CAZURI CLINICE

A rare case of unicentric plasma cell type Castleman's disease in the mediastinum

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REZUMAT

Caz rar de boală Castleman unicentrică de tip plasmocitar la nivel mediastinal

Boala Castleman este o afecțiune limfoproliferativă foarte rară (așadar puțin cunoscută), caracterizată prin hiperplazie ganglionară foliculară cu proliferare anormală vasculară interfoliculară. Poate fi clasificată în boală unicentrică sau multicentrică, în funcție de localizare, și în trei tipuri histologice: hialin-vascular, plasmocitar și mixt.

Articolul descrie cazul unei paciente de 51 de ani care s-a prezentat cu durere toracică precordială. Radiografia toracică și tomografia computerizată (CT) au ridicat suspiciunea unui anevrism aortic, iar pacienta a fost trimisă la Secția de cardiologie clinică intervențională de la Spitalul Clinic Militar de Urgență, unde s-a stabilit diagnosticul de tumoră medistinală prin ultrasonografie, coronarografie și CT.

Pacienta a fost transferată în clinica noastră, unde mediastinoscopia în scop diagnostic a fost non-diagnostică. S-a practicat ulterior toracotomie laterală cu tumorectomie ideală, prin care s-a stabilit diagnosticul de boală Castleman de tip plasmocitar.

Articolul prezintă mecanismele patogenice, simptomele, diagnosticul și tratamentul bolii Castleman.

Cuvinte-cheie: *boală Castleman, tip plasmocitar, tumoră mediastinală.*

ABSTRACT

Castleman's disease is a very rare (and thus little-known) lymphoproliferative disorder characterized by lymph node follicular hyperplasia with abnormal interfollicular vascular growth. It can be classified into unicentric and multicentric variants according to its localization, and into three histological types: hyaline-vascular, plasma cell and mixed.

We describe the case of a 51 year-old woman who presented with precordial chest pain. Chest X-Ray and computed tomography (CT) raised the suspicion of an aortic aneurysm, and the patient was sent to the Interventional Cardiology Clinic at the Military Emergency Clinical Center for Cardiovascular Diseases (CCUBCVA), where the diagnosis of mediastinal tumor was made by ultrasonography, coronarography and CT. The patient was transferred to our clinic, where a diagnostic mediastinoscopy proved histolopathologically inconclusive. A lateral thoracotomy with ideal tumorectomy was decided upon and carried out, and the diagnosis of plasma cell type Castleman's disease was established.

This paper discusses the pathogenic mechanisms, symptoms, diagnosis and treatment of Castleman's disease.

Key words: *Castleman's disease, plasma cell type, mediastinal tumor.*

Introduction

Castleman's disease is a very rare lymphoproliferative disorder characterized by a follicular hyperplasia of the lymph nodes, with abnormal interfollicular vascular growth. This disease is also known as: angiofollicular lymph node hyperplasia, gigantic lymph node hyperplasia, lymphoid hamartoma, angiomatous lymphoid hyperplasia and gigantic benign lymphoma[1].

Case presentation

We describe the case of a 51 year-old female who presented with precordial chest pain. Chest X-Rays (Fig. 1) revealed an enlargement of the mediastinum and computed tomography raised the suspicion of an ascending aortic aneurysm. The patient was sent to the Interventional Cardiology Clinic at CCUBCVA, where a series of paraclinical tests were carried out: ECG – sinus rhythm without repolarization changes; echocardiography – ascending thoracic aorta 42 mm in diameter, hyperechogenic walls, supple valves, intact septa, normal kinetics and pericardium free of fluid. Coronarography showed permeable epicardic coronary arteries, and left atrial and ascending thoracic aortic ectasia. A new thoracic CT scan showed a large (4.4 x 5.2 x 7 cm) mass, located paratracheally to the right in the superior mediastinum, imprinting on the trachea and displacing it to the left, while also displacing the vena cava anterolaterally and adhering closely to it for 3-4 cm. The tumor was intensely contrast-enhancing and heterogeneous in structure, and presented small calcified

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Figure 1.
Posteroanterior chest x-ray – a right homogeneous paramediastinal opacity

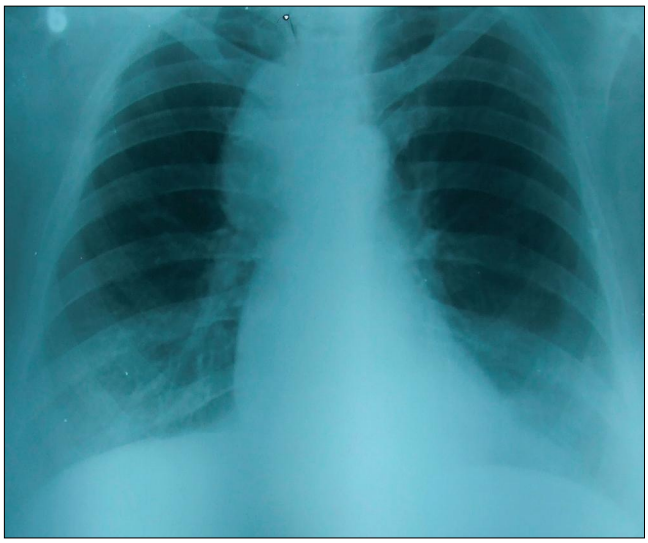

Figure 2.
CT scan – Large contrast-enhancing mediastinal tumor

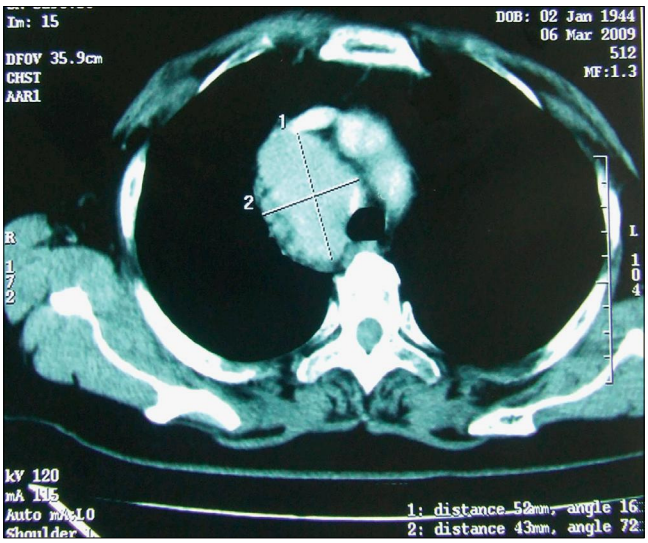

Figure 3.
CT scan – Large mediastinal tumor displacing trachea to the left and vena cava anterolateral