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Primary epithelioid leiomyosarcoma of the endothoracic fascia

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Abstract

The case we describe is, to our knowledge, the first primary epithelioid leiomyosarcoma of the endothoracic fascia reported in the literature. The endothoracic fascia is localized between the costal parietal pleura and the inner face of the thoracic wall and contains connective, adipose, smooth muscular tissues, lymphoid aggregates and vessels. Malignant tumors of the endothoracic fascia are difficult to identify because they quickly infiltrate the lung, the chest wall and the mediastinum. When discovered, these tumors are classified as primary soft tissue sarcomas of these sites. In the present case, early diagnosis of an asymptomatic lesion allowed us to establish its origin in the endothoracic fascia. The CT characteristics suggested a non-aggressive lesion, with clearly defined profiles and without signs of infiltration of the adjacent lung, fat and cartilages. Histological differential diagnosis and role of radiology in the management of the case are discussed. As radiological techniques spread and improve, we expect the diagnosis of endothoracic fascia primary tumors could become more frequent.

Keywords: Endothoracic fascia; Leyomiosarcoma; Epithelioid leyomiosarcoma

1. Introduction

In the text of *Terminologia Anatomica* [1] the fasciae of the trunk are named as parietal, extraserosal and visceral. Fascia parietalis is defined as a "generic term of the fascia, which lies outside the parietal layer of a serosa (pericardium, peritoneum, pleura, tunica vaginalis) and lines the wall of the cavity bodies. The parietal fascia of the thorax is called endothoracic fascia. It covers the internal surface of the sternum, the internal intercostal, subcostal and transverse thoracic muscles, and the diaphragm. Dorsally, it merges with the thoracic portion of the prevertebral fascia. Superiorly, it is continuous with the scalene or Sibson's fascia, which forms the suspensory ligament of the pleura. Inferiorly, the endothoracic fascia is continuous with the transverse (endoabdominal) fascia, which lines the entire wall of abdomen" [2] (Fig. 1). So, endothoracic fascia is by definition equivalent to the parietal abdominal or endoabdominal fascia [3]. The structure of the endothoracic fascia is composed of connective, adipose, smooth muscular tissues, lymphoid aggregates and vessels. During the dissection, the endothoracic fascia is found firmly attached to the costal pleura, and during operation there is no surgical cleavage plane between them. The visceral pleura has an interconnection with the transverse thoracic muscle, the inner thoracic vessels, ribs, intercostal and infracostal muscles, the initial tract of the intercostal nerves and vessels, the sympatethic trunk, the azygos vein and the hemiazygos vein. At these anatomical sites, the endothoracic fascia can give rise to benignant or malignant neoplasm originating from its components. To our knowledge, a primary malignant or benignant neoplasm arising from the endothoracic fascia has never been reported before in any scientific magazine, nor we have found an international tumor classification of benign, malignant primary or secondary tumors of the endothoracic fascia in any pathologic anatomy book.

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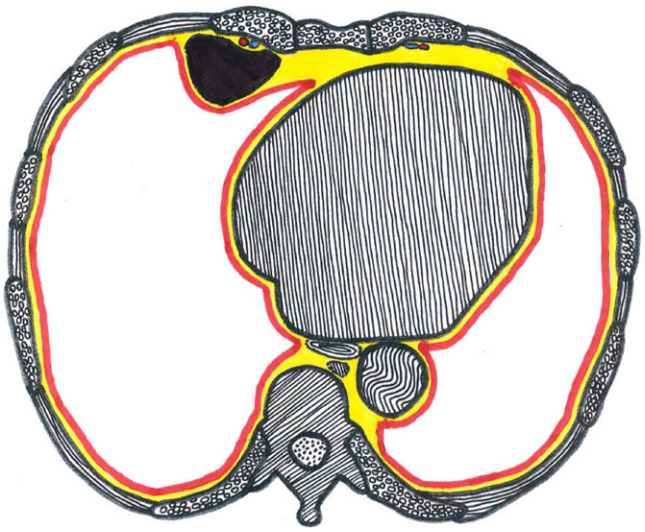

Fig. 1. Endothoracic fascia and epithelioid leiomyosarcoma. In this drawing, adapted from the CT scan, it is represented the pleura and endothoracic fascia (in yellow) and its connections to anatomical site (in gray).

