To cite this article: Adeoye PO, Abubakar HI, Dominic AA, Udo R. Primary mediastinal small round cell tumor suggestive of Ewing's sarcoma associated with pneumothorax: a case report. Curr Thorac Surg 2026;11(2):214-219.

Case Report

Primary mediastinal small round cell tumor suggestive of Ewing's sarcoma associated with pneumothorax: a case report

Peter O. Adeoye[1,2], Hassan I. Abubakar[2*], Akolo A. Dominic[3], Robert Udo[2]

[1]Department of Surgery, Division of Thoracic and Cardiovascular Surgery, University of Ilorin and University of Ilorin Teaching Hospital, Kwara, Nigeria
[2]Department of Surgery, Division of Cardiothoracic Surgery, Federal Medical Centre Keffi, Nasarawa, Nigeria
[3]Department of Anatomic Pathology and Forensic Medicine, Federal Medical Centre Keffi, Nasarawa, Nigeria

ABSTRACT

Ewing's sarcoma (ES) is a rare form of malignancy that commonly affects the bone. Extraskeletal ES (EES) accounts for 20% of ES cases. The mediastinum is an exceedingly rare primary site of EES and presents with non-specific symptoms that may mimic several other thoracic pathologies. We describe a suspected case of primary mediastinal ES presenting with pneumothorax in a 17-year-old female who presented with chronic cough, dyspnea, and chest pain. The patient had been commenced on empirical treatment for tuberculosis (a common diagnosis in our sub-region) for six weeks before presentation to our facility, due to a lack of improvement in symptoms. Chest computed tomography scan revealed a huge anterior mediastinal mass extending into the left hemithorax with pneumothorax on the right. She underwent a transthoracic percutaneous biopsy, and histological analysis revealed a monomorphic small-round-cell morphology, suggestive of primary mediastinal Ewing's sarcoma.

Keywords: mediastinal neoplasms, Ewing's sarcoma, pneumothorax, neuroectodermal tumors, extraskeletal

Corresponding Author*: Hassan Abubakar, MD. Federal Medical Centre Keffi, Nasarawa State, Nigeria.
E-mail: mrhassan.i.a16@gmail.com
DOI: 10.26663/cts.2026.031
Received 03.06.2026 Accepted 23.07.2026

Introduction

Ewing's sarcoma (ES) is a rare aggressive tumor postulated to be of neuroectodermal or mesenchymal origin with a slight male predominance. It has a higher incidence in Whites than in people of African and Asian descent, with peak incidence seen between the ages of 10 and 19 years. ES commonly affects the bones and rarely the extra-skeletal soft tissues. There is a paucity of data on the exact incidence of ES in Sub-Saharan Africa [1].

Extraskeletal ES (EES) has an incidence of 0.4/1,000,000 and represents about 20% of ES cases. EES has a bimodal distribution with peak incidence in individuals < 5years and > 35years old. EES, unlike ES of the bone, shows no predilection for biological sex and race. The upper thigh, buttocks, upper arm, and shoulders are the most common sites of EES [2]. The mediastinum is an exceedingly rare primary site of EES and thus presents a diagnostic dilemma due to nonspecific symptoms that may suggest commoner diseases such as Tuberculosis (TB) in high TB-burden countries [3]. We report a presumed case of primary mediastinal ES with a distinctive presentation of pneumothorax and review the literature.

Case Report

A 17-year-old female presented to the emergency department of our tertiary referral center with a three-month history of persistent cough, a two-month history of progressive shortness of breath, and right-sided chest pain of one-week duration, which worsened a day before her presentation. The cough was productive of whitish sputum, but there was no hemoptysis. There was a history of unintentional weight loss in the preceding month. She had been commenced on anti-tuberculosis (TB) drugs (isoniazid, rifampin, pyrazinamide, and ethambutol) empirically at a peripheral center, six weeks before her presentation. Her family denied a history of malignancy.

