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CASE REPORT

ASKINS TUMOR: A RARE CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Ewing's sarcoma of the chest wall forms a part of the Ewing's family of tumors which also includes Ewing's sarcoma and primitiveneuroectodermal tumour (PNET).Also known as the Askin tumour it was first described as a separate entity by Askin and Rosai in 1979[1].As the occurrence of Ewing's sarcoma and PNET among childhood tumours is 2%, data standard validated treatment regimens are yet to be formulated. We report a rare case of childhood Askin's tumor and its management at our center.

INTRODUCTION

A 2-year-old male child presented with a 12 × 8 cm, lump arising from left side of chest wall involving the entire axillary region associated with low-grade fever since two months (Fig 1). Parents of the patient thought swelling to arise from the shoulder joint hence presented to us in the orthopaedic OPD. Ultrasonography showed a large heterogenous hyperechoic necrotic mass arising from the left chest wall involving axillary region without any mediastinal infiltration .Plain radiograph depicted ribs, clavicle and humerus not involved by tumor mass. Computed tomography (CT) scan revealed a large exophytic, heterogeneously enhancing mass lesion in the left axilla. Possibility of Ewing's sarcoma, lymphoma or rhabdomyosarcoma was suggested. Patient was planned for an open incisional biopsyand the biopsy report suggested mesenchymal cells with a hemorrhagic background suggestive of rhabdomyosarcoma and immunohistochemistry was awaited. Metastatic workup was done and no metastasis were found in liver, lung brain and bone. Meanwhile based on the clinical, laboratory and radiologic findings, an initial diagnosis of a large chest wall rhabdomyosarcoma was suspected and neoadjuvant chemotherapy initiated including Vincristine, Adriamycin and Cyclophosphamide for 3 weeks duration. There was no clinical response to chemotherapy and the tumor rapidly progressed in size further. Owing to its fleshy fragile surface tumor even on utmost care and dressing with paraffin gauge used to bleed profusely. Child developed high spikes of fever, cachexia and severe anemia.

Figure 1 Tumor mass involving left axillary region extending towards anterior chest wall as well as posteriorly.]

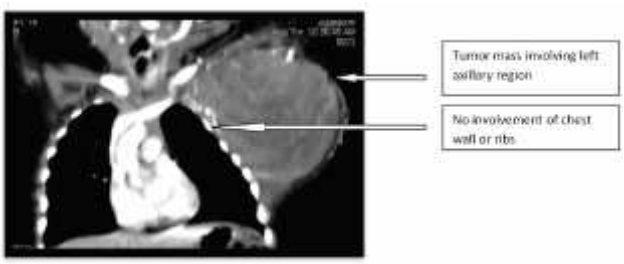

Figure 2 CT scan showing a well-defined extra osseus tumor involving the left axillary region.

Multiple blood transfusions were administered. Due to non responsiveness of the tumor to chemotherapy doubts were raised and so we decided repeat biopsy which revealed multiple tissue bits comprising of tumor cells arranged in sheets with many interspersed blood vessels suggestive of round cell tumors. Tumor cells were periodic acid Schiff (PAS) positive and PAS was diastase sensitive suggesting glycogen in the cells. On Immunohistochemistry, tumor cells showed focal membranous positivity for CD 99 and was negative for desmin, myogenin, LCA and HMB45.

DISCUSSION

Extra osseous malignancies of chest wall are a rare entity in the pediatric population. The reported incidence is 0.2 cases per million as described in a database review [2].The small round cell tumor described as chest wall tumors include Ewing's sarcoma, PNET, rhabdomyosarcoma, neuroblastoma and lymphoma in the pediatric population.[3,4]

The differentiation between Ewing's sarcoma and PNET is quite difficult clinically and upon radiographic investigations. Askins tumor as originally described by Askin et al is a malignant round cell tumour, that originates from the soft tissue of the chest wall, also called extra-skeletal Ewing's sarcoma or peripheral PNET. [5,6]

The usual presentation being a solitary mass, rarely involving the hemithorax[7] or asmultiple masses in the thoraco-pulmonary region (thoracic wall, lung, mediastinum, or pericardium).In our case the tumor originated as a small lump in the left axillary region and it rapidly progressed to a large size. Although no involvement of ribs or lungs was noted in our case. However due to late presentation the child was already emaciated and cachexic due to continous bleeding from the necrotic tumor mass and secondary infection in the tumor mass.

Askin tumour, also shows a neural differentiation that can be demonstrated byimmunohistochemical methods.

Similar to Ewing's sarcoma and PNET, these tumours have positivity for neural markers, such as neuronspecific enolase and also neuroendocrine markers, such as chromagranin and synaptophysin. These are also positive for MIC-2 gene which produces CD 99 and a cell membrane-like protein p 30/32 which arehighly sensitive but not specific products.

Our case showed positivity to CD99 on Immunohistochemistry According to the current consensus, the so-called Askin tumour is a variant of Ewing's sarcoma and PNET that involves the thoracopulmonary region.

Rarely, Askin tumours are found in the central nervous system. In the thoracic area, these tumours are invasive and prone to involve bone (ribs and scapula), invading the retroperitoneal space, and spreading to lymph nodes, adrenals, and liver. Askin et a1[1] reported that small round cell tumours of childhood and adolescence located in the thoracopulmonary region have a predilection for females, the median age for these being 14.5 years. 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 cytopathological investigation and immunohistochemical tests.

Treatment in Askin tumour consists of radical surgery, neo-adjuvant or adjuvant chemotherapy and radiotherapy. However the poor general condition of our patient didn't permit surgical resection.

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.[1]

As local recurrences after resection and metastases are frequently seen in Askin tumour, it has a poor prognosis and a short survival.[5]

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 reccurrence.[8]

Recent studies have shown that remission rate has improved from 30% to 75% with aggressive chemotherapy. Average survival has been reported to be eight months after the diagnosis.[1]

In our case we used VAC (vincristine, actinomycin D, cyclophosphamide) regimen but there was no regression of the tumor mass and the patient ultimately succumbed to death. Various chemotherapy regimens have been used that include VAC , VACA (vincristine, actinomycin D, cyclophosphamide, adriamycin) and VAC alternating IE (ifosamide and etoposide). [10] .The authors report this case so that actual incidence of this rare askins tumor can be reported and furthermore a treatment protocol can be formulated so that medical knowledge can be enhanced for medical fraternity.

CONCLUSION

Rarity of the disease and rapid progression of the disease towards fatal end are big hindrance for formulation of any standard treatment regimen. Askin tumour should be considered as an aetiologic possibility in a small-cell tumour of the thoracopulmonary region, especially in the young age group. Patients with such tumours should be treated surgically, with wide local excision wherever possible. Combination chemotherapy should be considered in patients with inoperable disease. More research and study oriented approach towards regression of tumor needs to formulate a standard treatment regimen.

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