

Case Report

Idiopathic Asymptomatic Aneurysms of Pulmonary Artery and Ascending Aort: A Case Report

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Abstract: The dilatation of pulmonary artery is a rare condition and called as pulmonary artery aneurysm. Aneurysm of both the pulmonary trunk and the ascending aorta is even rarer. Symptoms are due to aneurysm compression in adjacent anatomical structures. The main indicator of treatment is the pulmonary artery pressure. The prognosis and treatment of pulmonary artery aneurysm is unclear. Herein we present a case of a main pulmonary artery and ascending aortic aneurysms without underlying pathology. Because our case was asymptomatic, without a initiative cardiac lesion and/or pulmonary hypertension; we decided to follow-up him without operation and he was stable at 24-month follow-up.

Keywords: Ascending Aortic Aneurysm, Idiopathic, Pulmonary Artery Aneurysm

1. Introduction

It has been described for about a hundred years and many definitions have been defined. Pulmonary artery aneurysm (PAA) is a morphological abnormality which is characterized by focal dilatation of the vessel involving all three layers of the vessel wall. PAA is defined by a diameter of the pulmonary artery greater than 4 cm. [1] If an aneurysm does not involve all layers of the arterial wall it is called as pseudoaneurysm. Dissection and rupture of aneurysm are the main causes of death with PAA. There is no consensus in the literature on the treatment of PAA.

Aneurysm of both the pulmonary trunk and the ascending aorta is very rare. We report our experience of one main pulmonary artery and ascending aortic aneurysms with no underlying pathology case.

2. Case Report

A 49-year-old male patient with PAA detected in echocardiography and he was referred to our department for further evaluation, We performed computed tomography (CT).

CT scan revealed that the ascending aorta dilatation was 50 mm and the pulmonary trunk dilatation was 52 mm. (Figure 1).

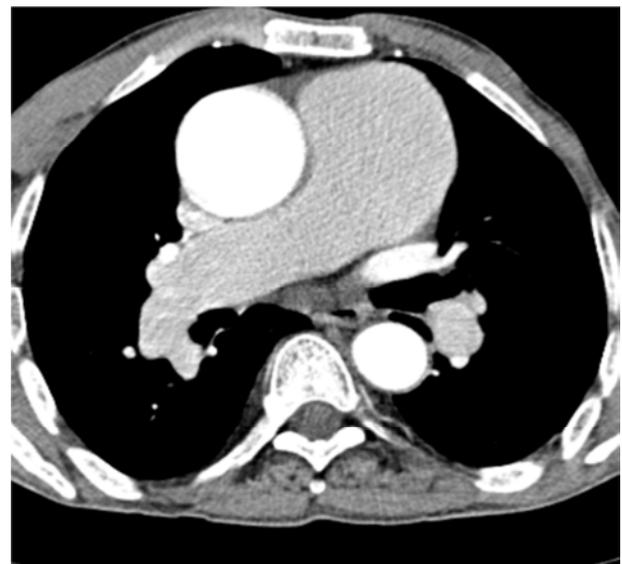


Figure 1. The computed tomography angiography showing the pulmonary and the ascending aortic aneurysms.

His physical examination was normal. The patient was in sinus rhythm, afebrile, and normotensive, and there were no features of cardiac failure. Electrocardiography study was normal. Heart sounds were of normal intensity. The pulmonary artery pressure estimated by means of echocardiography was normal (25 mm Hg).

No signs of intracardiac or extracardiac shunting were found. In his past medical history he only mentioned about controlled hypertension. There was no history of syphilis, tuberculosis, vasculitis, rheumatoid arthritis, or chronic obstructive airway disease. Serological tests were negative and antinuclear antibody level was not raised. Additional laboratory studies excluded inflammatory and collagen disorders.

The patient was diagnosed with idiopathic PAA and started metoprolol succinate 50 mg Daily.

He was asymptomatic so we decided to arrange regular follow-up visits for him. He was scheduled for follow-up every 6 months. He was recommended a routine follow-up every 6 months or when any cardiac or respiratory symptoms such as chest pain or dyspnea occurred. He was stable at 24-month follow-up. The diameters of both two main arteries are same as previous measurement.

3. Discussion

The dilatation of pulmonary artery is called as PAA. Normal diameters differ in different patients. The upper limit for adults of the main pulmonary artery (PA) diameter is 29 mm, and the upper limit of the interlobar PA is 17 mm in computed tomography. [2] However specific prevalence of PAA is unknown, it was reported in 1 out of every 14000 autopsies. [3] Eighty-nine percent of all PAAs were located in the main PA, whereas only 11% were located in the pulmonary branches. [4]

Fragility of the media with degeneration and disintegration of elastic fibers, high-wall tension and an abnormal pulmonary valve are the causes of pulmonary trunk aneurysm. Lack of pulmonary valve leaflets and narrowing of the ventricular arterial component may be associated with major PAA. Cystic medial necrosis is the most important histopathologic factor in the development of rare idiopathic PAA. [5]

Most patients with PAA are asymptomatic or have non-specific symptoms. Many patients are often accidentally diagnosed with the disease because they remain asymptomatic. Dyspnea, hemoptysis, chest pain, and cough are the unspecific symptoms that correlated with PAA. [4] Cyanosis, cough, increasing dyspnea, pneumonia, fever, and bronchiectasis may be because of bronchus compression by a large PAA. Hemoptysis may signify dissection or rupture of the aneurysm, which may necessitate urgent intervention [5] The incidence of pulmonary embolism is high in PAA patients. Idiopathic PAA is thought to be the cause of rupture, pulmonary artery dissection or sudden cardiac death. Most pulmonary artery dissections are diagnosed only at autopsy because of the high mortality rates. The most common site of dissection is the

PAA main body (at 80%) and in alive patients only 15% of the PAA dissections are diagnosed. [4] Severe dyspnea, retrosternal chest pain, central cyanosis, cardiogenic shock, and sudden death are the clinical symptoms of dissection. And the main cause of death is cardiac tamponade.