On examination, she had a temperature of 37.0 ^{0}C, blood pressure of 110/70 mmHg, heart rate of 120 beats/min, respiratory rate of 46 breaths/min, oxygen saturation of 98% in room air, and grade III digital clubbing. Chest examination revealed trachea deviation to the right, stony dull percussion note, and absent breath sounds on the left hemithorax. There was no significant peripheral lymphadenopathy, hepatosplenomegaly, or jaundice.

Chest X-ray showed an ill-defined homogenous opacity in the left hemithorax with a mass-induced mediastinal shift to the right, where a pneumothorax was also seen (Figure 1).

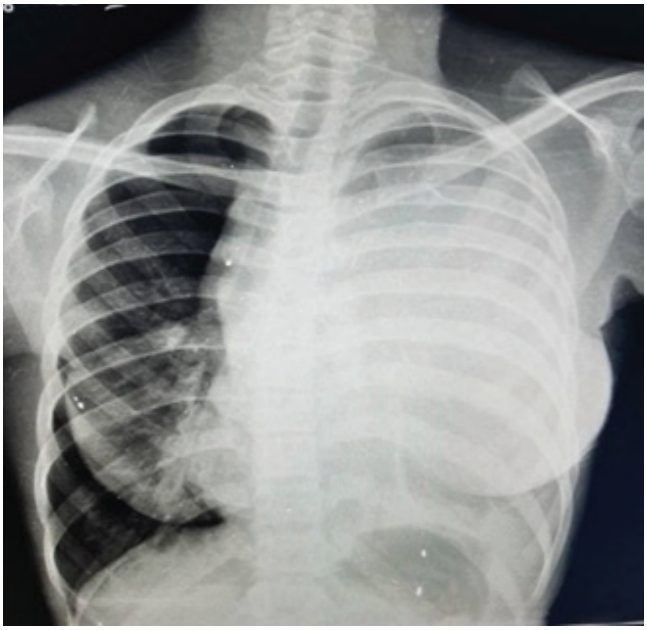

Figure 1. Plain chest radiograph showing a dense, homogeneous opacity throughout the left hemithorax with a massive mediastinal shift to the right, accompanied by a right-sided pneumothorax.

Chest CT scan showed a huge mixed-density anterior mediastinal mass measuring 17.3 x 16.4 x 12.6 cm with irregular margins and areas of necrosis extending into the left hemithorax. The mass occupies almost the entire left hemithorax with associated compression atelectasis of the left lung. It encases the arch of the aorta, superior vena cava, main pulmonary artery, and the descending thoracic aorta. It is adherent to the left side of the pericardium. Right-sided pneumothorax can also be seen (Figures 2a-d). No area of calcification, mediastinal lymphadenopathy, pleural effusion or hepatic metastasis noted.

Laboratory evaluation revealed a markedly elevated erythrocyte sedimentation rate (95 mm/hr). Total leucocyte count was 6.3×109/L, packed cell volume of 30%, serum urea: 3.8 mmol/L, creatinine: 75.4 μmol/L, and serum electrolytes were within normal limits. Sputum evaluation was negative for acid-fast bacilli, mycobacterium tuberculosis was not detected on GeneXpert, and cytology was negative for malignancy.