It is very difficult and rare to achieve an early diagnosis of a malignant lesion of the endothoracic fascia without involvement of adjacent anatomical structures. Such tumors are occasionally identified in patients who undergo radiological examinations for other reasons [4]. Malignant tumors of the endothoracic fascia typically grow within a very restricted anatomical space, quickly involving the chest wall or the lung tissue. For this reason, primary tumors of the endothoracic fascia are often included in the category of neoplasms of the chest wall, mediastinum and lung [4–6]. We describe here a case of primary malignant neoplasm of the endothoracic fascia (epithelioid leiomyosarcoma) with emphasis on the radiological and histological features of the lesion.

2. Clinical history

In July 2005 the incidental finding of a thoracic lesion was reported in a 76-year-old woman during a routine chest radiological examination. A subsequent computerized tomography (CT) revealed that the lesion was not localized in the lung but in the adjacent chest wall. The frozen section showed a mesenchimal malignant tumor. Since the neoplasia was well circumscribed, easily enucleated, and did not infiltrate the adjacent structures, a resection of the mass was performed. At this time the patient is healthy, disease-free, with no sign of relapse or recurrence.

3. Radiological findings

The lesion was localized in the extrapleuric antero-medial region of the right hemithorax, behind the internal mammal vessels, close to the costo-mediastinal sinus. The tumour was a 4 cm wide solid lesion, polilobulated, homo-

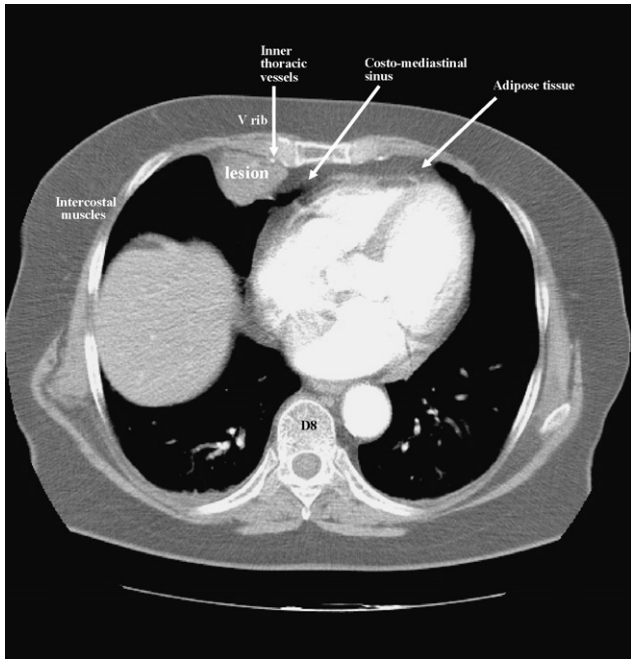

Fig. 2. Leiomyosarcoma of the endothoracic fascia. Contrast-enhanced CT shows a mass with density characteristics indicating a soft bulky tissue. This mass is located at the level of the extrapleuric antero-medial region of the right hemithorax, behind the internal mammal vessels, close to the costo-mediastinal sinus. Notice the position of the lesion in relation to the costo-mediastinal sinus.

geneously hypodense, without central necrotic areas, cystic degeneration or calcifications. We obtained a slight contrast enhancement after administering a contrast agent. The lesion lifted the parietal pleura (the so-called "lifted tent" effect) and compressed the intercostal muscles without signs of invasion of the lung parenchyma. There was no radiological evidence of infiltration of other structures within the thoracic wall (Fig. 2).

From the radiological characteristics, we characterized the lesion as being non-aggressive, with sharp profiles and no sign of infiltration of adjacent structures (lung, fat and cartilages).

A magnetic resonance imaging (MRI) was not performed.

4. Macroscopic findings

The specimen consisted of a soft pink 25-g pseudocapsulated nodule, with a maximum diameter of 4 cm. The cut surface showed a white-gray plurinodular structure with some gelatinous areas.

5. Histological findings

Microscopically, the tumor was confined between the endothoracic fascia and the other anatomical structures of the chest wall, and was composed of a variety of prolifer-