JCEI /
Journal of Clinical and Experimental Investigations
2015; 6 (1): 69-71
doi: 10.5799/ahinjs.01.2015.01.0489

CASE REPORT / OLGU SUNUMU

A rare cause of Ortner's syndrome: giant pulmonary artery aneurysm secondary to Behçet's disease

Ortner sendromu' nun nadir bir nedeni: Behçet hastalığına sekonder gelişen dev pulmoner arter anevrizması

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ABSTRACT

Behçet' s disease is a systemic autoimmune vasculitis of unknown etiology. It causes serious disability by affecting both arteries and veins. Hoarseness due to compression of the left recurrent laringeus nerve resulting from pathologies of the heart and intrathoracic great vessels is defined as Ortner's syndrome. The most common cause of Ortner's syndrome is left atrial enlargement due to mitral stenosis. Various intrathoracic pathologies may also be the reason. Beside, Ortner's syndrome due to primary pulmonary artery aneurysm as a feature of Behçet's disease is relatively rare. Herein, we report a case of a 78 year old female patient presenting with hoarseness and diagnosed as Ortner's syndrome resulting from a giant pulmonary artery aneurysm secondary to Behçet' s disease. *J Clin Exp Invest 2015; 6 (1): 69-71*

Key words: Ortner' s Syndrome, Pulmonary Artery Aneurysm, Behçet's Disease

ÖZET

Behçet Hastalığı nedeni bilinmeyen sistemik bir otoimmün vaskülittir. Hem arterleri hem de venleri tutarak ciddi sorunlara neden olabilir. Kalp ve intratorasik büyük damarların patolojilerine bağlı olarak sol rekürren laringeal sinirin kompresyonu sonucu gelişen ses kısıklığı Ortner Sendromu olarak ifade edilir. Ortner Sendromu' nun en sık nedeni mitral darlığa bağlı olarak sol atriyum genişlemesidir. Çeşitli intratorasik patolojiler bu tablonun nedeni olabilir. Bununla birlikte Behçet Hastalığı sonucu gelişen pulmoner arter anevrizmasına bağlı Ortner Sendromu nispeten nadirdir. Biz burada ses kısıklığı ile başvuran ve yapılan incelemeler sonucunda Behçet Hastalığı' na bağlı dev pulmoner arter anevrizması sonucu gelişen Ortner Sendromu olarak tanı alan 78 yaşında bir bayan hastayı sunuyoruz.

Anahtar kelimeler: Ortner Sendromu, Pulmoner Arter Anevrizması, Behçet Hastalığı

INTRODUCTION

Behçet' s disease is a multiorgan disorder of unknown origin and can cause complications in cardiovascular system, central nervous system and respiratory system resulting from vasculitis of both arteries and veins [1,2]. Pulmonary involvement rate in Behcet's disease is between 5-10 % and is seen most often in the form of pulmonary artery aneurysm (PAA) [3]. Vocal cord paralysis due to extralaryngeal cardiovascular pathologies, such as the thoracic aorta aneurysms and PAA is defined as Ortner syndrome. Left vocal cord paralysis due to PAA is quite rare [4,5]. Herein, we report a case of a 78 year old female patient presenting with hoarseness and diagnosed as Ortner's syndrome resulting from a giant pulmonary artery aneurysm secondary to Behçet' s disease.

CASE PRESENTATION

A 78 year-old female patient suffering from fever, fatigue, recurrent oral ulcers, and progressive hoarse-

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Received: 30.12.2014, Accepted: 15.01.2015

ness in the last 6 months admitted to our clinic. The medical history revealed that she had been following with the diagnosis of Behcet's disease for approximately 10 years and had received corticosteroid treatment previously. But she was not on a regular treatment regimen. Pathergy test for diagnostic evaluation of Behcet's disease was repeated and found to be positive.

On physical examination, arterial blood pressure was 100 / 60 mmHg and radial pulse was regular with a rate of 100 / min. Cardiovascular system examination was normal except tachycardia. In teleradiography, a prominent pulmonary artery was detected (Figure 1). On transthoracic echocardiography, right and left ventricular size and functions were normal, but the main pulmonary artery and its branches were excessively dilated . Doppler echocardiography revealed mild tricuspid and pulmonary regurgitation. Pulmonary artery systolic pressure measured through tricuspid regurgitant flow was 32 mmHg and diastolic and mean pressures of pulmonary artery were detected to be 14 and 18 mmHg respectively. In multislice CT angiography, main pulmonary artery was measured to be 55 mm at the level of the aorticopulmonary window. Left pulmonary artery was 32 mm and right pulmonary artery was 42 mm (Figure 2). The patient underwent endoscopic examination of the larynx and the left vocal cord was found to be fixed in the paramedian position.