If the pulmonary artery pressure is normal, the main pulmonary artery aneurysms do not show the same aggressiveness as aortic aneurysms. PAAs are usually seen in younger age groups than in aortic aneurysms and have an equally sexual incidence. [4] In our case at the end of the 2 years both diameters of ascending aort and pulmonary artery were same as the initial dimeters.

When the chest radiograph shows a significant enlargement of the left pulmonary hilum it is often accidentally diagnosed. For PAA the gold standard diagnostic tool is pulmonary angiography. But two-dimensional transthoracic and transesophageal echocardiography, magnetic resonance imaging and CT can help in diagnosis. [4] In our case aneurysm was diagnosed by echocardiography.

It has defined 4 pathological criteria for an idiopathic PAA: simple dilatation of the pulmonary trunk with or without involvement of the rest of the arterial tree, the absence of intracardiac or extracardiac shunts, the absence of chronic cardiac or pulmonary disease, and the absence of arterial disease such as syphilis or more than minimal atheromatosis or arteriosclerosis of the pulmonary vascular tree. [4] According to these criteria our patient had idiopathic pulmonary artery aneurysm.

Congenital causes are the major reason for PAA formation. To date, more than 50% of cases are associated with congenital heart disease. The most common association is patent ductus arteriosus, followed by ventricular and atrial septal defects. [4] In the past before antibiotic therapy the most common cause of PAA were tuberculosis and syphilis. Primary or secondary pulmonary hypertension, chronic pulmonary embolism, or pulmonary valve stenosis, systemic disorders, such as Behçet's syndrome, Osler's disease or Marfan syndrome, or with other congenital and acquired cardiovascular diseases are possible etiologies of PA aneurysm. The most common type of vasculitis associated with pulmonary aneurysms is Behçet's disease and Hughes-Stovin syndrome [6]

In Behçet's disease, systemic findings (recurrent mouth and genital ulcers, uveitis) and PAA involving the right lower lobe arteries, often accompanied by thrombosis, is seen. In Hughes-Stovin syndrome, recurrent venous thrombosis and PAA are seen. Focal pulmonary artery enlargement is seen in PAA which is called 'Rasmussen's aneurysm' after pulmonary tuberculosis [7]

Only 5 patients were diagnosed as idiopathic pulmonary artery aneurysm in a series of 51 pulmonary artery aneurysm repair in Mayo clinic [8]

We do not have precise information about the treatment of pulmonary artery aneurysm and indications for surgical repair. In terms of surgical outcome, mortality and morbidity data are not available because a large series of PAA patients is not published. To replace the PA and the pulmonary trunk with a

conduit starting in the right ventricular outflow tract is the most common procedure.

Van Rens et al. [9] suggested that the long-term outcome of pulmonary artery aneurysm was favorable without surgical treatment. They reported a case of idiopathic left pulmonary aneurysm with a follow-up period of 40 years.

Kuwaki et al. suggested surgical repair of major pulmonary aneurysms regardless of underlying disease or etiology if low operative risk is present. [10] Surgery is suggested for patients with an aneurysm having a diameter of 60 mm or greater. [11] Dacron graft replacement, replacement with combination of Dacron prosthesis and bioprosthesis, aneurysmorrhaphy, and pulmonary allograft repair are the suggested surgical techniques.

Patients should be re-evaluated regularly, because idiopathic PAA seem to be a relatively benign condition and conservative treatment seems suggestable for asymptomatic patients with PAA with no significant PAH and stability in PAA diameter.

Surgery should strongly be considered in case of compression of adjacent structures, thrombus formation in the aneurysm sack, ≥ 5 -mm increase in the diameter of the aneurysm in 6 months, the appearance of clinical symptoms, evidence of valvular pathologies or shunt flow, and verification of PAH. [4]

Sugimoto et al reported a case with aneurysm of both the pulmonary trunk and the ascending aorta concomitant with bilateral bicuspid valves. [12] They achieved surgical repair for the pulmonary artery and ascending aortic aneurysms, concomitant with bilateral bicuspid semilunar valves successfully in a 63-year-old woman.

There is a report that explains the association of pulmonary aneurysm with syphilis, ankylosing spondylitis, rheumatoid disease, rheumatic fever, Reiter's syndrome, Takayasu's disease, and giant-cell arteritis. [13]

Our case report examines simultaneous aneurysms of the ascending aorta and the pulmonary trunk. But our case was not associated with any connective tissue disorder, had no evidence of arteritis, and was unrelated to the mentioned disease and disorders.

4. Conclusion

The dilatation of pulmonary artery is a rare condition and called as pulmonary artery aneurysm. Symptoms are due to aneurysm compression in adjacent anatomical structures. The prognosis of PAA is unclear. This is the compelling nature of the disease, and when it is diagnosed, it is impossible to determine when the disease has began. The pulmonary artery pressure is the main indicator of therapeutic options. PAA with low pulmonary artery pressure has a relatively better prognosis

However, in patients with high pulmonary artery pressure, the risk of future complications is high and surgery should be

considered for these patients. Aneurysmectomy and repair or replacement of the right ventricular outflow tract seem to be the preferred methods. Idiopathic low pressure PAA may have a good prognosis as in our patient without surgery. In PAA patients without pulmonary hypertension, it is considered that the long term aneurysm rupture is very low. The progression of the aneurysm will be better known with regular long-term follow-up of patients with normal pressure PAA

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