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Askin Tumor: A Case Report of a Rare Tumor

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ABSTRACT

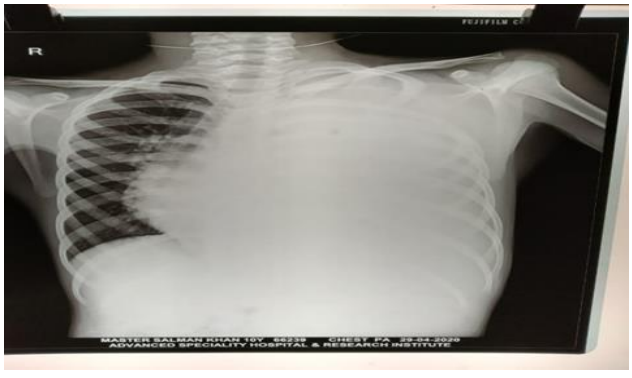
Askin tumor is a rare malignant neoplasm of neuro-ectodermal origin that emerges from the soft tissue of the thoraco-pulmonary wall and has aggressive activity. This paper presents a study on a 10-year-old boy having breathing difficulty and mass on the anterior portion of the left thoracic wall. On examination, there was a mass on the left anterior chest wall measuring 13x13 cm2, which was hard, tender, and fixed to the chest wall. There was associated pleural effusion and infiltration into the left lung. Histopathology confirmed primitive neuro-ectodermal tumor (Askin tumor) with IHC marker CD99 positive. Chemotherapy was used to treat the patient. Early detection of this tumor would necessitate a high index of suspicion, allowing for a better prognosis and result. The overall prognosis is poor due to the higher rate of local recurrence.

1. Introduction

Askin tumor belongs to the Ewing group of sarcoma with small round blue cell tumors.[1] It is a rare malignant neoplasm of neuro-ectodermal origin with aggressive behavior originating from the soft tissue of the thoraco-pulmonary wall.[2] It predominantly occurs in males and patients under 20 years of age; defined by reciprocal translocation t (11:22) (q24:q12) with EWS-FLI-1 fusion gene[3, 4] and believed to be of neural crest origin. The patients usually present with a painful chest wall mass, cough, fever, dyspnoea, and weight loss. Cremone et al.[5] diagnosed the above indications with histologic and immunohistochemical analysis. The standard treatment for an Askin tumor consists of anterior chemotherapy, complete surgical resection, adjuvant chemotherapy, and radiotherapy.[6, 7]

2. Case presentation

A 10-year-old boy attended the Department of Radiation Oncology, RIMS Imphal, with two months history of breathing difficulty and swelling (on the left chest wall). It was associated with fever and dry cough for the last four days. The patient's past and familial histories were unremarkable. On examination, a mass on the anterior portion of the thoracic wall measured 13x13cm2, which was hard, tender, and fixed to the chest wall, as illustrated in Fig. 1. (a). Fig. 1. (b) represents the chest radiograph of the patient with massive pleural effusion.



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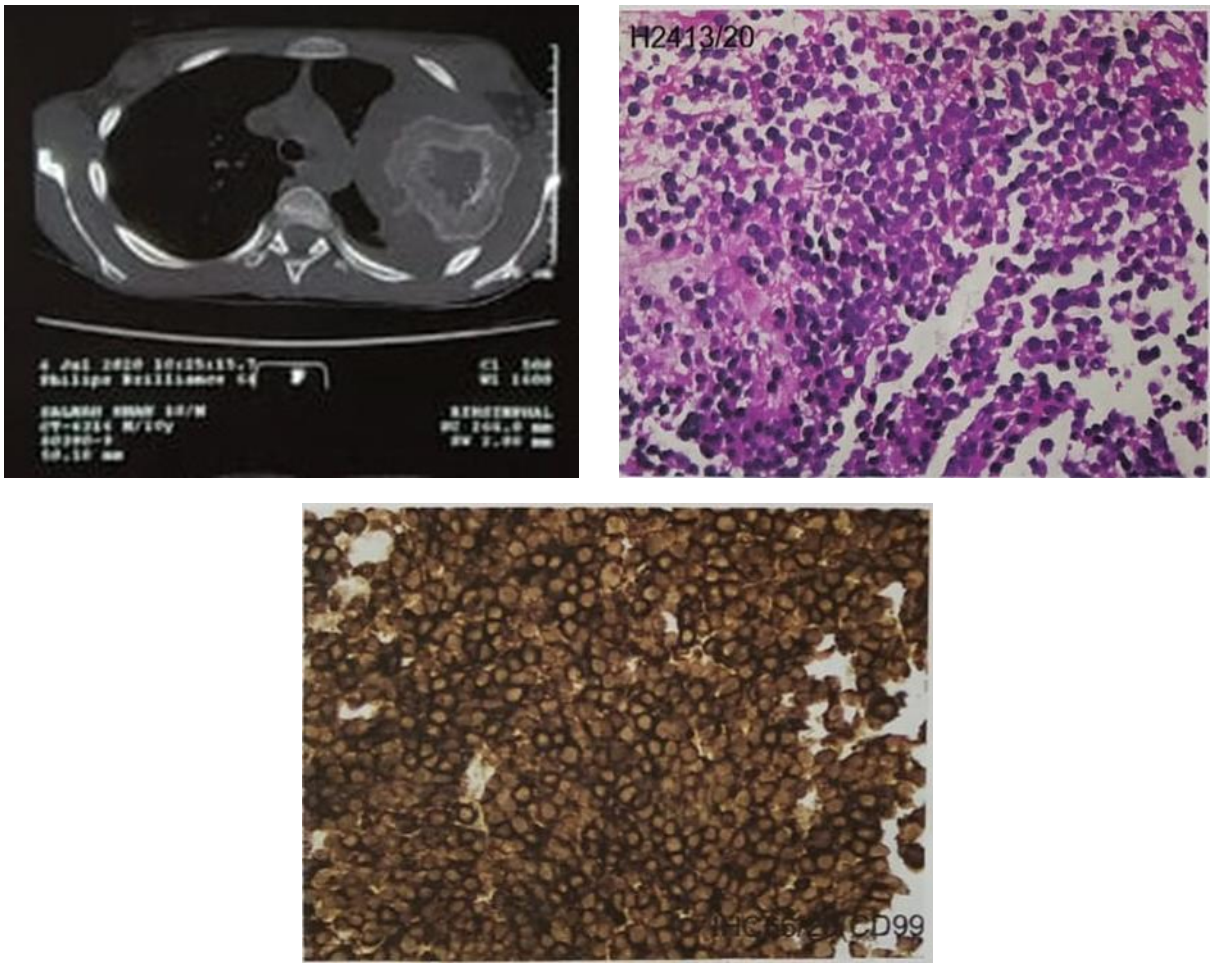

