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Thymic cyst as a differencial diagnosis of acute thoracic aortic disease

Cisto tímico como diagnóstico diferencial de doença aguda da aorta torácica

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Abstract

Patient was referred to our Service with acute thoracic pain and diagnosis of intramural hematoma of the ascending aorta for surgical correction. The diagnostic investigation showed a cystic tumor involving the ascending aorta causing restriction of the right ventricular inflow. After resection, the histologic analysis disclosed the diagnosis of thymic cyst. The aim of this study is to describe a rare mediastinal tumor that may simulate a radiologic feature with characteristics of intramural hematoma and may result in a wrong therapeutic approach.

Descriptors: **Mediastinal Cyst. Thymus neoplams. Diagnosis, differential.**

Resumo

Paciente encaminhada de outro serviço com história de dor torácica aguda de forte intensidade com diagnóstico de hematoma intramural na aorta ascendente para correção cirúrgica. Após investigação diagnóstica, identificou-se tumoração cística no mediastino anterior, que envolvia toda a aorta ascendente e que produzia restrição ao enchimento diastólico do ventrículo direito. A análise histológica do tumor ressecado revelou o diagnóstico de cisto tímico. Objetivo deste relato é descrever tumoração mediastinal de baixa prevalência, que pode produzir imagem radiológica com características de hematoma intramural e resultar em conduta terapêutica equivocada.

Descritores: **Cisto mediastínico. Neoplasias do timo. Diagnóstico diferencial.**

INTRODUCTION

Primary tumors and mediastinal cysts are uncommon, affecting patients of all ages (though they are more frequent in young and middle-aged adults), and typically identified during routine examinations. The benign lesions are usually asymptomatic, while the malignant lesions are associated with chest pain, pleural effusion, diaphragmatic paralysis, weight loss and/or fever. The diagnosis is confirmed by biopsy and histological analysis, but the clinical suspicion of a specific lesion can be based on its location, the patient's age, the presence or absence of symptoms and the association with systemic diseases. Most lesions of the anterior mediastinum (structures committed between the sternum and pericardium) are of thymic origin, but there are recent reports in the literature about thymic lesions (thymoma and thymic cyst) located in the middle mediastinum. In the case of tumors of the anterior mediastinum, the thymic cyst occupies the ninth position, being preceded by the following neoplasms: thymoma, germ cell tumor, lymphoma, lymphangioma, hemangioma, lipoma, fibroma and fibrosarcoma [1-3].

Thymic cysts are uncommon mediastinal tumors corresponding to 1% to 3% of the lesions in the mediastinum [4]. These cysts are rarely symptomatic, however, due to their mass effect, which is associated with

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This study was carried out at Heart Institute (InCor) of University of São Paulo Medical School.

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symptoms of dyspnea, cough or chest pain. Tomography and chest resonance are the diagnostic methods used to provide accurate information about the characteristics of mediastinal tumor masses.

CASE REPORT

79-year-old female patient, referred to the Emergency Unit with presentation of severe chest pain and dyspnea that began 15 days prior, with diagnosis of Stanford type A aortic dissection (intramural hematoma) after a chest tomography at the local hospital. The patient presented with underlying diseases: systemic arterial hypertension and ischemic cardiomyopathy with a left ventricular ejection fraction of 35% (two previous infarctions and two stent angioplasties in the anterior interventricular artery).

During the diagnostic investigation at our hospital, there were no changes in physical examination and the ECG revealed inactive area in the anterior wall, normal cardiac enzymes and enlargement of mediastinum. In addition to septal and anterior akinesia, the transthoracic echocardiogram revealed mild pericardial effusion with signs of restriction to diastolic filling of the heart chambers on the right side and ascending aorta within normal limits. The chest angiotomography revealed an ascending aorta with maximum caliber of 38 mm without signs of dissection, large pericardial effusion and an anterior mediastinal cystic measuring 11.5cm x 6.8cm x 9.0cm involving the ascending aorta, with an image compatible with a pericardial cyst (Fig.1a and 1b). The patient underwent surgical resection of the tumor via longitudinal sternotomy with identification of unilocular cystic lesion measuring approximately 13cm x 7cm and completely covering the ascending aorta (Fig. 2a). The histological analysis revealed that these areas were thymic tissue areas mixed with lymphoid tissue (Fig. 2b). The patient recovered well and remained asymptomatic, with no recurrence of the tumor six months after the procedure.

