptom in 60% of cases. Seventy-five percent of Askin's tumors have been reported in white female patients younger than 20 years.¹¹ Rarely, Askin's tumors are found in the central nervous system. In the thoracic area, these tumors are invasive and prone to destroying bone (ribs and scapula), invading the retroperitoneal space, and spreading to lymph nodes, adrenals, and liver. Once resected, they recur with extremely high frequency. The most common recurrence sites are the skeleton, the sympathetic chain, and the original site. It shows a neural differentiation that can be demonstrated by immunohistochemical and ultrastructural methods.

Positivity for neural markers, such as neuron specific enolase and also neuroendocrine markers, such as chromagranin and synaptophysin are typical of Askins tumour which are also seen in Ewing's sarcoma and PNET. These are also positive for MIC-2 gene which produces CD 99 and a cell membrane-like protein p 30/32 which are highly sensitive but not specific products. PNET of the chest wall should be considered in a child with a chest wall mass. CT is valuable for evaluating tumor extension at diagnosis, the effects of chemotherapy, and assessing tumor recurrence after surgery. However, CT can overestimate infiltration into the pleura, lung or diaphragm, and it would be better evaluated by ultrasonography. MRI is superior to CT for evaluation of tumor extension, and may be considered complementary to CT, particularly for very large tumors of the chest wall¹². Kabiri and colleagues emphasized on the difficult histological diagnosis, and demonstrated the importance of complete removal of the tumor for survival¹³. Pain is the only or the main symptom in 60% of the cases. Radiological characteristics range from a unilateral chest wall mass to pleural fluid, invasion to the adjacent lung parenchyma, pulmonary nodules and sometimes lymphadenopathy. The diagnosis of Askin tumour rests on cyto-pathological investigations and immunohistochemical tests. The established treatment of this tumor is neo-adjuvant chemotherapy followed by surgical excision of the tumor and post operative chemotherapy with or without radiotherapy^{14,15}. The neo-adjuvant chemotherapy results in better regional management of the tumor, less extensive surgery and can treat the distant metastasis. Currently, Doxorubicin is added to the regimen in the majority of protocols. Additional drugs administered in most patients are Ifosfamide and Etoposide. Operation with wide margins is ideal, but occasionally is feasible in patients with chest wall tumors. Patients with positive margin of tumor after surgery need post operative radiation.

The overall survival is approximately 60% at 5 years. The late effects of local therapy include scoliosis, restrictive lung disease, hypoplasia of soft tissue, and secondary tumors. The best prognosis can be provided by surgical treatment with wide resection. Recurrences in the primary tumour site are important in differentiating these tumours from other tumours in children and adolescents. As local recurrences after resection and metastases are frequently seen in Askin tumour, it has a poor prognosis and a short survival. The most common recurrence sites are the skeleton, sympathetic chain and the original site. Indicators of poor prognosis include advanced age, metastatic disease, extraosseous primary tumour and recurrence. Aggressive chemotherapy in recent studies have shown that remission rate has improved from 26% to 65%. Various chemotherapy regimens have been used that include VAC (vincristine, actinomycin D, cyclophosphamide), VACA (vincristine, actinomycin D, cyclophosphamide, adriamycin) and VAC alternating IE (ifosamide and etoposide)

CONCLUSION

Askin tumour is to be considered as an etiologic possibility in a small-cell tumour in thoracopulmonary region, especially in the young age group. Patients should be treated surgically, with wide local excision wherever possible. Combination of chemotherapy should be considered in patients who are non-operative

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