Fig. 1. Askin Tumor. (a) Swelling over left chest wall. (b) Massive pleural effusion. (c) HRCT thorax showing expansile lytic lesion with lobulated soft tissue component. (d) A small round cells malignant tumor with scanty cytoplasm and hyperchromatic nuclei arranged in sheets, clusters, and peritheliomatous patterns. (e) CD99/MIC-2 positive.

The child underwent HRCT thorax that revealed an expansile lytic lesion with a lobulated soft tissue component and calcifications in the lateral aspect of the left 3rd rib infiltrating and abutting into the left lung, as well as an underlying gross left pleural effusion and total lung collapse (suggestive of the neoplastic lesion) as displayed in Fig. 1. (c). FNAC from the lesion showed small round cells with scanty cytoplasm and relatively large nuclei, as shown in Fig. 1. (d). Trucut biopsy from left chest wall for histopathological examination showed primitive neuro-ectodermal tumor (Askin tumor) with IHC marker CD99 positive, as illustrated in Fig. 1. (e). His routine investigations were uneventful.

The final diagnosis was a primitive neuro-ectodermal tumor (Askin tumor). The patient underwent chemotherapy with cyclophosphamide, vincristine, and dactinomycin 9 weekly cycle. The tumor regressed significantly following 1st cycle of chemotherapy (cyclophosphamide-D1, 2, 22, 23, 43, 44, VCR-D1, 8, 15, 22, Dactinomycin-D22, 23, 24). The patient was advised for surgery after the first cycle, but the patient family refused and lost to follow up. Moreover, the patient reappears after two months with the complaint of pain over the swelling and vomiting. On examination, swelling (measures~9x9cm^2), hard, tender, and fixed to the chest wall. The patient was administered chemotherapy with the same chemo regime, but unfortunately, the response was not good, and we referred to a higher center for radiotherapy.

3. Discussion

Askin's tumor is a rare tumor with aggressive behavior. It was first reported in 1979 by Askin and Rosai. The disease predominantly occurs in males, and 80% of diagnosed patients are under 20 years of age; however, there are reported cases in elderly patients.[8] Askin's tumor exhibits common respiratory manifestations. Chiefly, it involves cough, chest pain, fever, difficulty in breathing. It may begin with localised abnormal sensation and hardly as a pathological fracture or metastasis-related symptoms.[9] Chest radiograph in Askin's tumor may show classical onion-peel appearance or just homogenous opacity. However, in our case, it showed consistent attenuation. Non-Hodgkin's lymphoma, small cell osteosarcoma, and metastatic neuroblastoma during the histologic examination should be considered differentials.[9, 10] Immunohistochemistry distinguishes and confirms peripheral neuro-ectodermal tumors from round cell tumors.[4, 9, 11, 12] The tumor tested positive for NSE, CD99, and vimentin in immunohistochemistry.

Askin tumor treatment must include radical resection, radiotherapy, and aggressive chemotherapy (cyclophosphamide, vincristine, doxorubicin, Ifosfamide, Etoposide, and Dactinomycin). The most important role is played by surgical therapy. However, because of its position, it must be customized. Although it is a well-known fact that Ewing sarcoma is highly responsive to radiation, radiation therapy should be used with caution, especially in younger children, as there is a high chance of the chest wall defect of developing bones

and neurodevelopmental complications. Although radiotherapy was avoided initially due to irregular follow-up, non-responsive to chemotherapy, and the tumor's aggressive nature, we have decided on radiation therapy.[13] Even though a 96-month durable survival period has been observed, the average life span is eight months. However, unfortunately, the patient died six months after being diagnosed.

4. Conlcusion

Askin's tumor is a rare and aggressive tumor of childhood. Its diagnosis is challenging because of its rarity and its clinical polymorphism. It needs high concern of this tumor to identify at the earliest for better result and a great chance of recovery. This study has explained a case of a 10-year-old boy. Despite chemotherapy, the patient died four months later. Because of its destructive nature, troublesome course, and recurrence tendency, long-term follow-up is warranted.

Conflict of Interest

The authors declared that there is no conflict of interest.

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