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Idiopathic Asymptomatic Main Pulmonary Artery Aneurysm: Surgery or Conservative Management? A Case Report

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ABSTRACT

Idiopathic main pulmonary artery aneurysm is a very rare entity and there are no clear guidelines for optimal treatment. Operative treatment is recommended for patients with a risk of rupture, which is not well defined. We present an unusual case of a 53-year-old woman with an idiopathic main pulmonary artery aneurysm. Our case is asymptomatic, without a causative cardiac lesion and/or pulmonary hypertension; therefore, we did not operate on our patient and she was stable at 22-month follow-up.

INTRODUCTION

Idiopathic main pulmonary artery aneurysm is a rare anomaly. The cause of pulmonary artery aneurysm and optimal treatment strategies are not clear. Some authors prefer conservative management [Casselman 1997] and others prefer surgery [Finch 1983; Chen 1996; Kiron 2001]. As reported in the literature, operative treatment is commonly recommended, but the localization and size of the aneurysm and the risk of rupture is important for choosing` treatment [Finch 1983]. In 1947, Deterling and Calget reviewed pulmonary artery aneurysms and reported them as either central or peripheral [Deterling 1974]. Pulmonary artery aneurysm is most commonly associated with other congenital cardiovascular lesions, most frequently being patent ductus arteriosus, ventricular septal defect, and atrial septal defect. Other causes and predictors are atherosclerosis, infections (syphilis, tuberculosis, and bacterial endocarditis), cystic medial necrosis, and some types of vasculitis, hypertension, and trauma [Barrater 1998; Ozyazicioglu 2001]. But sometimes there are no causes and even no predictors, and these lesions are called idiopathic pulmonary artery aneurysms. Most reported cases relate to autopsy findings. This entity seldom has been observed in a patient's lifetime [Jaffin 1983].

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CASE REPORT

A 53-year-old woman with an abnormal superior mediastinal silhouette on chest x-ray was referred to our clinic. She had no symptoms whatsoever. Auscultation revealed 1/6 systolo-diastolic murmur over the left sternal border and other physical and neurological examinations were normal. The patient was in sinus rhythm, had no ischemic findings on electrocardiography, and was normotensive. Blood tests were all within normal range. Chest x-ray showed a widening of the mediastinum with no cardiomegaly and normal lung fields. No previous chest roentgenogram was available. A computed tomographic scan of the chest revealed an aneurysm of the main pulmonary artery with a maximum diameter of 6.1 cm. Dilatation of the pulmonary artery started 1 cm above the pulmonary valves and there was no intraluminal thrombus (Figures 1-3). Screening for rheumatic and autoimmune diseases as well as diagnostic evaluation for tuberculosis, syphilis, and Behcet and Marfan syndromes were negative. Echocardiography showed an additional pulmonary valve insufficiency of grade 1/4 and there was no stenosis. Coronary angiography was normal. Right and left cardiac catheterization was performed to confirm the main pulmonary artery aneurysm, but no other cardiac abnormalities or shunts were found (Figure 4). Catheterization documented right atrial pressure of 4 mmHg, right ventricular pressure of 38/6 mmHg, pulmonary arterial pressure of 30/8 mmHg, and pulmonary capillary wedge pressure of 7 mmHg.

DISCUSSION

Idiopathic main pulmonary aneurysm is a rare entity. The etiology and pathogenesis of pulmonary artery aneurysms are not well known. The incidence of pulmonary artery aneurysm is about 8 cases in every 100,000 autopsies [Casselman 1995].

