CASE REPORT

Ewing Sarcoma of Chest Wall (Askin's Tumor) in a 3-year-old Child: A Case Report

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Abstract

Fever and difficulty in breathing in a child are common symptoms encountered in routine practice. Often, the underlying cause will be infectious etiology due to bacterial, viral, or tuberculous. However, a malignancy of the paraspinal soft tissue region mimicking infectious disease clinically in a child is challenging to diagnose and treat. Here, we report the Askin's tumor of the chest wall diagnosed in a child presented with fever, worsening dyspnea, and unilateral lung whiteout on a chest radiograph to insist on the importance of extensive and earlier evaluation even for common clinical symptoms.

Keywords: Askin's tumor, Child, Ewing sarcoma, Malignancy.

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Introduction

Fever and difficulty in breathing in a child are common symptoms encountered in routine practice. Often, the underlying cause will be infectious etiology due to bacterial, viral, or tuberculous. Chest X-ray is the first investigation that gives a clue to the underlying cause of dyspnea. In children presenting with worsening dyspnea, unilateral lung whiteout/opaque hemithorax is one of the chest X-ray findings. The most common etiologies for this finding are large pleural effusion, pneumonia involving all lobes of hemithorax, obstruction of the main bronchus, and intrathoracic malignancies involving the chest wall, pleura, lungs, and mediastinum.[1]

Malignancies in children that give unilateral whiteout lung appearance on chest radiographs are pleuropulmonary blastoma, Askin's tumor, Ewing sarcoma, and neuroblastoma.[2,3] These have the potential to grow large enough to shift the mediastinum to the opposite side, causing severe worsening dyspnea. Ultrasound chest or computed tomography (CT) chest is advised for further evaluation of opaque hemithorax. We report the Askin's tumor of the chest wall diagnosed in a child presented with fever, worsening dyspnea, and unilateral lung whiteout on a chest radiograph to insist on the importance of extensive and earlier evaluation even for common clinical symptoms.

Case Description

A 3-year-old male child presented with complaints of fever on and off and progressively worsening difficulty in breathing for a 1-month duration. Parents gave a history of low-grade intermittent fever and not responding to antibiotic therapy. The child also had difficulty in breathing, cough, poor food intake, and poor activity for 1 month. On examination, the child was drowsy, febrile, breathing with the use of accessory muscles of respiration, and oxygen saturation of 88%. There was no pallor, icterus, cyanosis, or peripheral edema. He had tachycardia and tachypnea with normal blood pressure. Respiratory sounds were absent on the left side hemithorax. Other systems were unremarkable. His blood glucose and electrolyte levels were normal. Complete hemogram, erythrocyte sedimentation rate, and C-reactive protein were normal. Nasopharyngeal coronavirus disease 2019 reverse transcription polymerase chain reaction was negative. The chest X-ray showed homogenous opacity on the left hemithorax with tracheal and mediastinal shift to the right side (Fig. 1A). CT of the thorax showed some characteristic heterogeneous enhancing solid and cystic mass arising from the paravertebral soft tissue of left hemithorax eroding posterior aspect of sixth rib with massive left-sided pleural effusion compressing left lung, bronchus, and shifting heart to right side (Figs 1B and C). The histopathology from CT-guided biopsy showed small round cells with scanty cytoplasm and round nucleus with numerous mitotic figures consistent with features of malignant small round cell tumor (Fig. 1D). CT abdomen showed no evidence of ascites or metastasis. Immunohistochemical analysis showed tumor cells to be positive for cluster of differentiation 99 (CD99), vimentin, friend leukemia integration 1 transcription factor (FLI-1), and negative for leucocyte common antigen, desmin, and cytokeratin. Considering the origin of a lesion from the thoracopulmonary region, classic small round cell tumor histology, and immunohistochemical findings, the most probable diagnosis is Askin's tumor. The child was started on neoadjuvant chemotherapy with etoposide and carboplatin.

