Journal of
Medical Case Reports

CASE REPORT

Open Access

Ewing sarcoma presenting in the lung: a case report

Mohammad Berro[1], Abdulrahman Al Balkhi[1], Yahia Ranjous[1*], Khalil Kashour[1,3], Mohamad Shbat[1,2] and Hussain Chaban[1,2]

Abstract

Background Ewing sarcoma is a malignant round-cell tumor that primarily affects bones in children. It can also arise in extraosseous tissues, such as the lung, kidneys, and liver. The presentation symptoms of Ewing sarcoma may include cough, dyspnea, and chest pain.

Case presentation This report details the history of a 15-year-old Syrian boy with a previous diagnosis of Hodgkin lymphoma who presented with chronic shoulder pain. Imaging studies revealed an 80 mm mass in the apex of the left lung, which was confirmed through histopathological examination to be Ewing sarcoma following a computed-tomography-guided biopsy. The patient received multiple cycles of chemotherapy and subsequently underwent surgical resection of the remaining mass.

Conclusions This case highlights the rare occurrence of Ewing sarcoma in the lung and the unusual clinical presentation of shoulder pain without other accompanying symptoms.

Keywords Ewing sarcoma, Primitive neuroectodermal tumors, Extraosseous, Pulmonary

Background

Ewing sarcoma (ES) is a rare small round-cell tumor originating from the neural crest, accounting for 10–15% of primary bone tumors and ranking as the second most common bone malignancy in children and adolescents [1, 2]. Although ES primarily occurs in bones, it can manifest in various locations such as the lungs, kidneys, and liver [3]. The Ewing sarcoma family of tumors encompasses four different subtypes: Ewing sarcoma of the bone, peripheral primitive neuroectodermal tumor (pPNET), Askin tumor, and extraosseous Ewing sarcoma (EES) [4]. The occurrence rate of EES in pediatric cases with ES is 22%, with the thorax being the most prevalent location [5].

Primary Ewing sarcoma in the lung, first described by Hammar *et al.* in 1989, is a rare presentation for Ewing sarcoma, with around 55 cases reported in the last decade [6, 7]. The clinical manifestations of pulmonary ES can vary widely, often presenting with symptoms such as dyspnea (54.3%), cough (55.3%), chest pain (50%), fever (17%), and hemoptysis (14.9%) [7]. To the best of our knowledge, only one case has reported shoulder pain accompanied by chest pain as a symptom for pulmonary Ewing sarcoma [7, 8].

In this report, we present a case of a 15-year-old male pateint who experienced persistent shoulder pain for 7 months, without any other accompanying symptoms. Subsequent investigations unveiled the presence of an extraskeletal primary Ewing sarcoma in the lung.

*Correspondence:
Yahia Ranjous
yahran2@gmail.com
[1] Faculty of Medicine, Damascus University, Damascus, Syria
[2] Thoracic Surgery Department, Assad University Hospital, Damascus University, Damascus, Syria
[3] Plastic Surgery Department, Al-Mouwasat University Hospital, Damascus University, Damascus, Syria

Case presentation

In January 2023, a 15-year-old Syrian male patient presented with chronic shoulder pain for the past 7 months. The patient's medical history revealed a previous diagnosis of Hodgkin lymphoma at the age of 7 years, for which he received chemotherapy treatment and was monitored for 5 years. Physical examination revealed dullness on percussion and decreased respiratory sounds at the apex of the left lung, without involvement of lymph nodes in the area. Computed tomography (CT) scan revealed the presence of an 80 mm mass at the apex of the left lung. On the basis of the latest information (tumor size 80 mm, intact lymph nodes, absence of metastasis), the tumor was classified as T2N0M0. Subsequently, a CT-guided needle biopsy was conducted on the left lung apex. The histology of the specimen showed necrosis and low metabolic activity in the tumor cells. On immunohistochemical staining, the tumor cells were diffusely positive for CD99 and negative for LCA and DESMIN. On the basis of these findings, the case was diagnosed as Ewing sarcoma/PNET. Chemotherapy treatment was initiated, and the patient received 14 doses, with the last one administered 6 weeks before the surgery.

A follow-up CT scan demonstrated improvement in the size of the mass (measuring 39×37 mm) compared with the previous scan (Fig. 1).

Subsequently, a positron emission tomography (PET)-CT scan revealed an irregular density measuring 30×40 mm at the apex of the left lung with no metabolic activity. Therefore, the tumor was classified as T1N0M0. Consequently, in October 2023, a surgical procedure was performed to remove the mass, which had adhered to the first rib. Therefore, both the first and second ribs had to be excised (Fig. 2). Following the surgical intervention, the patient was transferred to the intensive care unit before being discharged from the hospital and sent home. After 5 months from the surgery, follow-up examinations showed metastasis to the left testicle. Therefore, a second surgery was performed, and the testicle was resected.

