Case Report

Askin tumor: A rare neoplasm of thoracopulmonary region

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ABSTRACT

Askin tumor is a rare neoplasm of thoracopulmonary region. But it mimics other common pediatric disorders, such as empyema, lymphoma, and tuberculosis, posing a great diagnostic and therapeutic challenge to the treating clinicians. So it is of utmost importance to make an early diagnosis and proper referral/treatment in such cases. We highlighted diagnostic challenge, treatment, and favorable outcome of a case that presented to us.

KEY WORDS: Adolescent, askin tumor, neural crest

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INTRODUCTION

Ewing sarcoma, peripheral primitive neuroectodermal tumors and Askin tumors belong to Ewing family of tumors with a variable degree of neuroectoderm differentiation. Histologically, they all show small round blue cells. Askin tumor is a primitive neuroectodermal tumor of thoracopulmonary region 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 months.[1] Askin tumor 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] Because of rarity of condition, there is no defined treatment guideline for this condition. Most centers follow multimodality treatment of chemotherapy, surgery, and radiotherapy.

CASE REPORT

A 14-year-old girl presented to pediatric emergency with chest pain on the right side of the body, excessive irritability for last 15 days, and difficulty in breathing for 3 days. There was no history of fever, cough, blood mixed vomitus, swelling anywhere, trauma, convulsions, yellowish discoloration of eyes and urine, or blood transfusion. A history of tubercular contact was present in her mother 4 years back.

At presentation in emergency: Examination revealed sick dyspneic girl with vitals; respiratory rate (32/min), pulse rate (88/min), blood pressure (102/60 mmHg), mild pallor, pedal edema, and normal jugular venous pressure. Inspection of respiratory system revealed positive trail sign on left side, fullness of intercostals space on right side, movement decreased on right side, and apical impulse was not visualized. Palpation confirmed the shifting of trachea to left side. There was decreased movement of right chest wall with tenderness present in right infraaxillary and inframammary area. Tactile vocal fremitus was decreased on right side of chest. There was stony dullness sound on percussion of right side of chest in all areas. Air entry was decreased/absent in the right side of chest in all areas. Examination of left side of chest was normal. Rest of the systemic examination was normal.

The child was stabilized with respiratory support and intravenous fluids. Diagnostic tap revealed straw colored

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fluid with cobweb coagulum formation. Fluid biochemistry revealed protein 2.7 g/dL and sugar 78 g/dL. Total cell count was 80/mm^3 with 70% neutrophils and 30% lymphocytes. Adenosine deaminase level was 20 U/L. Gram stain and Acid fast satin were negative. Tuberculin test was negative. Pleural fluid and blood culture was sterile. Blood biochemistry: Hemoglobin, total leukocyte count, liver function test, renal function test, viral markers for hepatitis B surface antigen and human immunodeficiency virus were normal. Urinary catecholamine metabolite was normal. Chest radiography revealed right side radioopaque hemithorax [Figure 1]. Sonography of chest revealed massive fluid in right pleural cavity causing mediastinal shift and displacing abdominal organs downward. Right parietal pleura show multiple heteroechoic deposits (largest measuring 4.7 cm × 1.9 cm). Computed tomography showed well-defined oval heterogeneously enhancing soft tissue dense lesion arising from right lateral chest wall with involvement of pleura and subpleural fat plane and associated permeative erosion of right fifth rib. There was right-sided hydropneumothorax with multiple enhancing nodular pleural deposits with few heterogeneously enhancing nodules in right collapse lung [Figure 2]. Sonography-guided biopsy showed round cell having scanty basophilic cytoplasm. Staining for glycogen granules was negative. Immunohistochemistry showed CD 99 positivity [Figure 3]. The child was managed with respiratory support, intravenous antibiotics, intercostal drainage tube, chemotherapy, and radiotherapy. Her condition improved with supportive treatment and was not on any respiratory support before starting chemotherapy. She was started on combination of six drugs chemotherapy protocol (VACAc/IE): Vincristine (V; 1.5 mg/m^2), doxorubicin (A; 80 mg/m^2), cyclophosphamide (C; 1200 mg/m^2), actinomycin D (Ac; 1.5 mg/m^2), ifosfamide (I; 9 g/m^2); etoposide (E; 600 mg/m^2). Four cycles (three weekly) of above drugs were planned according to Italian Sarcoma Group/Scandinavian Sarcoma Group III protocol. This was to be followed by radiotherapy with following protocol: Total radiation dose was 54 Gy, 1.5 Gy twice daily, 5 days/week/36 fractions. She tolerated first cycle of chemotherapy well.

