# Extra-osseous 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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**Case Report**

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# Abstract

Extraosseous Ewing's sarcoma of the lungs is a rare soft tissue tumor of pediatric and adolescent usually found in the extremities. In this case report, we describe such a case in a four year-old-male child 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. This rare and fatal malignancy has been misdiagnosed and managed inappropriately because of low suspicion of index of taking this into the differential diagnosis.


# Introduction

Ewing’s sarcomas are rare neuroectodermal tumors that primarily arise from the bone and are the second most common primary bone tumor (1). This tumor, firstly described by James Ewing in 1921 as an endothelioma of bone (2). Extraosseous Ewing’s sarcomas are extremely rare neuroectodermal tumors, and Hammer et al had reported the first case in 1989(3). Here we report a case of primary extraskeletal Ewing’s sarcoma affecting the lungs that was diagnosed at our tertiary care teaching hospital, Mogadishu, Somalia. The role of an integrated approach, considering the clinical, radiological and pathological features of the disease has been highlighted.

# Case Report

A 4-year-old child complained of shortness of breath, cough, left site chest pain and hemoptesis for two months before admission to Mogadishu Somali Turkish Training and Research Hospital. On respiratory examination, air entry of right site of the lung was markedly decreased on auscultation, and the percussion note was dull on the left hemithorax. Other systems examinations were unremarkable. Over the previous year, the family reported that they visited up to 5 different hospitals due to similar complaints that the patient presented to our institution. The last institution that the patient referred to our hospital done chest ray and then they were diagnosed as massive left side pleural effusion and planed to insert a chest tube. After insertion of the chest tube, the patient's condition worsened, and they referred him to our hospital. When the patient reached to our hospital, we send him blood tests and a chest x-ray. His blood investigations were unremarkable, but the chest x-ray demonstrated a completely homogenous opacification in the left hemothorax with extensive mediastinal shift towards the right side and a chest tube was in the between 7 and 8 intercostal space of the left chest wall. The tube is twisted and turned upward due to solid mass lesion (Fig. 1) and we decided to admit the patient for further evaluation. Then we removed the chest tube, and decided to do ultrasound since it is first choice diagnostic modality for pediatric patients, therefore we performed a thoracic ultrasound to role out effusion, and unexpectedly ultrasound demonstrated a large hypoechoic heterogeneous solid mass with hipervascular in clour doppler examination and multiple areas of cystic degeneration occupied by the entire left hemithorax (Fig. 2a and 2b). For further additional imaging is required for lesion charactrisation, Contrast enhanced chest CT-scan was performed that revealed large heterogeneous enhanced mass with multiple cystic necrosis hypodense areas involving the whole left hemithorax associated with compression of the

mediastinum to the right side. No calcification had seen within the mass (Fig. 3a and 3b). Also, we have done a whole-body CT scan and no metastasis had detected. A true-cut biopsy was performed and histopathologically confirmed as an Ewing sarcoma of the chest wall. After admission, one day letter we referred to the oncology center, unfortunately before they started management the child passed away.

# Discussion

Extraosseous Ewing's sarcomas (EES) are extremely rare neuroectodermal tumors, and Hammer et al had reported the first case in 1989(3). In our case, the child was four years old. But the usual age scope of the preponderance of the published cases was in the second decade of life, indicating a possible early presentation or delayed diagnosis in our study (1).

Although the diagnosis of EES primarily based on histology, also clinical and radiological features had a significant role in the diagnose of EES and differential diagnosis from other sarcomas (4). An especially CT scan is useful in describing the extent of the tumor and confirm that the soft tissue mass is completely extraosseous. In the present case, an enhanced chest CT-scan had performed that revealed heterogeneously enhanced mass with multiple cystic necrosis hypodense areas involving the whole left hemithorax associated with compression of the mediastinum to the right side. No calcification had detected within the tumor. Also, we have done a whole-body CT scan, and no metastasis had demonstrated. Extraosseous Ewing's Sarcoma is an aggressive tumor having local recurrences and metastases but, the treatment of choice is an early surgical intervention with intensive chemotherapy and radiation therapy to eliminate any residual microscopic disease (5). Even though our patient had an aggressive character of the tumor, fortunately, no metastasis had detected.

In conclusion, primary extraosseous Ewing's sarcoma is an extremely rare soft tissue neuroectodermal tumor that is histologically imperceptible from the other osseous form. Due to its rarity and aggressive character, it has no particular guidelines for the management of this disease and needed combined modality treatment. Also, it should be considered in the differential diagnosis of children and young adults presenting with primary pulmonary mass.

# Declarations

### Ethical Approval:

The child  was treated in accordance with the ethical standards of institutional and national committees on human experimentation and with the Declaration of Helsinki (1975, and subsequent revisions). According to the guidelines of the Ethics Committee of Somali Turkey Recep Tayyip Erdogan Hospital institutional approval was not required to publish the case details Informed Consent.

The child's parents provided permission (written informed consent) for the publication of the details of their child and the images shown in the figure.

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Conflict of interest and Funding

None.

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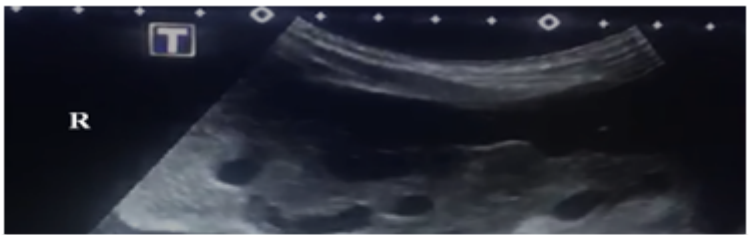
# Figures

**Figure 1**

AP chest x-ray view showed complete opacification of the left hemithorax Due to resistance of the solid lesion tube twisted and extends upward.




## Figure 2

**a:** Chest ultrasound shows large hypoechoic containing multiple cystic changes in the solid mass lesions.

**b:** Chest doppler ultrasound revealed a large hypoechoic solid lesion having vascularity in some areas.

## Figure 3

**a:** Coronal chest CT showed Complete opacification at the left hemithorax heterogeneously enhanced with hypodense necrotic areas ( blue arrow) and marked mass effect and shift of the mediastinum towards the right. While the black arrow shows twisted tube.

b: An axial chest CT showed mediastinum structures markedly shift of the to the right and reduced right lung volume.