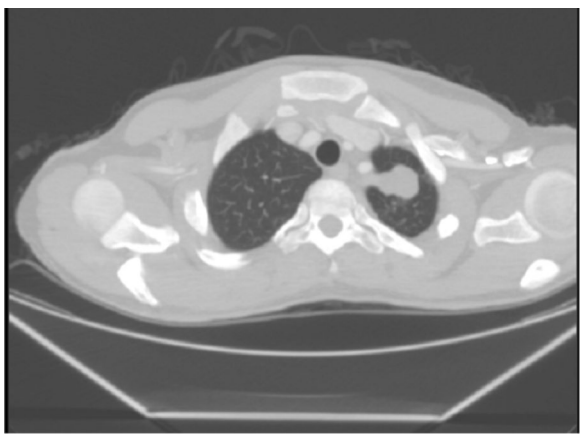

Fig. 1 Thoracic computed tomography, illustrating the reduction in the size of the mass (measuring 39×37 mm)

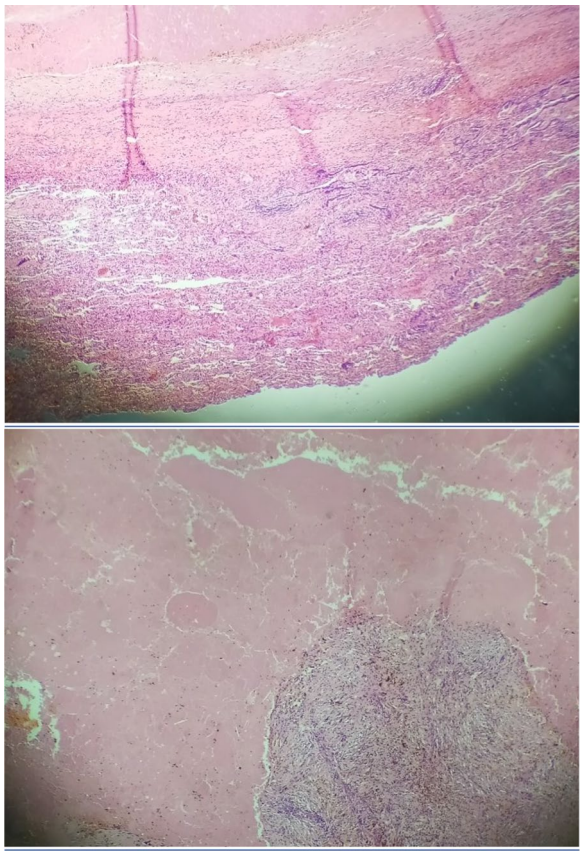

Fig. 2 Microscopic examination of the resected mass, revealing inflammatory infiltrate, granulation tissue, fibrosis with abundant collagen, necrosis, and hemosiderin deposition

Discussion

Ewing sarcoma is part of a tumor family that shares clinical and histological similarities. The two primary members of this family are Ewing sarcoma and peripheral primitive neuroectodermal tumors (PNETs). These tumors can be distinguished by their degree of neurological differentiation [9]. While Ewing sarcoma typically arises from bones, primary pulmonary Ewing sarcoma cases are rare and sparsely documented in medical literature [7]. Diagnosing Ewing sarcoma in the lung can be challenging due to its nonspecific symptoms, such as cough and dyspnea, which resemble those of other common diseases [10].

The use of fluorodeoxyglucose (FDG)-PET/CT has significantly improved tumor detection, including extraosseous Ewing sarcoma. FDG-PET/CT is highly effective in identifying rapidly enlarging masses,

making it an invaluable investigative tool. However, it is essential to emphasize that pathological examination remains the primary diagnostic method, particularly immunohistochemical staining for CD-99 [8]. Pathologically, Ewing sarcoma is characterized by small round blue cells and scant eosinophilic cytoplasm. Immunohistochemical staining reveals a robust positive reaction for glycoprotein p30/32 (CD99). This protein, produced by the MIC2 gene, serves as a reliable marker for diagnosing Ewing sarcoma [11].

Intensive chemotherapy has shown promising outcomes as an adjuvant treatment before surgery, resulting in long-term survival rates of approximately 60–70%. However, recent studies have explored alternative approaches for refractory Ewing sarcomas. Among these options, pazopanib has emerged as a potential treatment when conventional therapies fail to achieve desired results [12, 13].

