

Askin Tumor: Malignant Round Cell Tumor in Thoraco-pulmonary Region in a Young Male Child

Swati Goyal¹, Gaurav Chahal², Alok Kumar Yadav², Rahul Raja²

¹Resident, Department of Radio-diagnosis, Eras Lucknow Medical College, Lucknow, U.P., India.

²Resident, Department of Orthopaedics, Eras Lucknow Medical College, Lucknow, U.P., India.

ABSTRACT

Askin's tumor is a rare tumor arising from the chest wall. It is known to be a subset of Ewing's sarcoma histologically presenting as a small round blue cells. It is a highly malignant tumor with poor prognosis. 17-year-old teenager visited in our department with complaints of chest pain and swelling. Chest skiagram revealed a homogenous radio opacity in the right mid/ lower hemithorax. NECT and CECT thorax was done and was suggestive of two lesions in chest. One large lytic lesion involving the right lower ribs (6th, 7th) associated with large heterogeneous soft tissue mass on medial side. The other causing destruction of body of D8 vertebra associated with a paravertebral soft tissue mass. The diagnosis was further confirmed by biopsy of 6th rib which was suggestive of small round cell tumor.

Keywords: Askin tumour, Thoraco-pulmonary

INTRODUCTION

Askin tumour was defined by Askin and Rosai^[1] in 1979. However the cytogenetics and the molecular biology, allowed including it with the sarcoma of Ewing and the neuro-epithelioma within a single entity under the term of primitive neuro-ectodermal tumors (PNET). The frequency of Ewing's sarcoma and PNET among childhood tumours is 2%, with the occurrence of Askin tumour is unestablished due to the rarity of the disease. Askin tumour is a rare tumour of childhood and usually presents with respiratory symptoms. Its occurrence in adults is much rarer. To date, no definitive treatment and prognosis is made. We report a case of Askin tumor in a 17-year-old male child and review the different data from the literature.

CASE REPORT

A 17-year-old male child, resident of Lucknow, non-smoker, non-alcoholic, presented with chest pain, back pain and swelling for the past seven months. He was earlier being treated symptomatically with oral antibiotics and was partially relieved. However, later he developed progressive breathlessness and vague chest pain over the right hemithorax. Chest radiograph posterior-anterior view revealed a homogenous radio opacity in the right mid/ lower hemithorax [Figure 1].

Name & Address of Corresponding Author

Dr. Alok Kumar Yadav
Junior Resident
Dept of Orthopaedics
Eras Medical college
Lucknow, UP, India
E mail: alokiyadav3001@gmail.com

NECT and CECT thorax was done and was suggestive of two lesions in chest [Figure 2]. One large lytic lesion involving the right lower ribs (6th, 7th) associated with large heterogeneous soft tissue mass on medial side [Figure 3]. The other lesion was seen causing destruction of body of D8 vertebra associated with a paravertebral soft tissue mass [Figure 4]. The diagnosis was further confirmed by biopsy of 6th rib which was suggestive of small round cell tumor [Figure 5]. Excision biopsy revealed almost complete replacement by a tumour composed of sheets of cells with scanty cytoplasm. The nuclei were hyperchromatic with "salt and pepper" chromatin and inconspicuous nucleoli. Nuclear moulding and brisk mitosis was noted. Perivascular pseudo-rosettes was seen at places. The histology was consistent with a malignant round cell tumour. CECT of the whole abdomen, brain and bone scan did not show any evidence of metastatic involvement.

DISCUSSION

Dickinson et al^[2] in a 45-year database review found the prevalence of Askin's tumour to be of 0.2 cases per million. Ewing's sarcoma, PNET, rhabdomyosarcoma, neuroblastoma and lymphoma all are small round cell tumours encountered in children and young adults.^[3,4] Although Ewing's sarcoma and PNET are easily differentiated microscopically from other entities in this group, the differentiation amongst Ewing's sarcoma and PNET is very difficult. This malignant round cell tumour that originates from the soft tissue of the chest wall is also known to be extra-skeletal Ewing's sarcoma or peripheral PNET.^[5,6] Askin tumour develops as a solitary mass, few occasions involving the hemi-thorax^[7] or as multiple

masses in the thoraco-pulmonary region. Its histogenesis is uncertain and would result from the migration of cells from the neural crest during the embryonic life [8]. These tumors have positivity for neural markers, such as neuron specific enolase and also neuroendocrine markers, such as chromagranin and synaptophysin. Askin tumour are also positive for MIC-2 gene which produces CD 99 and a cell membrane-like protein p 30/ 32, highly sensitive but not specific products. According to the current concept, Askin tumour is a variant of Ewing's sarcoma and PNET involving the thoracopulmonary region.

