

Giant pulmonary artery aneurysm due to undiagnosed atrial septal defect associated with pulmonary hypertension

Atrial septal defektin sebep olduğu pulmoner hipertansiyon sonucu gelişen dev pulmoner arter anevrizması

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Introduction

Pulmonary artery aneurysm is a rare condition. It may develop as a result of variable etiologies such as congenital heart disease, infections, vasculitis, arteriovenous communications, trauma and connective tissue diseases (1). Due to its rare occurrence, little is known about natural evaluation; therefore there are no suggested guidelines for the optimal treatment as for aortic aneurysms (2). We present a case with a huge pulmonary artery aneurysm (PAA) of the pulmonary conus and bilateral main pulmonary arteries with a distal branch extension. Pulmonary hypertension due to undiagnosed wide atrial septal defect (ASD) at the older age was the etiology of this giant aneurysm.

Case Report

A 55-year-old male with a 9-year history of dyspnea was referred to our hospital for clinical evaluation. He had progressive shortness of breath and reduced exercise capacity. His prior medical evaluation showed pulmonary artery hypertension and large pulmonary artery aneurysm. He has been checked for the Behçet's disease due to large pulmonary artery aneurysm, which is a second common site of arterial involvement (3) but diagnose for Behçet's disease was not established. His physical examination was normal except a palpable systolic impulse within the second left anterior intercostal space and 2/6 systolic murmur at the apex. At his chest radiography there was a prominent bilateral pulmonary artery enlargement (Fig. 1A), which was measured 6 cm from the chest roentgenogram. In order to detect the pulmonary vasculature lesions, magnetic resonance angiography had been performed and it confirmed the 5.5 cm large aneurysm of the main pulmonary artery and bilateral main pulmonary arteries with an extension to the distal branches (Fig. 1B). In our institution he was investigated for the secondary causes of the pulmonary hypertension and pulmonary aneurysm because of their associations with other clinical conditions (2).

Due to easy accessibility of the echocardiography it was performed to rule out any congenital or acquired etiology of pulmonary hypertension. Echocardiography showed 4 cm in size wide prominent secundum type ASD with a hypertrophied right ventricular and right atrial enlargement but normal left ventricle (Fig. 2). In color Doppler examination there was no right to left shunting across the defect due to high right ventricular pressure. At the Doppler echocardiography severe tricuspid and mild mitral regurgitation were obtained. Measured pulmonary artery systolic pressure was 130 mmHg. Right heart catheterization was performed to measure pulmonary pressures precisely. Pulmonary artery pressures could not be obtained due to failure to accent but the right ventricle pressures were measured as 110 mmHg systolic and 5 mmHg diastolic. Atrial septal defect was confirmed by passing through it and oxygen saturations demonstrated a step-up within the right atrium due to shunting across the defect. After confirmation the diagnosis and etiology he was presented to institution's cardiovascular committee. Surgical closure of the ASD was not considered due to established Eisenmenger's syndrome and it was decided to follow-up medical treatment with calcium channel blockers and anticoagulation.

Discussion

Aneurysm of the pulmonary artery is a rare disease with an estimated incidence of about 8 cases in every 100.000 autopsies (1-2). Aneurysm is defined when pulmonary artery dilatation greater than 4 cm (4). Although it may be related with a variety of etiologies approximately 50% of aneurysms are associated with congenital heart diseases mostly due to pulmonary hypertension (2, 5). Other causes are vasculitis especially seen in Behçet's disease, infections (usually in the pre-antibiotic era mostly due to tuberculosis and syphilis), arteriovenous fistulas, connective tissue disorders and trauma (2). The clinical presentation may be entirely asymptomatic or non specific such as chest pain, cough, and shortness of breath may accompany this disorder (1). In our

case dyspnea was related to pulmonary hypertension and partly was a result of bronchial compression due to enlarged aneurysm. Pulmonary artery aneurysms are located most frequently in the right lower lobar arteries followed by the right and left main pulmonary arteries (3). The enlargement of central pulmonary arteries, with attenuation of peripheral vessels, and oligemic peripheral lung fields are classic finding in pulmonary hypertension (6). However, in this case, the diffuse extension of the aneurysm from truncus to distal branches of the bilateral pulmonary artery was prominent in comparison to other cases reported in the literature where pulmonary hypertension usually was related to main pulmonary trunk involvement.