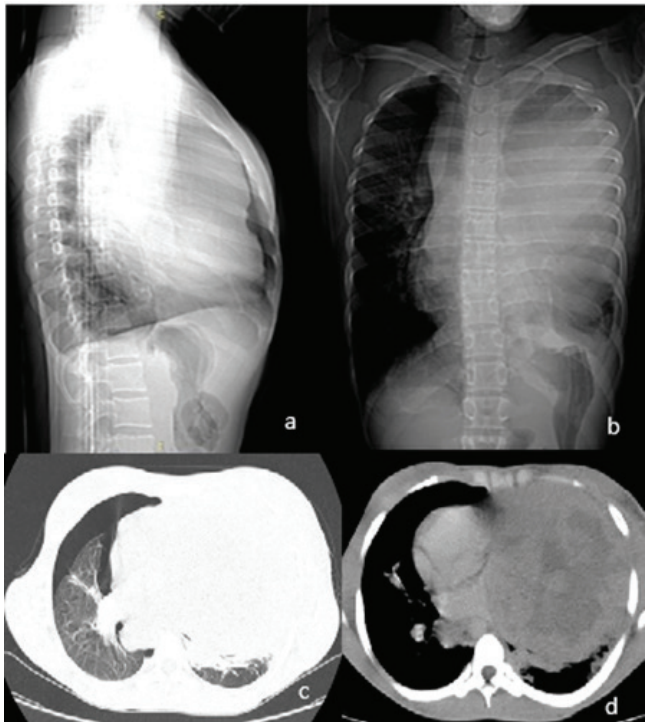

Figure 2. Chest CT scan showing a huge mixed-density anterior mediastinal mass measuring 17.3 x 16.4 x 12.6 cm with irregular margins and areas of necrosis extending into the left hemithorax. The mass occupies almost the entire left hemithorax with associated compression atelectasis of the left lung. It encases the arch of the aorta, superior vena cava, main pulmonary artery, and the descending thoracic aorta. It is adherent to the left side of the pericardium, and a right-sided pneumothorax can also be seen (**a-d**).

She had a closed thoracostomy tube drainage for the right-sided pneumothorax and symptomatic treatment. Anti-TB medications were discontinued following the results of laboratory investigations, and she underwent a trans-thoracic percutaneous Tru-cut biopsy of the mediastinal tumor. She was discharged after 7 days of admission following improvement of symptoms and was adequately counseled to see the next scheduled specialist clinic with the histology report.

Histological sections showed sheets of monotonous primitive small round blue cells with a high nuclear-to-cytoplasmic ratio, hyperchromatic nuclei, some irregular nuclear contours with fine stippled chromatin, inconspicuous nucleoli, and scanty cytoplasm with an indistinct membrane. No mitotic activity or areas of necrosis or hemorrhage were seen; features highly consistent with a small round-cell malignancy, such as Ewing's sarcoma (Figure 3).

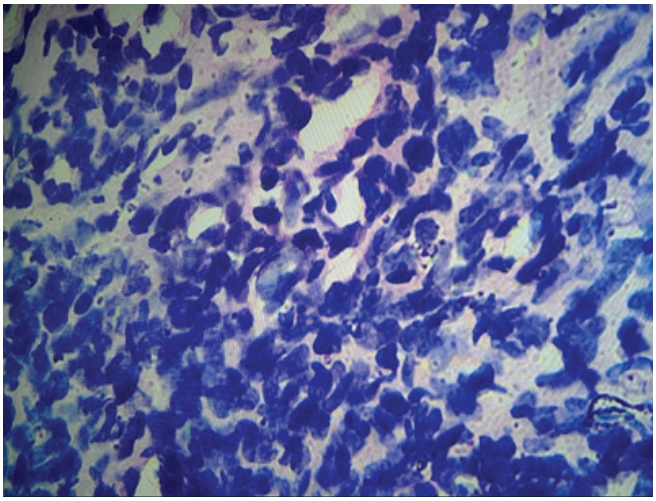

Figure 3. Hematoxylin and eosin staining of the tumor showing small round blue cells with enlarged nuclei, fine stippled chromatin, inconspicuous nucleoli, and scanty clear to eosinophilic cytoplasm with indistinct membrane. Features consistent with classical Ewing's sarcoma (H&E, ×400).

Unfortunately, our patient only re-presented after four weeks of initial discharge with worsening of her symptoms, as her parent had attributed her illness to a "spiritual attack", which stems from a magico-religious thought system of disease causation that is not uncommon in our sub-region [4]. In a sad turn of events, our patient succumbed to her illness within 2 days of re-presentation to the clinic. A deceased patient's permission form was signed by the patient's next of kin for the publication of this case report.