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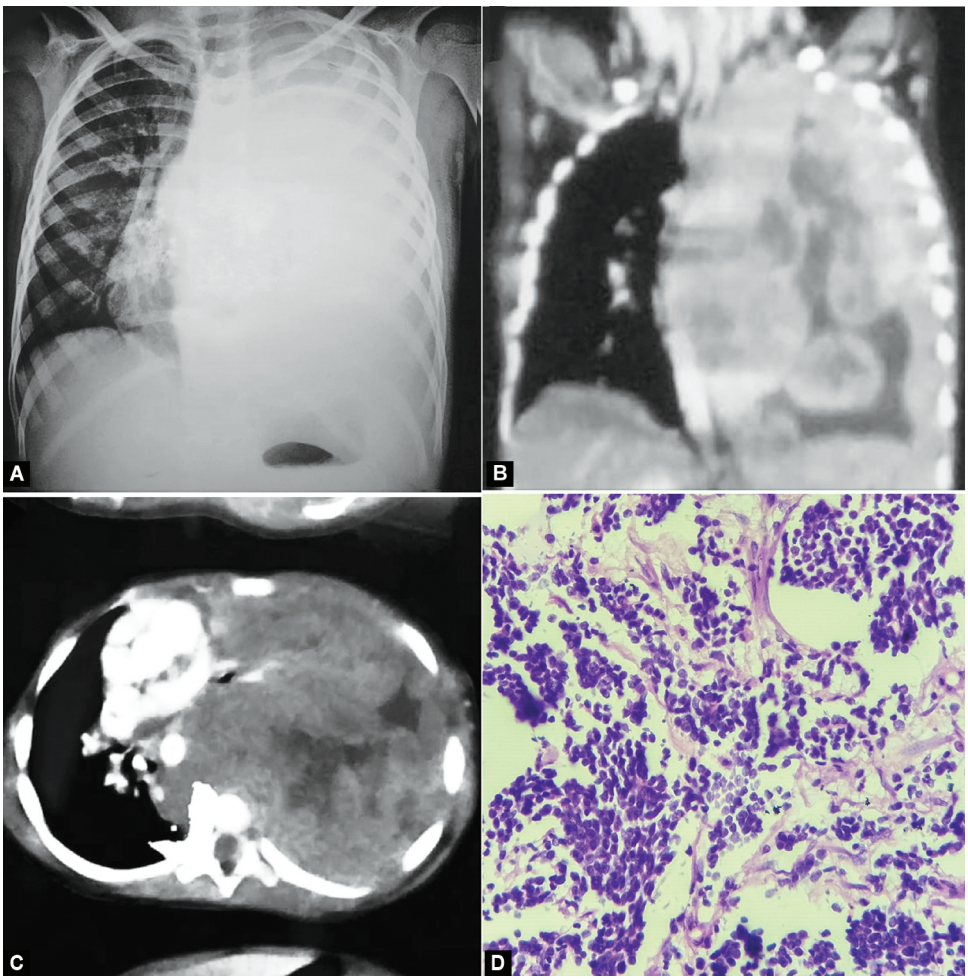

Figs 1A to D: (A) Chest X-ray showing homogenous opacity on left hemithorax with tracheal and mediastinal shift to right side; (B and C) contrast-enhanced CT thorax showing heterogeneous enhancing solid and cystic mass arising from the paravertebral soft tissue of left hemithorax with massive left-sided pleural effusion compressing left lung, bronchus, and shifting heart to right side; (D) Histopathology showing small round cells with scanty cytoplasm and round nucleus with numerous mitotic figures consistent with small round cell tumor

Unfortunately, the child's condition deteriorated progressively in spite of treatment and he died due to respiratory failure within a week of starting chemotherapy.

Discussion

Askin's tumor is a rare, aggressive primitive neuroectodermal tumor (PNET) arising from paravertebral soft tissues of the thoracopulmonary region, classified as Ewings sarcoma/PNET by the World Health Organization in 2002.[4–6] The prevalence of the disease is 0.2 cases/million.[7] The clinical picture usually includes subacute onset and slow progression of nonspecific symptoms like palpable mass in the thorax, fever, cough, pleuritic chest pain, dyspnea, and weight loss.[4,5]

Chest radiograph usually shows unilateral whiteout lung/opaque hemithorax due to a large infiltrative Askin's tumor. On CT, these tumors typically show ill-defined mass with heterogeneous attenuation and multiple loci of cystic degeneration. Contrast administration shows enhancement of solid components. Rarely pleural effusions and calcifications can be seen.[6] Magnetic resonance imaging shows heterogeneous mass due to numerous areas of necrosis, hemorrhage, and cystic degenerative changes.[5,6] Smaller tumors tend to be homogeneous.

Histologically, characteristic round, small, and slightly differentiated cells organized in strands are seen along with occasional pseudorosettes. Electron microscopy shows neurosecretory granules, and immunohistochemical markers like CD99, FLI-1, neuron-specific enolase, and vimentin are typically positive.[5] Inflammatory disorders and other malignancies are differential diagnoses. Clinical symptoms of Askin's tumor mimic inflammatory disorders like osteomyelitis, empyema, and tuberculosis. The differential diagnoses for chest wall soft tissue malignancy in children include lymphoma, osteosarcoma, and rhabdomyosarcoma.

Combined multimodality therapy, such as chemotherapy, radiotherapy, and wide surgical resection is recommended for Askin's tumors. Even with such therapy, the prognosis of the patient remains poor.[7]

Conclusion

Askin's tumor is a rare, aggressive malignancy that should always be considered in a differential diagnosis of soft tissue mass lesion in the thorax for early diagnosis and treatment.

Consent to Participate

Written consent was obtained.

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