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Case Report

Huge idiopathic pulmonary artery aneurysm

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ABSTRACT

A pulmonary artery aneurysm is an uncommon anomaly. The clinical manifestations are mostly nonspecific, and management is controversial. We report a case of a 67-year-old woman with a main pulmonary artery aneurysm who did not take surgical intervention. Subsequently, there was no increase in size for 3 years.

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Introduction

A pulmonary artery (PA) aneurysm is a rare anomaly of the cardiovascular system, with an incidence of 1 in 14,000 in necropsies [1]. The definition of PA aneurysm is a PA with a diameter over 40 mm [2,3]. Most patients present with nonspecific symptoms such as hemoptysis, dyspnea, chest pain, and cough [4]. PA aneurysms can be caused by congenital, acquired, and idiopathic reasons [5]. We report this case, as we had a rare case of a 67-year-old woman with huge idiopathic PA aneurysm, who was treated well with conservative treatment.

Case presentation

A 67-year-old female came to the cardiology department of this hospital because of shortness of breath and chest pain. The

patient had been taking medications for hypertension and asthma for more than 10 years. On examination, the blood pressure was 120/80 mm Hg and the pulse 73 beats per minute. Chest radiograph showed cardiomegaly with dilated main PA (Fig. 1). The electrocardiogram showed normal sinus rhythm. The transthoracic echocardiography showed normal left ventricular systolic function with ejection fraction of 73%. There were mild tricuspid regurgitation and mild pulmonary regurgitation, and the pulmonary flow acceleration time was 83 msec. The diameter of PA was 64.5 mm in echocardiography (Fig. 2). The pulmonary angiography demonstrated main PA aneurysm (Fig. 3), and the pulmonary transvalvular peak pressure gradient was 52 mm Hg. The chest computed tomography (CT) angiography revealed a main PA aneurysm sized 65 mm in transverse diameter (Fig. 4A). It was diagnosed as idiopathic because the patient had no comorbidities that can cause PA aneurysm. We recommended surgery for the PA aneurysm, but

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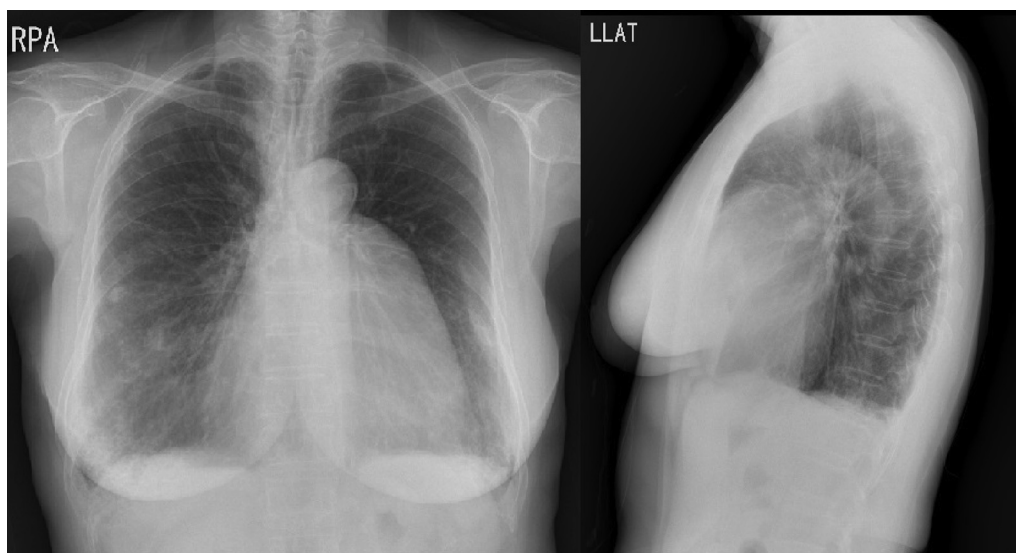


Fig. 1 – Chest X-ray showed cardiomegaly and a mass in the left hilum.

the patient refused surgery. The patient was managed with Beraprost sodium to control the pulmonary arterial hypertension (PAH) and discharged with medication. The patient was followed up for 3 years with CT angiography, and there was no significant increase in size of the PA aneurysm (Figs. 4B and C).