Discussion

ES typically has a rapid growth pattern with most patients harboring micro-metastasis at presentation. Clinical manifestation of primary mediastinal ES is highly variable with non-specific symptoms, this in addition to its very low incidence, poses a diagnostic dilemma. Patients with mediastinal ES can present with cough, wheezing, and chest tightness when the tumor compresses the trachea. Pleural invasion with resultant effusion or pneumothorax, as in this index case, as well as invasion of intercostal nerves, may cause chest pain and dyspnea. Invasion of the pericardium may present with pericardial effusion, while compression of the heart or great vessels can lead to mediastinal shift and superior vena cava syndrome [3, 5].

Imaging plays a vital role in the management of EES. Although CT scan features are non-specific, they remain

vital for assessing tumor size and resectability. Commonly seen features include a slightly hypodense soft tissue mass with areas of cystic degeneration on non-contrast CT, while contrast-enhanced medium demonstrates a mass with a mixed-density pattern. Calcification is seen in 10% of cases. Pleural effusion, pneumothorax, and pericardial effusion are manifestations of invasion of nearby structures that may be seen [6-8].

Histopathological analysis is required to confirm the diagnosis of EES. The histological picture of EES consists of a large number of uniform small round cells with a high nuclear-to-cytoplasmic ratio and scanty clear to eosinophilic cytoplasm [9].

Immunohistochemistry (IHC) is an indispensable tool in the diagnosis of ES. Positive staining for Cluster of differentiation 99 (CD99), Friend leukemia integration 1 (FLI-1), and NKX2 are currently acceptable markers for diagnosis of ES on IHC. Additionally, genetic testing is helpful for definitive diagnosis, and the presence of EWS-FLI1 fusion gene defines ES on genetic testing [9-11].

What makes this case particularly unique is the rare clinical presentation of a primary mediastinal tumor skewed to the left co-occurring with a contralateral pneumothorax and resultant paradoxical mediastinal deviation. In classic physiology, a right-sided pneumothorax exerts a positive pressure effect that shifts mediastinal structures toward the left hemithorax. However, in our patient, physical and radiological examinations revealed a severe tracheal and mediastinal shift to the right. This paradoxical finding is explained by the massive volume and dominant compressive force of the left-sided anterior mediastinal mass, which completely overwhelmed the contralateral pressure of the right pneumothorax. Such a presentation can easily mislead clinicians, especially in resource-limited settings where symptoms might be misattributed solely to common infectious pathologies like tuberculosis.

A major limitation of this report is the lack of immunohistochemical (IHC) confirmation and molecular or genetic testing (such as detecting the EWSR1-FLI1 fusion gene), which are vital to establish a definitive diagnosis of Ewing's sarcoma. Due to an inadequate tissue block from the initial biopsy, coupled with the patient's late re-presentation and rapid demise, these ancillary studies could not be completed. Consequently, a broad differential diagnosis for small round blue cell tumors of the mediastinum must be considered. This includes lymphoma, embryonal rhabdomyosarcoma, neuroblastoma, poorly differentiated synovial sarcoma, and newer entities like BCOR or CIC-rearranged sarcomas [12]. While the cytomorphological features were highly suggestive of extraskeletal Ewing's sarcoma, the diagnosis remains presumptive rather than definitive.

Early diagnosis and treatment can improve the prognosis of EES. Treatment involves a combination of systemic chemotherapy and local control either by surgery or radiotherapy. Generally, induction with multidrug chemotherapy is given before local control. Surgery is the preferred method of local control for resectable tumors, as the long-term outcome when radiotherapy is used alone is less favorable, even though ES is radiosensitive [13,14]. Autologous stem cell therapy has also been used in the management of EES [9].

