

Giant Cystic Mediastinal Lymphangioma

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ABSTRACT

Cystic lymphangioma mainly occurs in children and develops from the lymphatic vessels. The present study reports the case of a 36-year-old female patient with a giant cystic mediastinal lymphangioma. She was asymptomatic and the diagnosis was accidental. Imaging studies are important for referral to the Cardio-Thoracic Surgery Department for surgical intervention in order to remove the lesion. It is important that in the case of an enlargement of the mediastinum observed in a thoracic X-ray, the possible diagnosis takes into consideration various hypotheses. Early diagnosis may prevent further growth and infiltration of structures that hinder the surgical approach.

LEARNING POINTS

- Cystic lymphangioma corresponds to approximately 0.7 to 4% of all mediastinal tumours.
- They are usually asymptomatic and, in adults, the diagnosis is accidental.
- After an imaging study with computed tomography and nuclear magnetic resonance, patients are usually candidates for surgical excision.

KEYWORDS

Cystic lymphangioma, mediastinal tumour, surgical approach

INTRODUCTION

Cystic lymphangioma, also known as cystic hygroma, is a congenital malformation originating from lymphatic hyperplasia and is one of several mediastinal pathologies, corresponding to approximately 0.7 to 4% of all mediastinal tumours^[1]. Its incidence is rare and the majority of lymphangiomas are observed in patients under the age of 5, with extremely few cases being reported in adulthood^[1-3]. The differential diagnosis is made with all mediastinal cystic structures, mainly teratomas, thymic cysts, pericardial and pulmonary cysts^[1,2,4,5].

They are usually asymptomatic and, in adults, the diagnosis is accidental. In children, the compressive symptoms are more frequent due to the lower available space, with frequent alterations of the cervical anatomy^[1]. When present, the most common symptoms are dyspnoea, respiratory insufficiency, hoarseness and superior vena cava syndrome^[2]. In most situations, the diagnosis is made from an enlargement of the mediastinum detected on a chest X-ray performed routinely^[1,2].

CASE DESCRIPTION

A previously healthy 36-year-old female patient resorted to the emergency department due to a condition characterized by a feeling of widespread weakness after breakfast accompanied by vomiting. She denied having other symptoms: chest pain, dyspnoea, cough or fever. The physical examination did not show any relevant alterations and, analytically, only leucocytosis of 13,700/ μ l and neutrophilia of 12,400/ μ l were evident. Her medical history included the removal of an ovarian teratoma at 18 years of age. She remained in the emergency



department under surveillance, with no new episodes of vomiting and/or nausea taking place. After an X-ray, it was evident that there was an enlargement of the medial mediastinum, probably related to a mediastinal mass (Fig. 1).

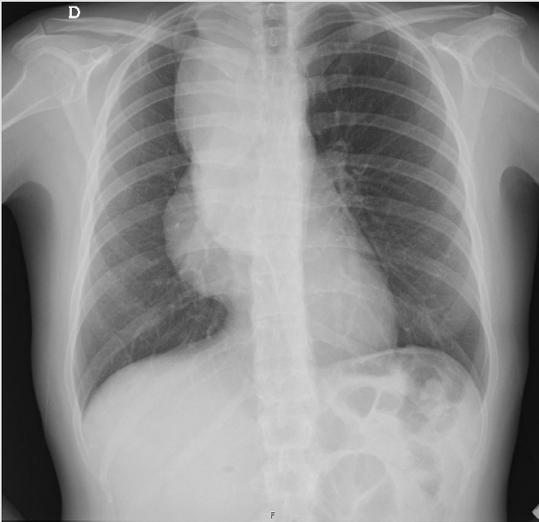


Figure 1. Enlargement of the medial mediastinum in the thoracic X-ray

A computed tomography scan was requested to exclude a vascular cause, with a voluminous formation of liquid content in the mediastinum and the subcarinal region being evident, possibly revealing a pleuropericardial cyst/cystic lymphangioma (Figs. 2 and 3).



Figure 2. Thoracic CT scan with a voluminous formation of liquid in the mediastinum and the subcarinal region



Figure 3. Thoracic CT scan with a voluminous formation of liquid in the mediastinum and the subcarinal region

The patient was admitted to the Internal Medicine Department for complementary studies, having performed respiratory function tests with CO₂ diffusion and plethysmography that revealed a normal respiratory function. A chest MRI scan was requested that confirmed an extensive lobulated, multilocular, cystic lesion centered on the right side of the mediastinum and mediastinal pleura, approximately 200 mm long, 130 mm large from side to side and 140 mm in the anteroposterior dimension. The lesion conditioned the deviation of the vascular structures and the mediastinum, without invasion criteria, suggesting a diagnosis of thoracic cystic lymphangioma, with cervical extension. The patient was referred to the Cardio-Thoracic Surgery Department for surgical intervention in order to remove the lesion.

DISCUSSION

Cystic mediastinal lymphangioma (CML) is an extremely rare vascular tumour originating from the lymphatic vessels. In total, 90% are discovered in individuals under 2 years of age^[6]. After an imaging study with computed tomography and nuclear magnetic resonance, patients are usually candidates for surgical excision. The growth and infiltration of mediastinal structures make surgery difficult. Although other treatment methods, such as sclerotherapy and radiotherapy, have been reported in unresectable cases, they are generally ineffective and may result in haemorrhage and infection^[7]. Therefore, surgery remains the superior method for treatment with curative intent. Complete surgical resection remains the treatment of choice for lymphangioma in order to eliminate symptoms and prevent recurrences^[8]. Thus, it is important to study mediastinal enlargement seen in X-rays, considering the various hypotheses of possible diagnosis, and to investigate with complementary examinations, even when there is no associated symptomatology. Early diagnosis may prevent further growth and infiltration of structures that hinder the surgical approach. In summary, CML, particularly giant CML, is extremely rare in adults. Complete surgical resection provides a definitive histological diagnosis and prevents recurrence.

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