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Case Report

Extraosseous Primary Ewing's Sarcoma of the Lungs Mistakenly Diagnosed and Managed as Hemothorax: An Extremely Aggressive and Fatal Rare Tumor with an Unusual Site

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Abstract Extraosseous Ewing's sarcoma of the lungs is a rare soft tissue tumor of children and adolescents usually found in the extremities. In this case report, we present a 4-year-old male child with primary Ewing Sarcoma of the lungs who presented with shortness of breath, cough, left-side chest pain, and hemoptysis for two months. We reviewed clinical, radiological, and pathological findings for this rare malignancy.

Keywords Extraosseous Ewing's sarcoma; Lungs; Tumor; Computed tomography.

Introduction

Ewing's sarcomas are rare neuroectodermal tumors that primarily arise from the bone and are the second most common primary bone tumor (1). James Ewing first described this tumor in 1921 as an endothelioma of bone (2). Extraosseous Ewing's sarcomas are sporadic neuroectodermal tumors, and Hammer et al. reported the first case in 1989 (3). Herein, we report a primary extraskeletal Ewing's sarcoma affecting the lungs diagnosed at a tertiary care

hospital in Mogadishu, Somalia. Considering the disease's clinical, radiological, and pathological features, the role of an integrated approach has been highlighted.

Case Report

A 4-year-old child complained of shortness of breath, cough, left-side chest pain, and hemoptysis for two months before admission to Somalia Turkiye Training and Research Hospital. On respiratory examination, air entry of the right lung site was markedly decreased on auscultation, and the percussion was dull on the left hemithorax. Other systemic examinations were unremarkable. Over the previous year, the family reported visiting up to 5 hospitals due to similar complaints. The last hospital that referred the patient to our hospital performed a chest X-ray, detected a massive left-sided pleural effusion, and planned to insert a chest tube. After the chest tube insertion, the patient's condition worsened, and they referred him to our hospital. Blood tests and a chest x-ray were performed when the patient reached our hospital. His blood investigations were unremarkable, but the chest x-ray demonstrated a completely homogenous opacification in the left hemothorax with an extensive mediastinal shift towards the right side. In addition, a chest tube was detected between the seventh and eighth intercostal space on the left side. The tube was twisted and oriented upward due to a solid mass lesion (Figure 1). We admitted the patient for further evaluation and removed the chest tube. A thoracic ultrasound was performed to rule out effusion, and unexpectedly this scan demonstrated a large hypoechoic heterogeneous solid mass. It was hypervascular in color Doppler examination, and multiple areas of cystic degeneration were present in the left hemithorax (Figure 2).

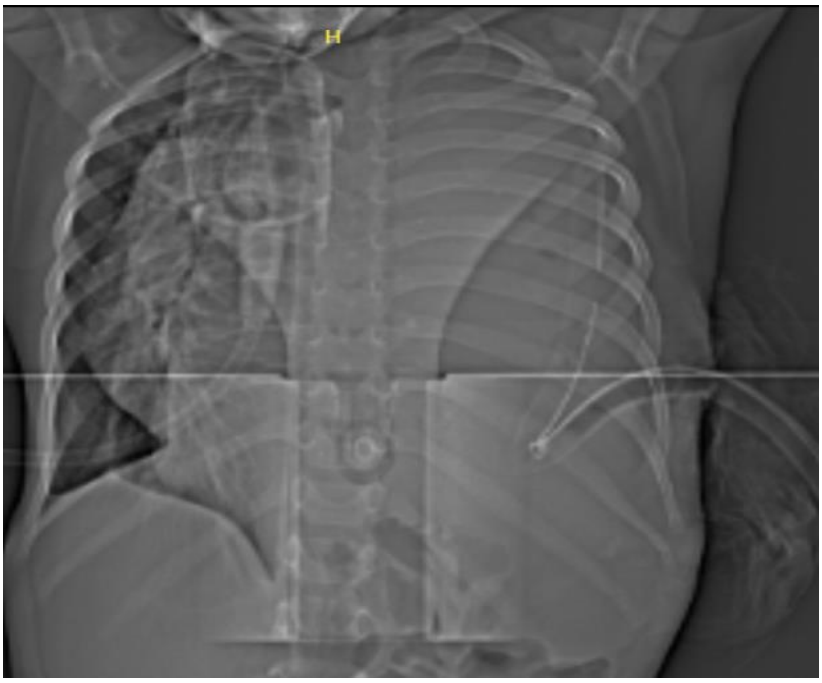

Figure 1. Frontal chest X-ray showing complete opacification of the left hemithorax, a chest tube that twisted and oriented upward.