Pulmonary artery aneurysm classification includes primary (congenital) and secondary (acquired) aneurysms. The classification of pulmonary artery aneurysms is shown in Table 1 [Baratter 1998]. Evaluation of a patient with pulmonary artery aneurysm should include echocardiography, computed tomography, cardiac catheterization, and coronary angiography to exclude other cardiac pathologies. Pulmonary

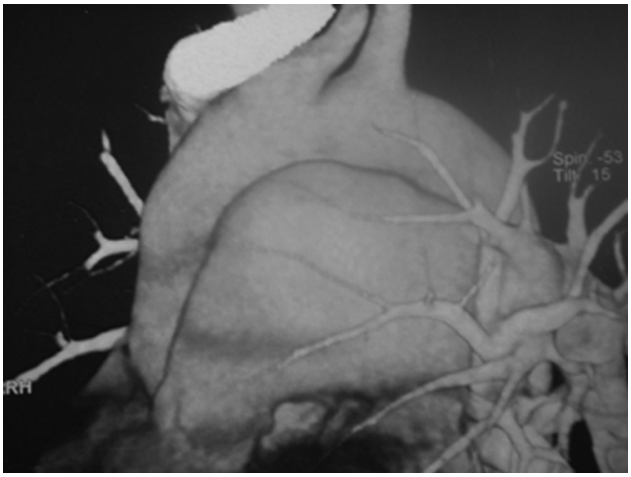

Figure 1. Multislice computed tomography scan shows the main pulmonary artery aneurysm.

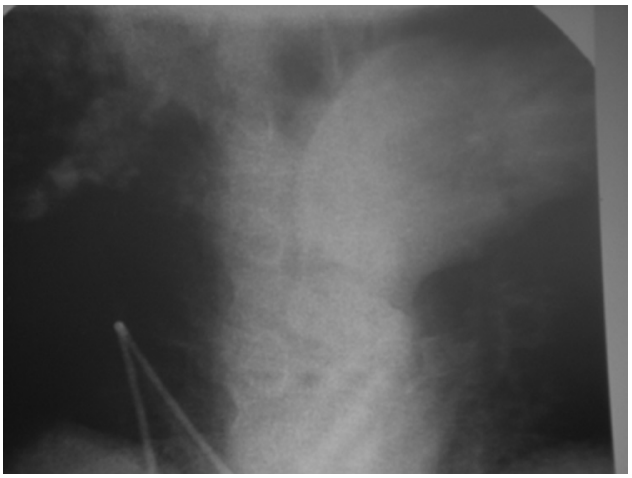

Figure 3. Computed tomography scan shows the main pulmonary artery aneurysm without intraluminal thrombus.

artery pressure, aneurysm size, and pathogenesis are very important for the choice of treatment strategy.

If an aneurysm is associated with (A) simple dilatation of the pulmonary trunk, with or without involvement of the rest of the arterial tree; (B) absence of abnormal intracardiac or extracardiac shunts; (C) absence of chronic cardiac pulmonary disease, either clinically or at autopsy; and (D) absence of arterial disease, such as syphilis, or more than minimal atheromatosis or arteriolar sclerosis, then it is considered an idiopathic pulmonary aneurysm [Green 1949]. Our patient fulfills the above criteria. We do not have any follow-up documents before the 22-month period, but undoubtedly the aneurysm should have a longer history. Ring et al reported on their follow-up with 4 patients with idiopathic pulmonary aneurysms. These 4 patients were followed between 12 to 46 years [Ring 2002]. Similarly, van Rens et al reported a patient that was diagnosed in 1959 with angiography who refused surgical intervention at that time. In the recent digital subtraction angiography of the patient, there was no change in the size of the pulmonary artery aneurysm [van Rens 2002]. Another case was reported by Rose et al, and the diagnosis was made when the patient was 15 months old and there was no proportional increase in the size of the aneurysm at the 29-year follow-up [Rose 2002]. All the cases described above were asymptomatic without pulmonary hypertension, as is our case. In contrast to these reports, Shimokawa et al reported a case that was diagnosed as pulmonary artery

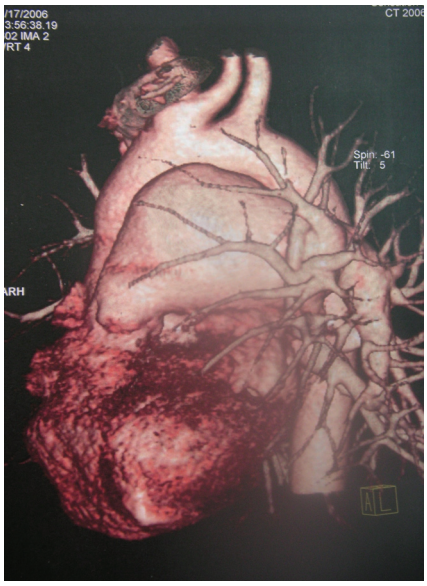

Figure 2. Multislice computed tomography scan shows dilatation of the main pulmonary artery started 1 cm above the pulmonary valves.