Our literature search identified six documented cases of primary mediastinal ES. Three of the cases were in males and 3 females; the ages ranged from 15 to 66 years. As in our case, the anterior mediastinum was the primary site of the tumor in 3 cases; 2 were located in the posterior mediastinum, and 1 involved both the anterior and posterior mediastinum. Cough was present in 3 out of 4 cases involving the anterior mediastinum, a finding fairly consistent with this index case, while pain was the predominant symptom in the posterior mediastinal tumors. There was an associated pleural effusion in 3 of the 6 cases, and 1 of the 3 cases with pleural effusion had pericardial effusion in addition. Notably, our case is distinguished by the presence of a primary mediastinal small round cell tumor complicated by pneumothorax, a finding we believe is unprecedented in prior case reports. At a minimum, all cases were diagnosed using histology, while 5 included further IHC testing and 3 involved genetic studies. Four of the six patients had surgery as their primary treatment [3,5,7-9,15].

In Table 1, we summarize the management and outcome of six cases of primary mediastinal ES previously reported in the literature.

Table 1. Management and outcome of some previously reported cases of primary mediastinal Ewing's sarcoma.

Study/ Year	Age/ Sex	Presenting complaints	CT scan Features		Method of Diagnosis	Treatment	Outcome
			Primary Site	Description			
Li X et al. [3] 2023	15/F	Cough Wheezing	Anterior mediastinum	15.7x11.7x10.5 cm moderately heterogenous enhancing mass with multiple mediastinal nodes and invading left lung with ipsilateral pleural effusion	Histology + IHC + Genetic testing	Chemotherapy	Follow-up
Su C et al. [5] 2024	32/M	Chest pain Back pain	Posterior mediastinum	4x4x3 cm well-defined mass with heterogenous density	Histology + IHC + Genetic testing	Surgery	Follow-up
Halefoglu AM et al. [7] 2012	19/F	Pain in the left upper arm	Posterior mediastinum	16x15x15cm heterogenous mass with smooth edges extending to the left hemithorax and supraclavicular region, plus contralateral mediastinal shift	Histology + IHC	Surgery	N/A
Cui M et al. [8] 2023	66/M	Chest tightness	Anterior mediastinum	Giant cystic and solid mass with moderate enhancement and enlarged mediastinal nodes. Invasion of SVC, Pericardium with pericardial effusion, and right lung with ipsilateral pleural effusion	Histology + IHC	Surgery	Metastatic pericardial effusion reappeared two months after surgery. Death
Caltavituro A et al. [9] 2023	30/F	Cough Dyspnea	Anterior mediastinum	Well-defined solid mass extending to the left hemithorax compressing the superior lung lobe and contralateral mediastinal dislocation	Histology + IHC + Genetic studies	Surgery + adjuvant chemotherapy + autologous stem cell transplant	Relapse after a few months of surgery. On follow-up after adjuvant therapy.
Ata F et al. [15] 2021	16/M	Cough Dyspnea	Anterior and posterior mediastinum	18cm mediastinal mass extending to the left hemithorax with ipsilateral pleural effusion and contralateral basal nodule and mediastinal shift	Histology	N/A	N/A

Abbrev.: M: Male, F: Female, CT: Computed tomography, IHC: Immunohistochemistry, N/A: Not applicable (i.e.: details not provided by publishing authors).

In conclusion, primary mediastinal tumors consistent with Ewing's sarcoma are rare, aggressive malignancies that pose significant diagnostic challenges. This case highlights a highly unusual presentation characterized by a contralateral pneumothorax and paradoxical mediastinal shift. In high-burden regions, such presentations can mask underlying malignancies and lead to empirical tuberculosis treatment, delaying diagnosis. Although the lack of immunohistochemical and molecular confirmation in this case limits a definitive diagnosis, clinicians should maintain a high index of suspicion for small round cell malignancies when evaluating massive mediastinal tumors with atypical intrathoracic pressure dynamics.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support.