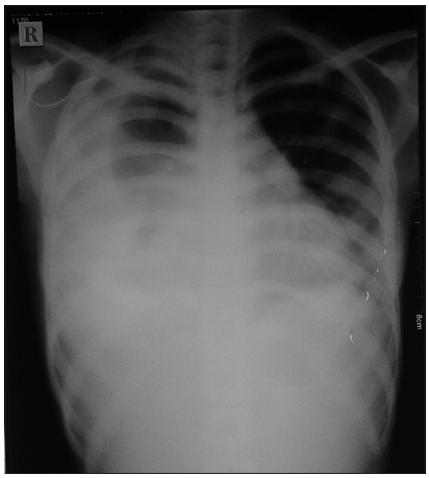

Figure 1: Radiograph of chest showing radioopaque right side

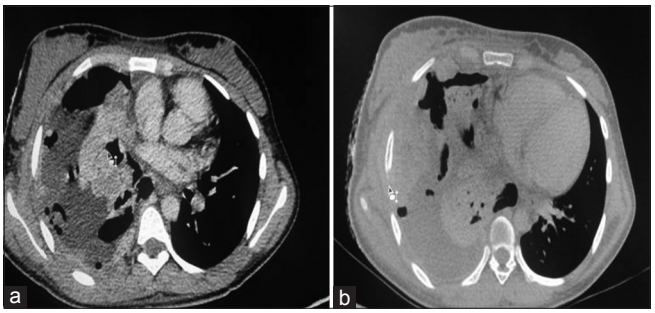

Figure 2: (a) Well-defined oval heterogeneously enhancing soft tissue dense lesion arising from right lateral chest wall; and (b) Erosion of right fifth rib

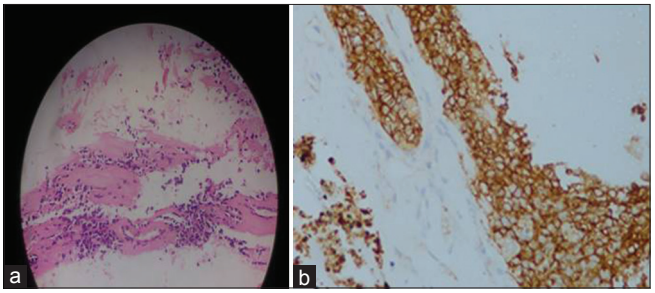

Figure 3: (a) Round cells having scanty basophilic cytoplasm; and (b) CD 99 marker positivity

DISCUSSION

The present case presented to us with right side thoracic mass with right side pleural effusion that was straw colored initially, later on became hemorrhagic. Possibilities of tubercular versus malignant lesions were kept. Malignant lesions included lymphoma, Ewing sarcoma, neuroblastoma, rhabdomyosarcoma, and primitive neuroectoderm tumor. Diagnosis of Askin tumor was established based on the following features: Aggressive nature of presentation and biopsy findings (round cell with basophilic cytoplasm and CD99 positivity). Rest of the malignant lesions were ruled out based on negative findings: Lymphadenopathy and splenomegaly (lymphoma), glycogen granules in cytoplasm (Ewing sarcoma), elevated urinary catecholamine (Neuroblastoma), spindle-shaped cells with acidophilic cytoplasm (rhabdomyosarcoma).

We managed our case with supportive treatment, chemotherapy, and radiotherapy. Chemotherapy protocol (VACAc/IE) was started in line with Italian Sarcoma Group/Scandinavian Sarcoma Group III.[5,6] Local treatment in the form of radiation therapy was planned as parents refused to go for surgery. Till now she has completed four cycles of chemotherapy and is able to perform her activities of daily living well.

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. There are only few case reports from India citing its rarity, clinical presentation, imaging findings, and outcome.[7,8] Recently, a large study by Laskar *et al*. showed the following characteristics in 104 patients: Mean age at presentation (17.9 years), male/female (73/31), swelling (73/104), pain (3/104), swelling and pain (33/104), swelling and dyspnea (8/104), pain and dyspnea (3/104), swelling, pain, and dyspnea (6/104), right hemithorax (53/104), left hemithorax (50/104), sternum (1/104), lung metastasis (19/104), bone metastasis (3/104), liver metastasis (2/104), brain metastasis (1/104), pleural effusion (44/104), lymphadenopathy (11/104), and bone marrow involvement (6/104).[9] Patients were treated with multimodality treatment (neoadjuvant chemotherapy, chemotherapy, surgical, radiotherapy). Poor prognostic indicators were age >18 years, poor response to induction chemotherapy, and presence of pleural effusion. In the above study, the overall survival rate was better in nonmetastatic group compared with metastatic group (65% vs 14%) at the end of 2 years.

There is scarcity of literature highlighting the long-term outcome in such group of patients. First case report citing long-term survival was published in the year 1998.[10] Subsequently, few reports had been published in English literature citing the presence of extensive disease at presentation as poor prognostic factor.[11,12] So, there is a need for development of International registry for such rare cancer patients to document problems associated with long-term survival. 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.[13] Our case presented with pain and dyspnea, right hemithorax opacity, and right-sided pleural effusion with lung and pleural metastasis. The presence of pleural effusion was poor prognostic factor, but localized disease, age (14 years), good response to initial cycle of chemotherapy were favorable factors suggesting good outcome in this patient.

This is a rare tumor mimicking tuberculosis, lymphoma, Ewing sarcoma, neuroblastoma, and rhabdomyosarcoma. Thus, it poses a huge diagnostic and therapeutic challenge to treating clinicians. Timely diagnosis and early intervention can improve the outcome in such patients.

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Conflicts of interest
There are no conflicts of interest.

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