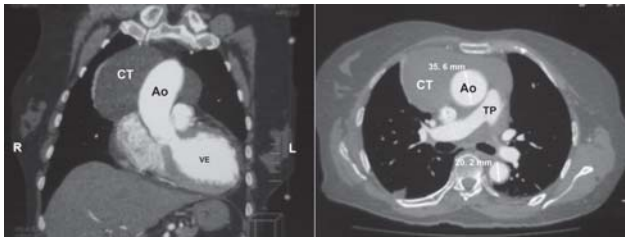

Fig. 1 ***Thoracic aorta angiotomography*** *– CT chest image in sagittal position (at left) and in axial position (at right) showing cystic formation encircling the ascending aorta.*
Ao: Aorta; CT: Tymic Cyst; VE: Left Ventricle; TP: Pulmonary Trunk

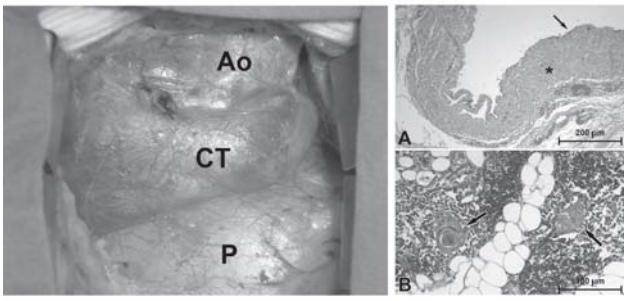

Fig.2 – ***Thymic Cyst*** *– 2a – Intra-operative image of the tymic cyst after longitudinal sternotomy. 2b – Hystological image of the tymic cyst, characterized by: A. Fibrosis (asterisk) partly covered by flat epithelial cells (arrow); B. Tymic cysts residues, characterized by presence of Hassall's corpuscles (arrow) admixed in lymphoid tissue. (Hematoxylin-eosin; increase of 200ì;100ì).*
Ao: Aorta; CT: Tymic Cyst; P: Pericardium

DISCUSSION

Thymic cysts represent about 1% of all mediastinal masses. When only the cystic thymic lesions are analyzed, the incidents range from 12% to 30% [5]. They are more frequently located in the anterior mediastinum, and can also be found in the neck, depending on thymus development.

Thymic cysts can be unilocular or multilocular. The unilocular cysts are probably of congenital origin (derived from embryologic thymic tissue), with a thin fibrous capsule composed of cuboid or columnar, squamous or transitional epithelium. In most cases, there is a clear serous fluid inside this ephithelium, which can also be thick, blood or heterogeneous, and its wall could present traces of cholesterol, signs of chronic inflammation, hemorrhage or calcification [6]. The most important feature, which allows for the definitive diagnosis of the lesion, is the presence of thymic tissue in the cyst wall characterized by a corticomedullary differentiation, where the Hassall's corpuscles can be found in about 50% of cases. The multilocular cysts are usually acquired lesions and commonly result from infection, trauma or neoplasia, as occurrs in Hodgkin's disease or in association with other diseases such as the Human Immune Deficiency Syndrome and auto-immune diseases such as Sjogrën's Syndrome.

The symptoms vary according to the tumor's size and location. The tumor can cause chest pain, dyspnea, cough, dysphagia and symptoms of low cardiac output [7]. They are usually found between the 3rd and 6th decade of life, usually through routine radiological examinations or cervical or thoracic surgeries. The characteristics of such tumors under chest tomography are similar to a lesion with

low-attenuation homogeneous structure, usually between 0 and 20 Hounsfield units. It is worth noting that, when performed by non-experienced professionals, the radiological analysis of thymic cyst images - completely encircling the ascending aorta - can result in an incorrect diagnosis and, as a result, errors in the therapeutic approach.

The treatment of unilocular thymic cysts is controversial. Some authors believe that resection is necessary for diagnosis because it deals with a histological diagnosis. Reports of malignant transformation and incipient neoplasia are also indications for surgery [8]. Multilocular cysts should also be removed so that any malignant degeneration is identified. Surgical resection is usually curative and can be performed safely and efficiently through videothoracoscopy, longitudinal sternotomy or thoracotomy.

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