Authors' contribution

POA,RU were responsible for the initial assessment, the request, and the interpretation of radiological findings. POA,HIA contributed to the biopsy and closed tube thoracotomy procedures. AAD was responsible for the histological analysis. POA, HIA, and AAD were responsible for drafting the manuscript. RU and HIA oversaw the literature review and prepared the figures. All authors have reviewed and approved the final manuscript.

Acknowledgements

We would like to thank the patient's parent for the consent to report this study and the nurses and clinical staff who dedicatedly provided care to the patient.

References

1. Spector LG, Hubbard AK, Diessner BJ, Machiela MJ, Webber BR, Schiffman JD. Comparative international incidence of Ewing sarcoma 1988 to 2012. Int J Cancer 2021; 149: 1054-66.
2. Abboud A, Masrouha K, Saliba M, Haidar R, Saab R, Khoury N et al. Extraskeletal Ewing sarcoma: Diagnosis, management and prognosis (Review). Oncol Lett 2021; 21: 354.
3. Li X, Qi S, Zhu T, Jiang Y, Wang W. Primary mediastinum Ewing's sarcoma with pleural effusion: A case report and literature review. Open Life Sci 2023; 18: 20220669.
4. Adegoke A. Factors influencing health beliefs among people in south west, Nigeria. Afr Res Rev 2008; 2: 177-97.
5. Su C, Zhu X, Zhang J. Primary mediastinal Ewing's sarcoma presenting with sudden and severe chest pain: a case report. Front Oncol 2024; 13: 1290603.
6. Wright A, Desai M, Bolan CW, Badawy M, Guccione J, Rao Korivi B et al. Extraskeletal Ewing sarcoma from head to toe: multimodality imaging review. Radiographics 2022; 42: 1145-60.
7. Halefoglu AM. Extraskeletal Ewing's sarcoma presenting as a posterior mediastinal mass. Arch Bronconeumol 2013; 49: 82-4.
8. Cui M, Zhai D, Liu Y, Zhou X, Wang T, Wang L et al. Case report: Primary mediastinal Ewing's sarcoma presenting with chest tightness. Front Oncol 2023; 12: 1020339.
9. Caltavituro A, Buonaiuto R, Salomone F, Morra R, Pietroluongo E, De Placido P et al. Extraskeletal Ewing's sarcoma of the mediastinum: Case report. Front Oncol 2023; 13: 1074378.
10. Tsuda Y, Zhang L, Meyers P, Tap WD, Healey JH, Antonescu CR. The clinical heterogeneity of round cell sarcomas with EWSR1/FUS gene fusions: Impact of gene fusion type on clinical features and outcome. Genes Chromosomes Cancer 2020; 59: 525-34.
11. Machado I, Yoshida A, López-Guerrero JA, Nieto MG, Navarro S, Picci P et al. Immunohistochemical analysis of NKX2.2, ETV4, and BCOR in a large series of genetically confirmed Ewing sarcoma family of tumors. Pathol Res Pract 2017; 213: 1048-53.
12. Dehner CA, Lazar AJ, Chrisinger JS. Updates on WHO classification for small round cell tumors: Ewing sarcoma vs. everything else. Hum Pathol 2024; 147: 101-13.
13. Hogendoorn P, Athanasou N, Bielack S, De Alava E, Dei Tos A, Ferrari S et al. Bone sarcomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. Ann Oncol 2010; 21 Suppl 5: v204-13.
14. Khanna N, Pandey A, Bajpai J. Metastatic Ewing's sarcoma: revisiting the "evidence on the fence". Indian J Med Paediatr Oncol 2017; 38: 173-81.
15. Ata F, Safwan Aljafar M, Mohammed AM, Mirza S, Zafar AA. Primary Mediastinal Ewing sarcoma presenting as a massive lung lesion with a mediastinal shift. Clin Case Rep 2021; 9: e04857.