

Case Report

Extraskelletal Ewing's Sarcoma Presenting as a Posterior Mediastinal Mass[☆]

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ABSTRACT

The Ewing's sarcoma family of tumors is an uncommon group of malignant neoplasms that may be located in both skeletal and extraskelletal regions. Extraskelletal Ewing's sarcoma (EES) is quite rare and predominantly involves the soft tissues of the trunk or the extremities. Herein, we report the case of a 19-year-old female patient who complained of left arm pain. Simple chest radiography revealed an opacity occupying almost the entire left hemithorax. Diagnostic imaging techniques demonstrated a solid contrast-enhanced mass in the posterior mediastinum. There was an evident mediastinal shift, and the left lung was collapsed. Even though lymphoma was considered as an initial diagnosis, a biopsy was taken and its histopathological analysis revealed EES. In the literature, there have been only a few case reports of ESS located in the mediastinum. We conclude that, although this is an unusual location, EES should be contemplated in the differential diagnosis of mediastinal masses.

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Presentación de un sarcoma extraóseo de Ewing como una masa mediastínica posterior

RESUMEN

La familia de tumores del sarcoma de Ewing es un grupo poco habitual de neoplasias malignas que pueden localizarse en regiones tanto óseas como extraóseas. El sarcoma de Ewing extraóseo (SEE) es poco frecuente y afecta de modo predominante a los tejidos blandos del tronco o de las extremidades. Describimos a una paciente de 19 años de edad que refirió dolor en el brazo izquierdo. La radiografía de tórax simple reveló una opacidad que ocupaba casi todo el hemitórax izquierdo y, tras realizar modalidades de diagnóstico por imagen, se demostró una lesión de masa realizada para el contraste, sólida, en el mediastino posterior. Era evidente una desviación mediastínica y el pulmón izquierdo estaba colapsado. Aunque, como diagnóstico inicial, se consideró un linfoma, la paciente se sometió a una biopsia y el análisis histopatológico reveló un SEE. Entre los estudios publicados, solo se han descrito unos pocos casos de SEE localizados en el mediastino. Concluimos que, aunque es una localización insólita del SEE, debe tenerse en cuenta en el diagnóstico diferencial de las masas mediastínicas.

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Palabras clave:

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Mediastino

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Introduction

In the family of primitive neuroectodermal tumors, Ewing's sarcoma stands out as part of a rare group of non-hereditary malignant tumors with a morphology of small-sized, round, blue cells.¹ These tumors generally originate in bone tissue, but they can occasionally originate in soft tissue, known as extraskelletal Ewing's sarcoma (EES). EES is predominantly observed in adolescents and young

adults between the ages of 10 and 30, and it is characterized by an aggressive course and a high associated relapse rate.² It is an uncommon process that especially affects the soft tissue of the extremities or torso. In this article, we described a case of EES that presented as a posterior mediastinal mass in a 19-year-old woman.

Case Report

A 19-year-old patient was admitted to our hospital for pain in the left arm. She presented no other symptoms. Both the physical examination and anamnesis were anodyne. Plain chest radiography revealed an extensive opacity that occupied

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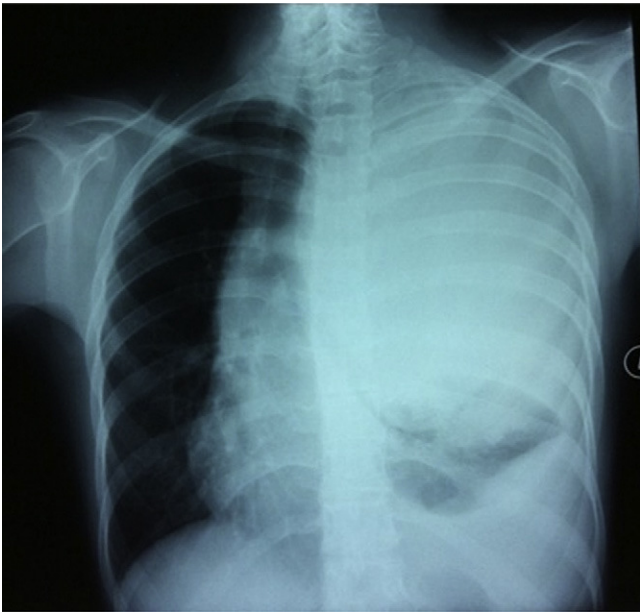


Fig. 1. Plain chest radiography (posteroanterior projection) showing a large opacity that occupies the entire left hemithorax.