After confirming the diagnosis, initiating chemotherapy is crucial to reduce tumor size. Subsequently, surgery should be performed to completely remove affected tissue due to the potential risk of tumor relapse [14].

Our case stands out due to the rare location of Ewing sarcoma in the lung and its unique presentation as shoulder pain without other accompanying symptoms. To the best of our knowledge, shoulder pain was only reported once by Ekin *et al.*, and it was accompanied by chest pain, unlike our case [7, 8].

Conclusions

Although Ewing sarcoma/PNET tumors are primarily observed within bones, it is essential not to disregard them as potential differential diagnoses for masses occurring outside the bone, including the lungs. This consideration is particularly significant when assessing children and young adults. A comprehensive understanding of Ewing sarcoma/PNET disease and available diagnostic methods can help radiologists and clinicians accurately diagnose and effectively treat this highly malignant tumor. By remaining vigilant and considering Ewing sarcoma/PNET as a potential diagnosis, healthcare professionals can ensure prompt and appropriate management of patients affected by this condition.

Abbreviations

CT	Computed tomography
PET	Positron emission tomography
PNET	Primitive neuroectodermal tumors
FDG	Fluorodeoxyglucose

Acknowledgements

The authors would like to thank the patient's parents for consenting to publish their son's case.

Author contributions

MB, AAB, YR, KK, MS, and HS took part in writing the manuscript. All authors read and approved the final manuscript.

Funding

The authors did not receive funding for this paper.

Availability of data and materials

The laboratory tests and imaging results are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Received: 5 February 2024 Accepted: 25 July 2024
Published online: 31 August 2024

References

1. Gurney JG, Severson RK, Davis S, Robison LL. Incidence of cancer in children in the United States. Sex-, race-, and 1-year age-specific rates by histologic type. Cancer. 1995;75(8):2186–95.
2. Tu J, Huo Z, Gingold J, Zhao R, Shen J, Lee DF. The histogenesis of Ewing sarcoma. Cancer Rep Rev. 2017. https://doi.org/10.15761/CRR.1000111.
3. Yue B, Chen P, Yin P, Wang J, Liu F, Zhao D, *et al*. Successful radical pneumonectomy for a primitive neuroendodermal tumor in the lung: a case report and review of the literature. Front Surg. 2021;28(8): 667467.
4. Abboud A, Masrouha K, Saliba M, Haidar R, Saab R, Khoury N, *et al*. Extraskeletal Ewing sarcoma: diagnosis, management and prognosis. Oncol Lett. 2021;21(5):354.
5. Ghandour M, Lehner B, Klotz M, Geisbüsch A, Bollmann J, Renkawitz T, *et al*. Extraosseous Ewing sarcoma in children: a systematic review and meta-analysis of clinicodemographic characteristics. Children (Basel). 2022;9(12):1859.
6. Hammar S, Bockus D, Remington F, Cooper L. The unusual spectrum of neuroendocrine lung neoplasms. Ultrastruct Pathol. 1989;13(5–6):515–60.
7. Fedeli MA, Marras V, Fara AM, Deiana A, Lobrano R, Cossu A, *et al*. Primary Ewing sarcoma of the lung: a systematic review of the recent literature. Ann Diagn Pathol. 2023;65: 152152.
8. Ekin S, Çobanoğlu U, Göya C, Erten R, Yıldız H. Ewing's sarcoma of the lung: a rare case. Tuberk Toraks. 2019;67(2):146–8.
9. Lahl M, Fisher VL, Laschinger K. Ewing's sarcoma family of tumors: an overview from diagnosis to survivorship. Clin J Oncol Nurs. 2008;12(1):89–97.
10. Anne S, Malempati AR, Palanki G. Primary Ewing's sarcoma of the lung: a rare case report. Indian J Thorac Cardiovasc Surg. 2020;36(4):409–11.
11. Öztürk S, KurtulusOzturk E, IsiksalanOzbulbul N, Acu B, Dundar E. Primary extraosseous Ewing's sarcoma of the lung: radiologic and pathologic correlation. Cureus. 2021;13(5):e14830.
12. Takigami A, Yamasawa H, Kurosaki A, Sakamoto N, Onuki T, Mato N, *et al*. Pazopanib confers a progression-free survival in a patient with Ewing's sarcoma/primitive neuroectodermal tumor of the lung. Intern Med. 2019;58(9):1335–9.

13. Aggerholm-Pedersen N, Rossen P, Rose H, Safwat A. Pazopanib in the treatment of bone sarcomas: clinical experience. Transl Oncol. 2019;13(2):295–9.
14. 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;26(13):1074378.

Publisher's Note