Figure 1A. Postero-anterior chest roentgenogram showing bilateral enlargement of the main pulmonary artery
White arrows point the right interlobar artery aneurysm of 6 cm in size

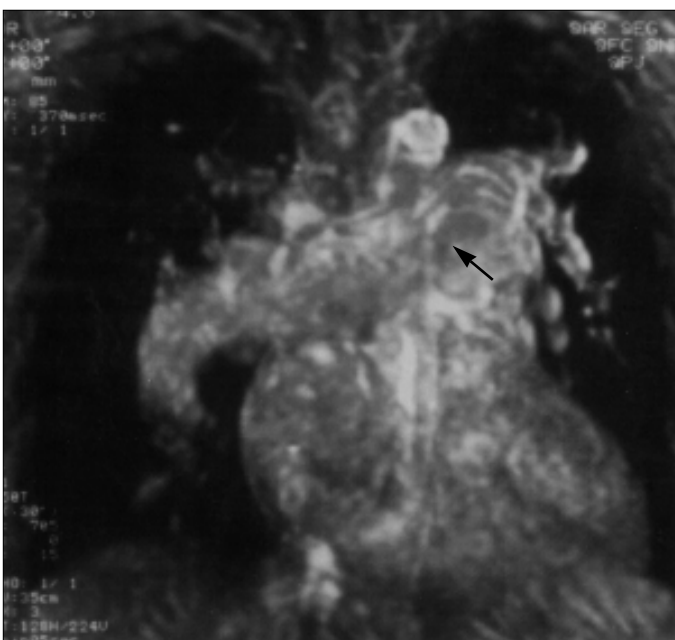


Figure 1B. Magnetic resonance angiography view of the aneurysm of pulmonary truncus and main branches

One of the three pathophysiological mechanisms of the pulmonary hypertension is pressure-volume overload caused by left to right intracardiac shunts, left ventricular dysfunction or valvular disease (7). Absence of valvular disease and normal left ventricular function and diameters in the echocardiography examination excluded the other etiologies that related to heart other than wide ASD (2).

Possible complications such as dissection, embolism, rupture, compression of the surrounding tissues may occur (2). Surprisingly, there were neither dissection nor rupture in the magnetic resonance imaging despite presence of established severe pulmonary hypertension. Being the most life-threatening complications, the risk of dissection is associated with pulmonary hypertension and/or connective tissue diseases while the risk of rupture increases with advanced age and the previous use of steroids (8, 9).

Thrombosis is the common complication in pulmonary hypertension of any etiology and may worsen the patient's functional class which is diminished as a result of pulmonary hypertension (7).

Even though little is known about the natural history of pulmonary artery aneurysm, there is no clear guideline about treatment especially for surgical indications (2). Surgical correction of the ASD was not considered for this patient due to established severe pulmonary hypertension.

In conclusion, we aimed to point out the importance of non specific clinical manifestations of the pulmonary artery aneurysms and pulmonary hypertension. Clinical presentation may be only with non specific shortness of breath however, unexplained dyspnea with or without accompanying pulmonary artery ecstasies at the routine chest roentgenogram are needed to be evaluated for the underlying etiologies causing pulmonary hypertension. Basic electrocardiogram showing right ventricular hypertrophy and atrial enlargement may lead physicians to suspect right ventricular pressure overload. Even though there are no clear guidelines for the surgical management for the pulmonary artery aneurysm, early diagnosis of the dilatation of the artery or the underlying correctable etiology may prevent the disease progression to inoperable stage.

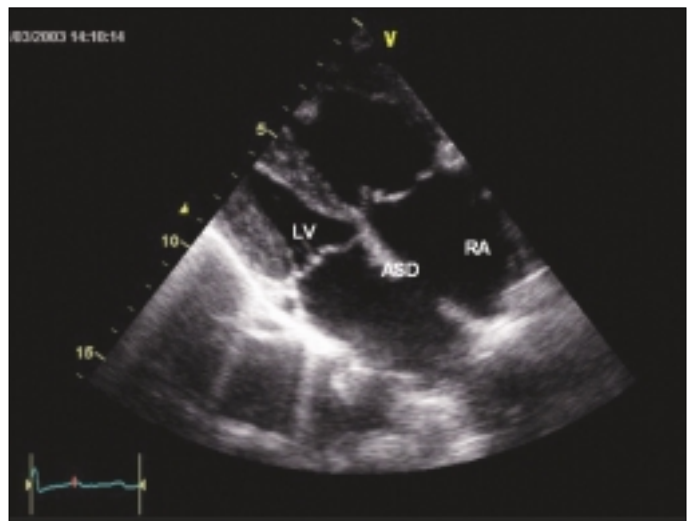


Figure 2. Echocardiographic view of a wide atrial septal defect

ASD- atrial septal defect, LV- left ventricle, RA- right atrium

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