AN UNUSUAL MASS OF THE CHEST WALL IN A 7 YEARS OLD CHILD: AN ASKIN’S TUMOUR

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Abstract
Askin’s tumour is a primitive neuroectodermal tumour developing from the soft tissues of the chest wall. It is commonly misdiagnosed due to features similar to other round blue cell tumours. The approach for its diagnosis and treatment is complex and requires a multidisciplinary team. The recommended treatment is chemotherapy and surgical excision mainly. The prognosis is dependent on many factors. We managed a child of Askin’s tumour using modalities like neo-adjuvant chemotherapy and surgical excision and found a good response.

Key Words: Chest wall mass, Askin’s tumour, Ewing sarcoma

Introduction:
The literature showed that frequency of Ewing’s sarcoma and PNET among childhood tumours is just 2%, that is the reason during the age of infancy and childhood, Ewing's sarcomas (ESs) and primitive neuroectodermal tumours (PNETs) considered to be very rare [1]. Askin’s tumour in childhood usually presents with common symptoms of upper respiratory tract infections. These are closely related neoplasms derived from the neural crest cells tumours [2]. In 1979 Askin et al separated PNETs originating from thoraco-pulmonary region and named Askin’s tumour. They belong to the family of the small round blue cell tumours and have no specific clinical and pathologic features which makes difficult to differentiate it from other tumours of their family [3]. Generally prognosis of these tumours is poor. The index case showed good response to multimodality treatment and till now no signs and symptoms of recurrence. Furthermore on searching literature we found that to date there are very few case reports from Pakistan.

Case Report:
A 7-year old boy presented to Oncology department with complaint of intermittent, dull and moderate to severe pain along with progressively increasing swelling on left lower chest wall for one year.

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Initially managed by as a case of pulmonary Tuberculosis for about three months with no response. On examination there was about 20X14 cm firm, non-tender, non-fluctuant swelling on left chest wall extending from 5th to 9th rib (Figure 1). All other physical parameters and labs including Alpha fetoprotein and Beta HCG were within normal range. Abdominal ultrasound showed complex multi loculated mild to moderate effusion involving lateral chest wall with internal septations and underlying consolidation. On CT scan, 19.3 x 12.7cm sized cystic mass in the left hemi thorax with displacement of mediastinum. The evidence of rib destruction was there but there was no mediastinal lymphadenopathy or bone involvement. (Figure 2). An Incisional biopsy of mass was done and proved out to be CD99 positive round blue cell tumour probably Ewing’s sarcoma (Figure 4). Patient underwent 6 cycles of chemotherapy (vincristine, ifosfamide, doxorubicin, etoposide). Post chemotherapy scan depicted reduction in mass size to just 6.0 x 2.6 cm located underneath the lower left ribs, inseparable from left lateral hemi diaphragm involving inner lower chest and upper abdominal wall but no lung infiltration (Figure 4). The case was discussed in tumour board; excision of tumour was planned. Left subcostal incision was made and tumour approached through trans-diaphragmatic route as it was involving the underlying ribs and diaphragm. After opening the diaphragm it was visible on under surface of lower ribs and excision was done (Figures 5-7).
Per operatively tumour was 5 x 6 cm arising from the periosteum of lower ribs and attached to pleura of left lower lobe and left hemi diaphragm. Periosteum was shaved and underlying Rib surface was smooth. Biopsies were taken from pleural site, diaphragm and periosteum of rib. Closure of diaphragm and abdomen done (Figure 9). Post-operative recovery of the patient was smooth and uneventful. He was initially shifted to ICU and then general ward after 3 days. Chest tube was removed after 5 days. Patient was referred back to oncology ward. On histopathological examination of resected mass there was no tumorous tissue and there was only fibrous tissue with necrosis. Also there was no residual tumour in pleura, diaphragm or rib periosteum. At three, six and nine months follow up patient is doing fine and completed his post-operative chemotherapy.

Discussion:

PNETs, a group of highly malignant tumours are of neuroectodermal origin that affects mainly soft tissue and bone. Due to indistinguishable clinical and pathologic features, Batsakis et al (1996) divided the PNET family of tumours into the following 3 groups; central nervous system PNETs, autonomic nervous system PNETs (Neuroblastoma) and peripheral PNETs (pPNETs) [4]. The annual incidence of PNETs is 2.9 per million population with a slight male preponderance. Askin's tumour, also named as ‘extra-skeletal’ Ewing’s sarcoma’ or ‘soft tissue Ewing’s sarcoma’ arising from the soft tissue of the chest wall and usually present in the second decade of life [5]. For extraosseous primary tumours, the most common primary sites of disease include the following: Trunk (32%), Extremities (26%), Head and neck (18%) and Retroperitoneum (16%) [6].
Askin’s tumour may present as a solitary mass, rarely involving most of the hemithorax or as multiple masses in the thoraco-pulmonary region (thoracic wall, lung, mediastinum, or pericardium). Mostly these tumours show a neural differentiation that can be demonstrated by immune-histochemical and ultrastructural methods. Similar to ES and PNET, regarding immunohistochemistry, these tumours show positivity for neural markers such as neuron specific enolase, CD99, vimentin including neuroendocrine markers, such as chromagranin and synaptophysin [7]. CD 99 was also positive in our index case. 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 [3] and usually presents with common respiratory symptoms. In Our Case lower ribs were also involved but responded to chemotherapy very well and no involvement was found per-operatively. Treatment is actually multimodal including chemotherapy, surgery and radiotherapy. Venkitaraman et al proposed remarkable remission rate from 26% to 65% after chemotherapy. Most commonly used chemotherapy regimens include VAC (vincristine, actinomycin D, cyclophosphamide), VACA (vincristine, actinomycin D, cyclophosphamide, Adriamycin) and VAC alternating IE (ifosamide and etoposide) [8]. The regimen used by our oncology team was vincristine, ifosfamide, doxorubicin, etoposide and patient responded well. The next important role to improve survival is complete surgical excision of tumour, though approach needs modification regarding exact location of tumour. In this case the surgeon approached the residual tumour through trans-diaphragmatic route. The extension of the tumour at diagnosis is the factor affecting prognosis although 70% 5-year survival is documented in literature [9]. Ewing sarcoma is usually sensitive to radiotherapy but needs to categorise the patient as far as selection is concerned especially in case of younger age patient [10].

No metastasis or relapse was seen in index case after successful excision and chemotherapy so radiotherapy was not given. Most common complications which can occur are in the form of relapses and musculoskeletal abnormalities, fortunately both of which were not present in our case, as tumour proved out to be chemosensitive and great reduction in size was there which was easy to excise without any thoracic wall deformity.
Conclusion:
Askin’s tumour is very rare in childhood. The most common presentation is with respiratory symptoms. High index of suspicion is required to diagnose it while being examined by primary physician whenever there is involvement of chest wall. Patients require treatment with chemotherapy; surgery and long term follow due to risk of recurrence.

References: