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Low Pressure Pulmonary Artery Aneurysm with Atrial Septal Defect

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Pulmonary artery (PA) aneurysm is a rare finding in the thoracic cavity, accompanied by pulmonary hypertension. Clinical presentation of PA aneurysms is usually asymptomatic. The guideline for PA aneurysm treatment is unclear. We report an unusual case of low pressure PA aneurysm associated with atrial septal defect in a 69-year-old man.

KEY WORDS: Low pressure · Pulmonary artery · Aneurysm · Atrial septal defect.

Introduction

Pulmonary artery (PA) aneurysm is a rare condition and has been associated with structural cardiac and vascular abnormality, pulmonary hypertension, infection, vasculitis, connective tissue disease and trauma.[1)] Most patients present with nonspecific symptoms and are referred with the abnormal mass on chest X-ray.[2)] The two-thirds of PA aneurysms are associated with pulmonary hypertension. There are no definitive guidelines about its optimal management. In asymptomatic low pressure PA aneurysm, possibility of rupture is low due to low PA pressure. A conservative treatment may be considered and follows up with echocardiography.

Case

A 69-year-old male was referred to our department for evaluation of a lung mass which is incidentally found on a routine chest X-ray. He was asymptomatic and physical examination did not reveal cardiac murmurs. There were no finding to doubt infection, vasculitis, collagen vascular disease, and trauma in the physical examination. Specific finding was not found in the laboratory finding. Chest X-ray showed left hilar enlargement (Fig. 1). Chest computed tomography showed aneurysmal dilatation of main PA (Fig. 2). The maximum diameter of the aneurysm was 56 mm. Transthoracic echocardiography showed PA aneurysmal formation with mild pulmonary regurgitation (Fig. 3A) and small atrial septal defect (Fig. 3B). The pulmonary to systemic flow ratio (Qp : Qs ratio) was 1.2. The pulmonary transvalvular peak pressure gradient was 8 mmHg. The right heart chambers appeared mildly dilated

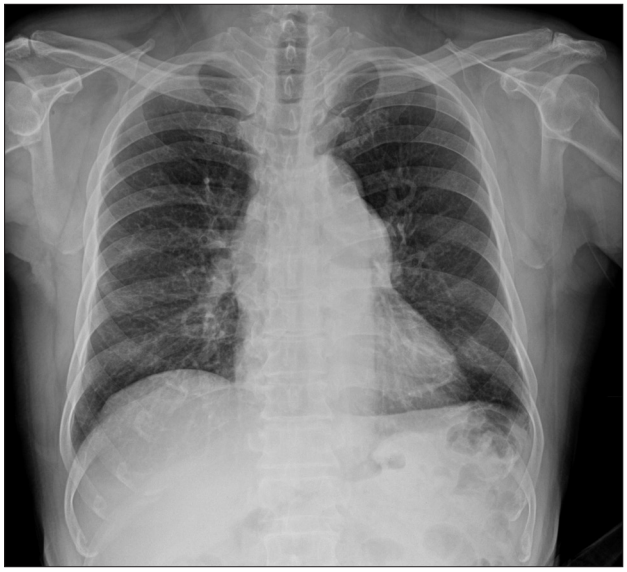

Fig. 1. Chest X-ray shows dilated main pulmonary artery.

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with pulmonary systolic artery pressure of 26 mmHg. He has been considered surgery for the PA aneurysm but refused it. And he was stable at 6 months follow-up.