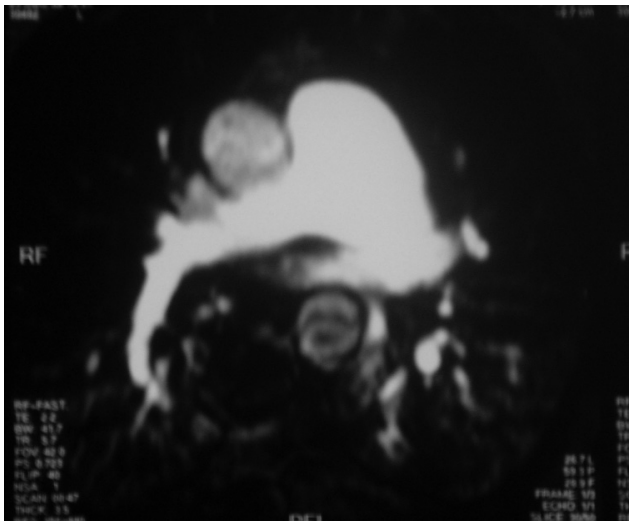

Figure 4. Cardiac catheterization image of the pulmonary artery aneurysm.

Classification of Pulmonary Artery Aneurysms (PAAs)

PAAs without an arteriovenous communication
Infection (mycotic aneurysms): tuberculosis (Rasmussen's aneurysms), syphilitic infection, other (bacterial, fungal)
Structural heart disease (congenital, acquired)
Structural vascular disease (Marfan, Behçet, vasculitis)
Pulmonary hypertension
Idiopathic (isolated, Huges-Stovin syndrome)
Trauma
PAAs with an arteriovenous communication
Congenital (isolated, associated hereditary hemorrhagic teleangectasia)
Acquired (infection, trauma)

aneurysm with a 6-cm diameter at the first operation of mitral valve replacement and tricuspid annuloplasty. The patient had mild pulmonary hypertension (47/16 mmHg). After 4 years of follow-up, the aneurysm reached a diameter of 10 cm and the patient was operated on because of the risk of rupture [Shimokawa 1997].

Symptomatic cases with significant pulmonary regurgitation or stenosis (which is enough to cause right ventricular dysfunction), pulmonary hypertension, or association with other cardiac lesions are candidates for surgery. Surgical intervention is commonly recommended when an aneurysm is diagnosed. This approach appears to be based on anecdotal reports of fatal aneurysm rupture [Deterling 1947; Finch 1983]. But the relationship between size and location of the aneurysm, pulmonary pressure, rate of diameter enlargement, and rupture remains unclear. According to these reports, treatment strategies for aneurysms elsewhere in the body may or may not be applicable for patients with idiopathic pulmonary artery aneurysm.

Our case was idiopathic and isolated, without pulmonary hypertension, as occurs in "low-pressure pulmonary artery aneurysm" patients, who may have a lower risk of rupture and progression. Our patient was stable at 22-month follow-up. We plan an elective surgical repair if signs of compression of adjacent vital structures, thrombus formation in the aneurysmal sack, or a ≥0.5-cm increase in the diameter of the aneurysm in 6 months are observed during the follow-up period. Surgical techniques differ for the management of aneurysms, including aneurysmorrhaphy, reconstruction with a pericardial patch, arterioplasty, and homograft or synthetic graft interposition [Arom 1978].

We think that the surgical intervention for a main pulmonary artery aneurysm should not be aggressive as it is for patients with an aneurysm of the thoracic aorta, especially when there is normal pulmonary artery pressure, as in our patient. However, the patient should be under close follow-up for at least a 6-month period, and if the patient has dyspnea on exertion, chest pain, hemoptysis, new onset of pulmonary artery insufficiency, or an increase in the diameter of the aneurysm, surgical management should be recommended.

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