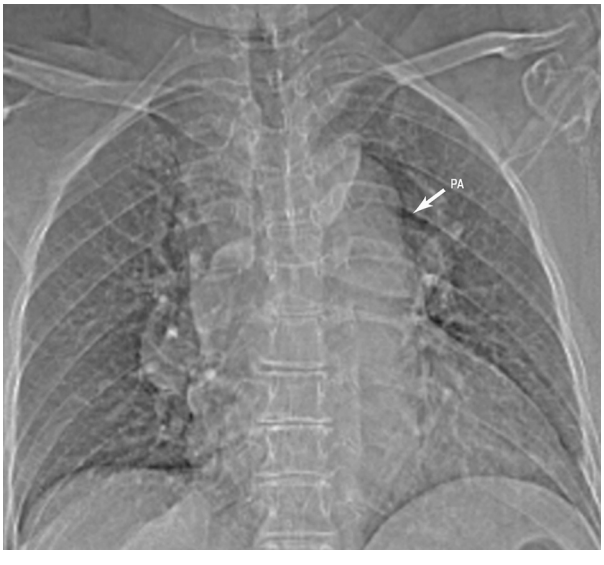

Figure 1. Antero-posterior chest x-ray shows aneurysmal main pulmonary artery and its branches in central and peripheral localizations

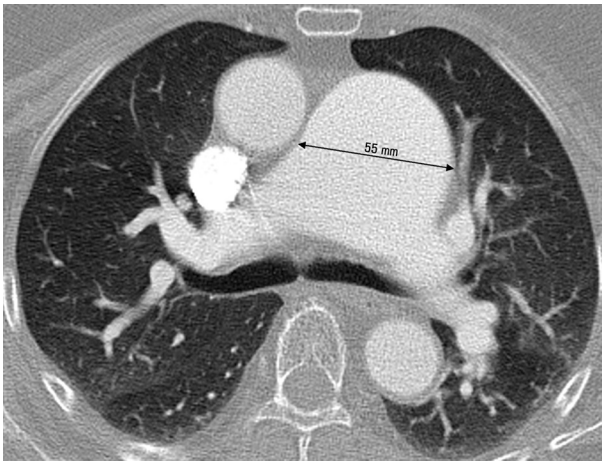

Figure 2. Axial contrast-enhanced CT image shows saccular aneurysm at the level of aorticopulmonary window

Surgery was not considered due to existence of Behçet 's disease, advanced age of the patient and extent of the aneurysm involving both pulmonary artery and its branches. Considering systemic symptoms and oral ulcers, the patient was thought to have an acute attack and medical therapy was started. Remission of fever, fatigue and oral ulcer was achieved with cyclophosphamide and corticosteroid therapy. A mildly improvement in hoarseness was also observed. The patient was discharged with follow up recommendations. The patient maintained asymptomatic on medical follow up.

DISCUSSION

PAA is a quite rare clinical condition [6]. The most common type of vasculitis associated with pulmonary artery aneurysms are Behcet's disease and Hughes-Stovin syndrome [7]. The patients with PAA are usually asymptomatic or may present with nonspecific complaints such as exertional dyspnea, fever, cough and hemoptysis. In some patients, compression of surrounding structures via the enlarged pulmonary artery or stretching of the pulmonary artery itself can cause symptoms. Hoarseness due to compression of the left recurrent laringeus nerve secondary to pathologies of the heart and intrathoracic great vessels is defined as Ortner's syndrome. The most common cause of Ortner's syndrome is left atrial enlargement due to mitral stenosis. Mitral valve prolapse, thoracic aortic aneurysm, aortic dissection, cor pulmonale and pulmonary artery dilatation secondary to increased pulmonary artery pressure with any reason have also been described in etiology of Ortner's syndrome [8,9].

Several etiologies have been described in the pathogenesis of PAA, namely, pulmonary hypertension (PH), congenital heart disease, Behçet' s disease, infections such as the formerly prevalent syphilis, arteriovenous fistulas, connective tissue diseases, atherosclerosis, and trauma [10, 11]. Our patient had none of these etiological factors except Behçet' s disease. Therefore, the point that makes this paper worthy of reporting is development of Ortner's syndrome due to pulmonary artery aneurysm probably resulting from Behçet's disease. Absence of increased pulmonary arterial pressure leads to the diagnosis of pulmonary artery aneurysm in our patient and hoarseness was due to enlargement of pulmonary artery. Beyond these clinical characteristics, the patients with PAA are also at high risk for dissection or rupture which are mortal [12]. Therefore, corrective surgery is recommended, but the risks and long-term results of surgical treatment are not well defined. In our case, while the absence of pulmonary hypertension reduces the risk of aneurysm rupture, the presence of Behçet's disease increases the risk [13]. Remission of the systemic symptoms and improvement in hoarseness with medical therapy led us to avoid surgery in this case. The improvement in hoarseness was probably due to resolution of the edema of the pulmonary arterial wall.

Our report is one of the rare PAA cases secondary to Behçet's disease causing Ortner's syndrome in the literature. Consequently, it should be taken into consideration that hoarseness in a patient with Behcet's disease may develop due to a PAA and further evaluation should be performed. Additionally, although surgery seems to be the effective treatment modality of PAA, in Behçet's disease steroids targeting the primary cause may provide clinical improvements and eliminate the need for surgery.

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