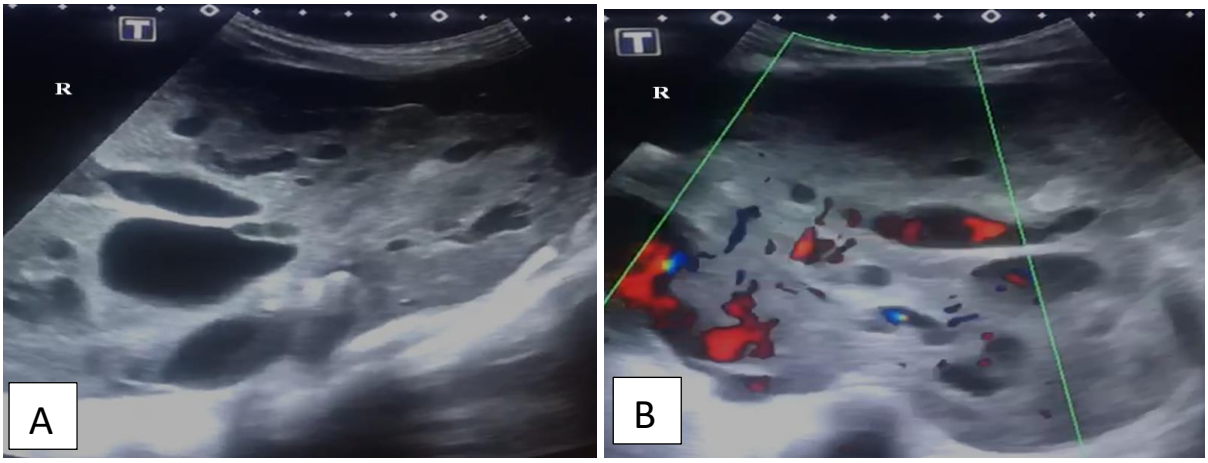

Figure 2. A-Chest ultrasound shows a large hypoechoic area containing multiple cystic changes in solid mass lesions. **B-**Chest Doppler ultrasound revealed a large hypoechoic solid lesion that is highly vascular in some areas.

A thoracal contrast-enhanced computerized tomography (CT) scan was performed to characterize the lesion further. This scan revealed a large heterogeneous, contrast-enhanced mass with multiple cysts and hypodense areas involving the entire left hemithorax. Also, the mediastinum shifted to the right side due to compression. No calcification was detected within the mass (Figure 3). Subsequently, a whole-body CT scan was performed, and no distant metastasis was detected. A tru-cut biopsy was performed, which revealed Ewing sarcoma of

the chest wall. The patient was referred to the oncology department. However, he died due to disease progression and respiratory failure soon after referral.

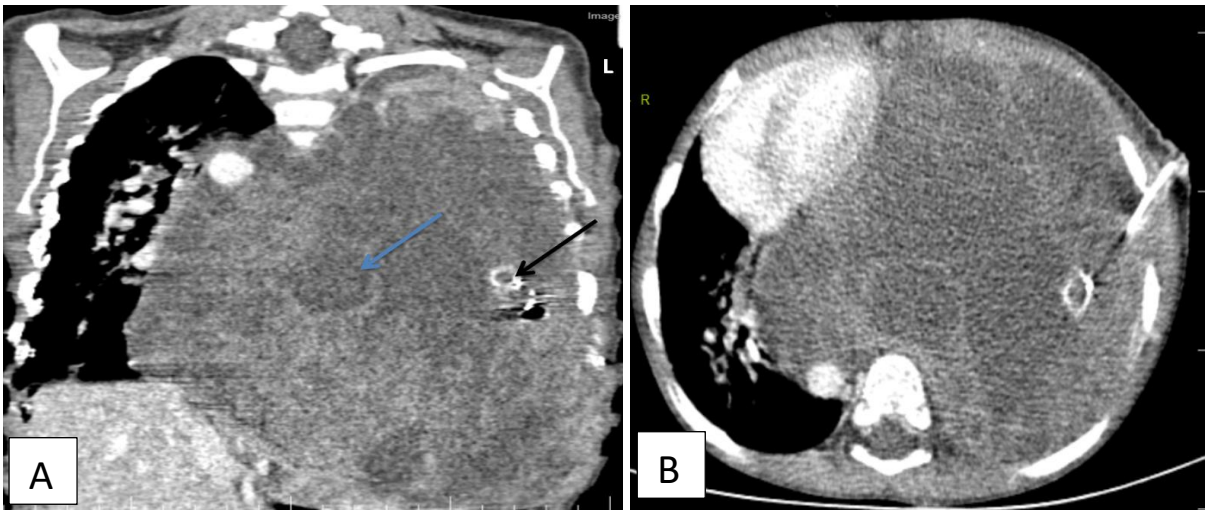

Figure 3. A-Coronal computerized tomography scan of the chest showing complete opacification at the left hemithorax heterogeneously enhanced with hypodense necrotic areas (blue arrow) and marked mass effect with a right-sided mediastinal shift. The black arrow shows the twisted chest tube. **B-** An axial section showing the right-sided mediastinal shift with markedly reduced right lung volume.

Discussion

Extraosseous Ewing's sarcomas (EES) is a sporadic neuroectodermal tumor initially described by Hammer et al. in 1989(3). Our case was a 4-year-old child. However, it is usually diagnosed in the second decade of life (1). This finding indicates a possible early presentation in our study (1).

Although the diagnosis of EES is primarily based on histology, clinical and radiological features also have a significant role in the diagnostic management of EES and its differential diagnosis from other sarcomas (4). A computerized tomography scan helps describe the tumor's extent and confirm that the soft tissue mass is extraosseous. In the present case, an enhanced thoracal CT scan revealed a heterogeneously enhanced mass with multiple cystic necrosis areas involving the entire left hemithorax associated with a right-sided mediastinal shift due to compression. No calcification was detected within the tumor. Also, a whole-body CT scan was performed, which excluded distant metastasis.

Typically, EES is an aggressive tumor with a high risk for local recurrences and metastases (5). Therefore, the treatment of choice is an early surgical intervention with intensive chemotherapy and radiation therapy to eliminate a potential residual microscopic disease. Our patient had no distant metastasis but died due to local disease-related complications.

Conclusion

Primary EES is a scarce soft tissue neuroectodermal tumor. Due to its rarity and aggressive character, there are no guidelines for managing this disease. In addition, since it is rare, it may be overlooked during the differential diagnosis stage of patients with malignant tumors. Although there is a high risk of distant metastasis, patients may also die due to the aggressiveness of the local disease and erroneous interventions. Therefore, performed needed combined modality treatment. Also, primary EES should be considered in the differential diagnosis of children and young adults presenting with primary pulmonary mass.

Informed consent

The child's parents provided permission (written and informed consent) to publish this case report.

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