Discussion

The patient presented in this report is a 67-year-old female who suffered from dyspnea and chest pain. She was diagnosed with PA aneurysm by echocardiography, pulmonary angiography, and chest CT angiography.

The PA aneurysm is an unusual anomaly. Deterling and Claggett [1] found only 8 cases of PA aneurysm in 109,571 postmortem examinations. The causes of PA aneurysm can be differentiated into congenital, acquired, and idiopathic causes [5]. More than 50% of all cases were associated with congenital heart disease. Increased flow caused by left-to-right shunt

results in increased hemodynamic shear stress on the vessel walls and therefore promotes aneurysm formation in the PAs. The three most frequent congenital heart defects associated with a PA aneurysm are persistent ductus arteriosus, ventricular septal defects, and atrial septal defects. Acquired causes include untreated syphilis and tuberculosis, Behcet's disease, and vasculitis of PA. Lung cancer and tumors arising from PA may also lead to PA aneurysm. Otherwise, it is classified as idiopathic PA aneurysm. Idiopathic PA aneurysm formation is rare, but an increasing number of cases are being reported in the literature [1,4,5]. PAH is an important cause of PA aneurysm formation. About two-thirds of PA aneurysm is associated with PAH [6]. Hence, the pathophysiology of PA

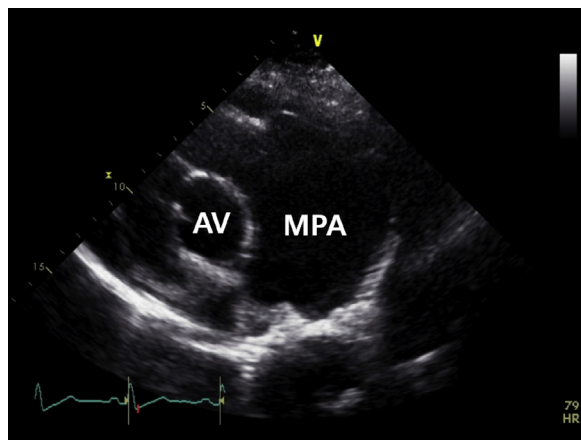


Fig. 2 – Transthoracic echocardiography revealed an enlarged main pulmonary artery. AV, aortic valve; MPA, main pulmonary artery.

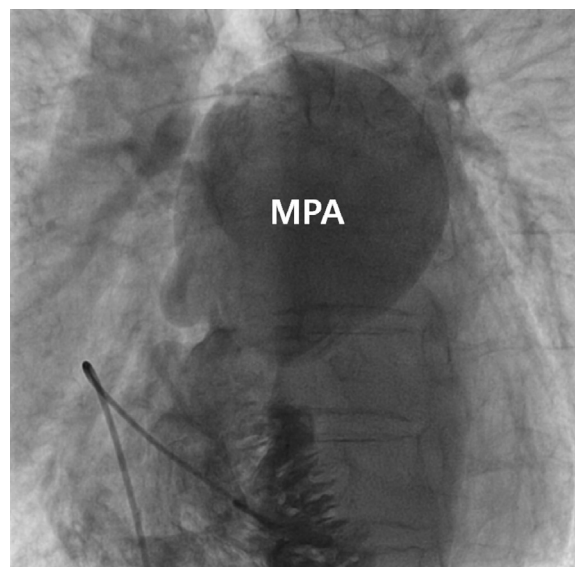


Fig. 3 – Pulmonary artery angiography demonstrated markedly dilated main pulmonary artery. MPA, main pulmonary artery.