almost the entire left hemithorax and caused a deviation of the trachea and the mediastinum towards the right (Fig. 1). Afterwards, computed tomography (CT) exploration better defined this lesion and revealed a solid heterogeneous mass that measured 16 cm×15 cm×15 cm (Fig. 2). The mass extended to the left supraclavicular region, with no skeletal destruction. It was characterized by smooth edges and it demonstrated a direct relationship with the mediastinal vascular structures, without obliterating them. The left lung was completely collapsed. After CT, the patient underwent magnetic resonance imaging (MRI) for more detailed evaluation. The mass originated at the posterior mediastinum, where it manifested low signal intensity in T1 and high signal intensity that was heterogeneous in the weighted MR images in T2, and, after the administration of gadolinium (0,1 mmol/kg), an intense contrast was demonstrated. We considered that the lesion may possibly have been a lymphoma or, although less likely, a primary lung carcinoma as an initial diagnosis. Positron emission tomography–computed tomography (PET-CT) was ordered, which revealed increased fluorodeoxyglucose (^{18}F FDG) uptake by the mass. For the definitive diagnosis, a biopsy was taken by CT-guided transthoracic needle aspiration. The histopathologic exam revealed layers

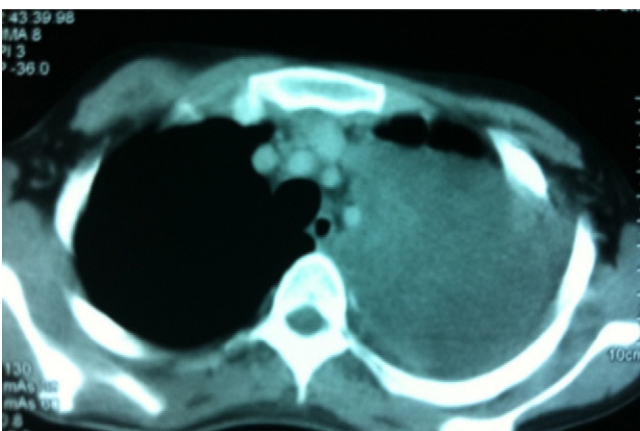


Fig. 2. Axial chest CT with contrast medium demonstrating a hypodense mass in the left hemithorax contiguous to the mediastinal vessels.

of small, round, blue cells with small round nuclei and limited cytoplasm. Immunohistochemistry demonstrated positive staining for CD99 and vimentin. It was negative for all lymphoma markers, including CD31, CD34, CD45, desmin and cytokeratins. Given these findings, the mass was diagnosed as an EES. Immediately afterwards, we consulted with the thoracic surgery department and an intervention was programmed to resect the lesion.

Discussion

EES is part of the primitive neuroectodermal family of tumors, a group of tumors made up of small round blue cells that are not hereditary and affect bone and soft tissue. They are characterized by the presence of translocation (11; 22) (q24; q12).³ Its incidence is predominantly in males, and the male:female ratio is 1,5:1.⁴ EES is a rare disease that affects above all the soft tissue of the extremities, torso, paravertebral and intercostal regions, head and neck, pelvis and peritoneum. Other exceptional locations of these tumors have also been described.⁵ The mediastinal location of EES is very rare. In a study done by Ahmad et al.⁶ including 24 patients with EES, posterior mediastinal mass was only identified in one.

EES can demonstrate clinical findings and imaging technique results that are very variable and, consequently, its diagnosis is based on histopathology. Tumors are frequently observed as slightly hypodense lesions that contain cystic areas on CT without contrast medium, and they demonstrate a heterogeneous pattern on CT with contrast medium.⁷ EES shares the histopathologic and immunohistochemistry findings with Ewing's sarcoma and, therefore, can be confused with embryonal neuroblastoma, lymphoma or rhabdomyosarcoma. Due to this reason, confirmation of the diagnosis should be based on positive staining for CD99 during immunohistochemistry.⁸

EES is a potentially curable disease. Nevertheless, in some cases surgery is required to resect the tumor with wide tumor-free margins together with chemotherapy based on multiple antineoplastic drugs and, in some cases, with radiotherapy in order to obtain favorable clinical outcomes. Therefore, the disease requires a multimodal, aggressive therapeutic strategy. In a retrospective study of 24 patients with EES, a global 5-year survival rate of 61% was seen.⁶ Another study estimated a 5-year disease-free survival rate of 60%-70% for localized disease when treated with chemotherapy based on multiple antineoplastic drugs and surgical exeresis.⁹

In conclusion, in children and in young adults who present with a mediastinal mass, even though EES is uncommon, it is always necessary to consider EES in the differential diagnosis. The reason is that early diagnosis followed by aggressive treatment with surgery and chemotherapy, with or without radiotherapy, produces prolonged survivals even in patients who present extensive disease.

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