Discussion

PA aneurysm is a rare disease and its estimated incidence is 1 in 14000 in autopsies.[1] The definition is a focal dilatation greater than 4 cm in diameter.[3] It has been associated with congenital heart disease, especially patent ductus arteriosus, pulmonary valve stenosis, atrial septal defect, pulmonary hypertension, infection, vasculitis, collagen vascular disease, trauma, idiopathic pulmonary aneurysm.[1][2][4] The two-thirds of PA aneurysms are associated with pulmonary hypertension.[5] The pathophysiology of the PA aneurysm is associated to vessel wall stress that leads to progressive dilatation or even rupture. The law of Laplace dictates that wall stress is directly proportional to the vascular pressure and radius of the vessel and is inversely related to the wall thickness.[6] However, in our patient, the natural history of low pressure PA aneurysm was unclear and has been thought to be due to inherent weakness of the arterial wall. Most patients with PA aneurysms are asymptomatic or unexplained dyspnea and are referred with the suspicion of a mass or vascular dilatation seen on chest X-ray.[2] The confirmative diagnosis may be made with echocardiography or computed tomography. There are no definite guidelines on the management of PA aneurysm due to its low prevalence. Surgical management may be needed. However, low pressure PA aneurysm is a benign condition with better survival than other etiologies and conservative treatment is may recommended such as our patient.[7]

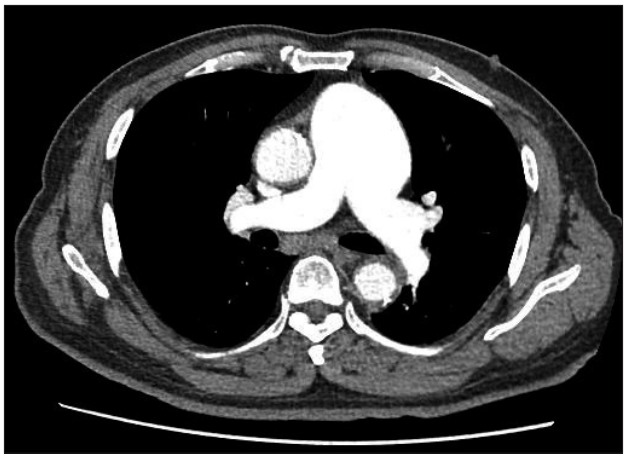

Fig. 2. Chest computed tomography demonstrates dilated main pulmonary artery and proximal branches.

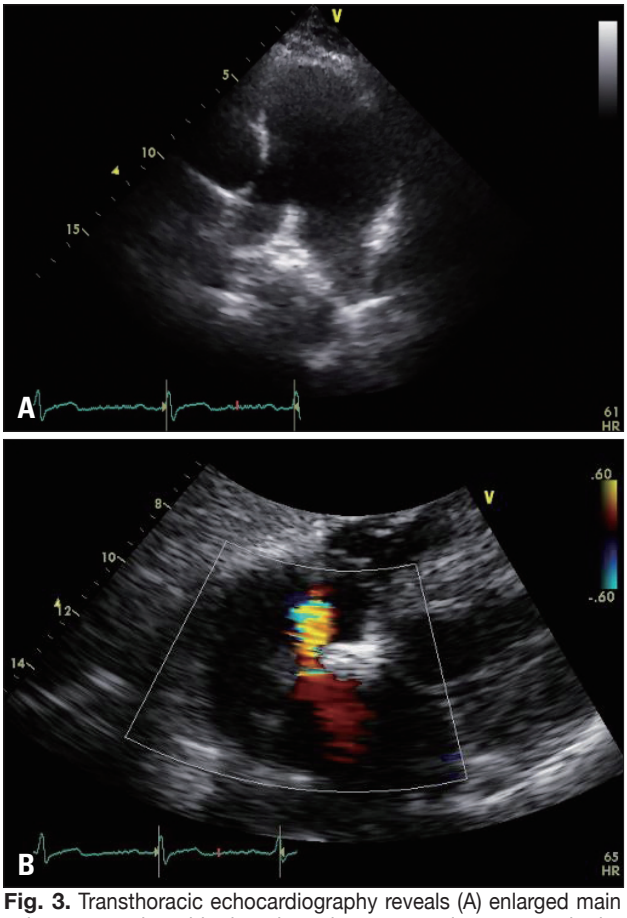

Fig. 3. Transthoracic echocardiography reveals (A) enlarged main pulmonary trunk and both major pulmonary arteries are seen in the basal short-axis view. (B) There is a small atrial septal defect in the subcostal view.

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