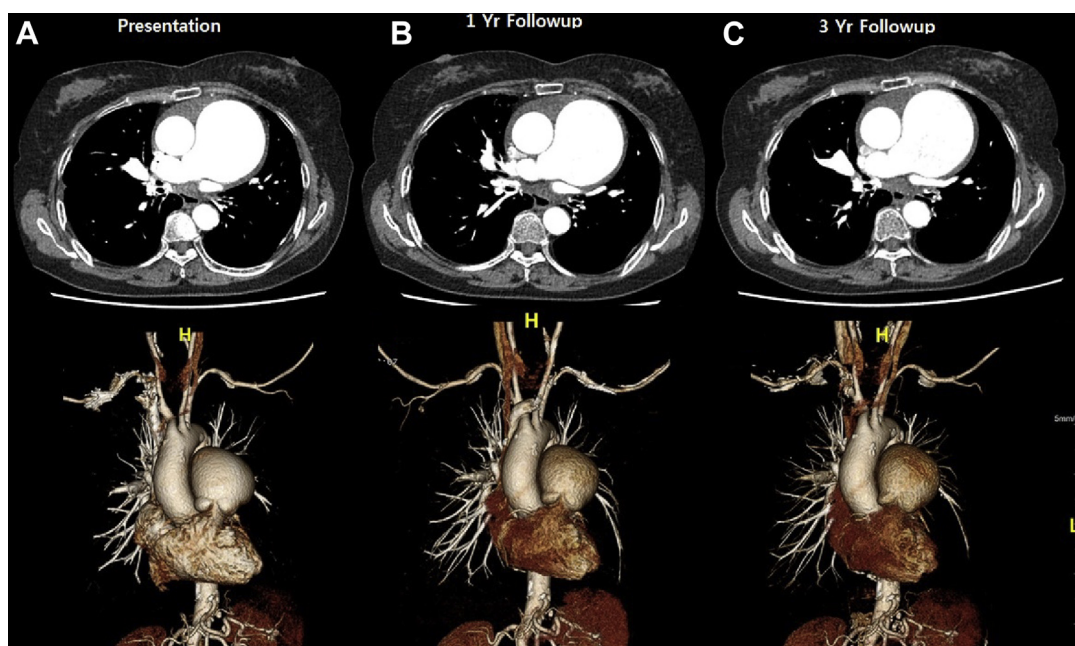


Fig. 4 – (A) Chest CT angiographic image (above) and three-dimensional reconstruction image (below) revealed a main PA aneurysm sized 65 mm in transverse diameter at initial admission; (B) 1-year follow-up; and (C) 3-year follow-up. There was no significant change in the diameter of the pulmonary artery aneurysm. CT, computed tomography; PA, pulmonary artery.

aneurysm might be associated with increased PA pressure. According to the law of Laplace, blood pressure and vascular radius are directly proportional, and wall thickness is inversely proportional to the wall stress [5]. Thus, increased PA pressure in PAH can cause structural changes in the vessel wall, leading to PA dilatation [7].

Most patients with PA aneurysm have no symptoms or just have nonspecific symptoms such as hemoptysis, abnormal chest X-ray, dyspnea, chest pain, and cough [1,4,8].

Various cardiac imaging modalities can be used for finding the PA aneurysm. Pulmonary angiography still remains as the gold-standard method for diagnosing the PA aneurysm. It can be used to check pressure gradients within the right side of the heart and evaluate extension of the aneurysmal structure into the vascular structure [8]. The role of noninvasive imaging modalities such as CT angiography and magnetic resonance angiography is expanding along with the development of image gathering and reconstruction techniques [9,10]. Trans-thoracic echocardiography or transesophageal echocardiography can also detect the PA aneurysm, and both are useful for assessing heart and valvular functions [5,11].

The natural history of proximal PA aneurysm is poorly understood because of limited number of cases. Possible complications of PA aneurysm include dissection, airway compression, and thrombus in PA. Dissection of PA aneurysm is a life-threatening complication. Among all reported cases, one-third of the patients died as a result of rupture [5,6,8].

There are no clear therapeutic guidelines for the PA aneurysm. Surgical repair can be recommended if the patient is symptomatic or the diameter of the PA is above 5.5 cm or for PA with complications such as dissection, airway compression, and thrombosis. Patients with PAH should be considered

for surgical treatment owing to risk of dissection and rupture. Otherwise, it can be managed with conservative treatment [5,6].

Conclusion

The incidence of the PA aneurysm is rare. Surgical repair is recommended if the diameter of the PA aneurysm is above 5.5 cm or increase in the diameter of PA aneurysm is above 0.5 cm in 6 months or PA aneurysm with PAH. However, in our case, the patient was stable, and the PAH was controlled well. The PA aneurysm was not enlarged for 3 years. Thus, the patient was successfully managed by medication alone.

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