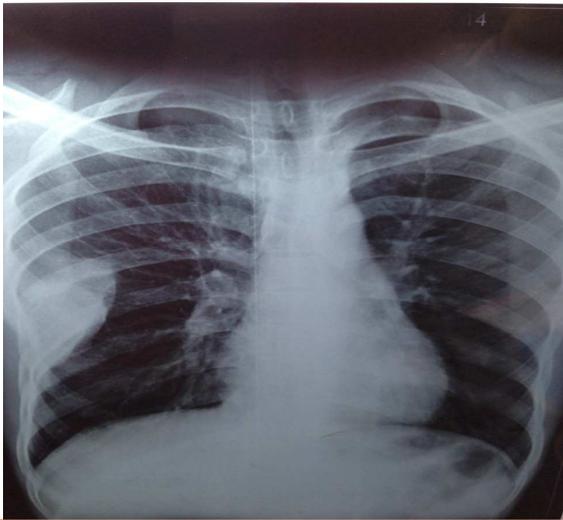


Figure 1: Chest radiograph (PA view)



Figure 2: CECT thorax

Very rare sometimes Askin tumours can also be found in the central nervous system. In thorax, these tumours are invasive and prone to involve bones of ribs and scapula, and into the retroperitoneal space, involving the lymph nodes, adrenals, and liver. Askin *ET al*¹ reported that small round cell tumours of childhood and adolescence located in the thoracopulmonary region are common in females than males, the median age for these being 14 and half years.



Figure 3: Heterogenous mass



Figure 4: Soft tissue mass

Radiological pictures range from a unilateral chest wall mass with invasion to the adjacent lung parenchyma with pulmonary nodules and with or without lymphadenopathy. The final diagnosis of Askin tumour rests microbiological examinations (cytopathological investigations and immunohistochemical tests). Treatment of Askin tumour involve radical surgery, neo-adjuvant or adjuvant chemotherapy and radiotherapy. Long survival of patient can be achieved by multimodel therapy but still prognosis is very poor. Out of which best prognosis can be provided by surgical treatment and wide resection. Recurrences at primary tumour

site are important in differentiating these tumours from other tumours in children and adolescents¹. Local recurrences of tumor after resection and metastases are less commonly seen, it has a poor prognosis and a short survival⁵. Commonest recurrence sites are the skeleton, sympathetic chain and the original site. Advanced age, metastatic disease, extra osseous primary tumour and recurrence are considered as poor indicators of disease.^[9]

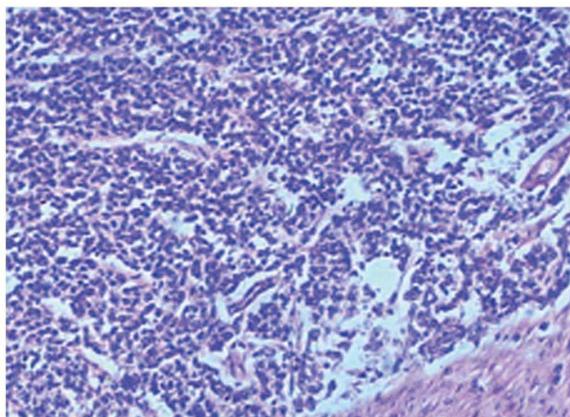


Figure 5: Small round cell tumour

Few recent studies have shown that remission rate has climbed up from 26% to 65% with aggressive chemotherapy. Multiple chemotherapy regimens^[10] have been used like VAC (vincristine, actinomycin D, and cyclophosphamide), VACA (vincristine, actinomycin D, cyclophosphamide, Adriamycin) and VAC alternating IE (ifosamide and etoposide). Average survival has been reported to be eight months after the diagnosis.^[11]

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 visiting to OPD with such tumours 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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