Aneurysmal Dilatation of the Pulmonary Trunk with Mild Pulmonic Stenosis

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We present the unusual case of a 72-year-old woman whose chest X-ray showed an abnormal left hilar shadow. A pulmonary angiogram revealed an aneurysm in the pulmonary artery with a diameter of 55 mm that extended from the main pulmonary trunk to its bifurcation. Mild pulmonic stenosis with a systolic pressure gradient of 18 mmHg across the pulmonic valve was recognized. Mild dilatation of the ascending aorta was also present. The pressure gradient across the pulmonic valve was lower than is typical for an aneurysmal dilatation, suggesting that this patient represented a case of idiopathic pulmonary artery dilatation. We suspected the presence of a congenital structural alteration common to the pulmonary artery and the ascending aorta. (Internal Medicine 34: 199-202, 1995)

Key words: pulmonary aneurysm, aortic dilatation, pulmonary angiogram, MRI

Introduction

The incidence of pulmonary artery aneurysm is extremely low, and its clinical significance is still unknown. The majority of cases are associated with congenital heart disease, most commonly with left-to-right intracardiac shunting (1-3). Infectious conditions such as mycosis and syphilis, and conditions such as aortitis and Marfan's syndrome are also associated with pulmonary artery aneurysm. Pulmonary stenosis is commonly associated with some degree of post-stenotic dilatation of the pulmonary artery, although aneurysmal changes rarely cause complications. Pulmonary artery aneurysm is more often associated with severe valvular stenosis. We evaluated a 72-year-old woman with a pulmonary artery aneurysm accompanied by mild pulmonary stenosis.

Case Report

A chest X-ray obtained as part of an annual health examination in May 1991 showed an abnormal left hilar shadow in a 72-year-old Japanese woman. A mediastinal tumor was suspected. However, she had no physical complaints and refused further examination. In January 1993, the same abnormality was observed on her chest X-ray and she was referred for further evaluation. She visited the Jichi Medical School Hospital in February 1993.

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2.67 m/s and the pressure gradient, estimated by the modified Bernoulli equation ($Ap=4v^2$), was 28.5 mmHg. Dilatation of the pulmonary trunk was also detected, suggesting a post-stenotic change in the pulmonary artery. No shunt flow was observed during transesophageal echocardiography.

CT and MRI of the chest showed aneurysmal dilatation of the main pulmonary artery extending to the point of bifurcation (Fig.2). The maximum diameter of the aneurysm in the MRI slice of the pulmonary trunk was 55 mm. Decreased thickness of the pulmonary arterial wall was observed, but no dissection was detected. Dilatation of the ascending aorta with a maximum diameter of 40 mm was also observed. The dimensions of the other parts of the aorta were normal.

Cardiac catheterization was performed. Right ventricular systolic pressure was 44 mmHg and pulmonary arterial systolic pressure was 26 mmHg. There was a peak-to-peak pressure gradient of 18 mmHg. The pulmonic valvular area calculated by pressure gradient and flow volume across the pulmonic valve was 1.4 cm². There was no left-to-right shunt. A right ventriculogram obtained from the lateral projection showed that the right ventricular cavity was of normal size, without hypertrophy. A systolic doming formation on the pulmonic valve and marked dilatation of the pulmonary trunk (maximum diameter 55 mm) were observed (Fig. 3). Biplane left ventricular cineangiography showed that the size and motion of the left ventricle were normal. No significant stenotic lesions were observed in coronary arteries.

**Discussion**

A pulmonary artery aneurysm of the pulmonary trunk, defined as a dilatation of more than 50 mm in diameter of the main pulmonary artery, is rare (1). Deterling and Clagett identified 8 such cases from reports of 109,571 autopsies (2). Groedal found the frequency ratio of pulmonary aneurysm to aortic aneurysm to be 1:250 (3). About half of the cases of pulmonary artery aneurysm are associated with congenital heart diseases, such as patent ductus arteriosus, atrial septal defects and ventricular septal defects, that cause volume and pressure overloading in the right cardiac system. Other causes include inflammatory changes in the artery resulting from mycosis and syphilis, trauma, primary pulmonary hypertension, cystic medial necrosis and aortitis (2, 3) (Table 1). Pulmonary aneurysms associated with pulmonic valvular stenosis are especially rare. Echocardiographic findings and data obtained from cardiac catheterization in the present case suggested the existence of pulmonic valvular stenosis.

There are only a few reports of pulmonary aneurysms associated with pulmonic stenosis with pressure gradients below 20 mmHg (1, 4). Van Buchem et al described a 21-year-old man in whom marked dilatation of the trunk of the pulmonary artery (73 mm in diameter) was observed, although there was only a slight difference in systolic pressure between the right ventricle and the pulmonary artery (15 mmHg) (4). It is possible that dilatation of the pulmonary artery slows blood flow, leading to a small decrease in pulmonary artery pressure, and resulting in only a small difference in systolic pressure between the right ventricle and the pulmonary artery. If so, the diameter of the pulmonary artery aneurysm would not necessarily correspond to the severity of the pulmonic stenosis. The present patient also showed a markedly dilated pulmonary artery with only mild pulmonic stenosis. The clinical significance of the pulmonic stenosis in this patient is still unclear. Although the mild pressure gradient may have a positive effect on the aneurysmal dilatation, this patient appeared to represent a case of idiopathic pulmonary artery dilatation.
A Case of Pulmonary Artery Aneurysm

Shilkin et al reported that 7 of 17 autopsy cases with pulmonary aneurysms showed medial necrosis of the aneurysmal wall (5). D’Arbela et al described a case of pulmonary aneurysm associated with patent ductus arteriosus (6). Pools of alcian blue-stained mucopolysaccharide were observed in the media, fragmented smooth muscle and elastic fibers of the pulmonary artery. We speculated that an abnormal structure of the arterial wall or the combination of atherosclerosis and pulmonic stenosis contributed to the aneurysmal changes in the present case.

The best therapeutic intervention for an aneurysm of the pulmonary artery is difficult to determine. Rupture has occurred in cases of pulmonary hypertension associated with congenital heart disease (5), Marfan’s syndrome (7) and mycosis (8). However, in a 10-year follow-up of several patients no clinical complications were observed (9, 10). There are no reports of rupture associated with pulmonic stenosis. In the present case, there was no marked change in the size of the aneurysm over 2 years, suggesting that the pulmonary artery was dilating very slowly. The patient has experienced no adverse symptoms or pulmonary hypertension, and is being carefully monitored.

References
2) Deterling RA, Clagett OT. Aneurysm of the pulmonary artery. Am Heart
Fig. 3. A right ventriculogram showed that the right ventricular cavity was of normal size without hypertrophy. A systolic doming formation of the pulmonary valve and gross dilatation of the main pulmonary artery were observed. Arrows indicate the position of the pulmonary valve. PA: pulmonary artery, PV: pulmonary valve.

Table 1. Cause of Dilatation of the Pulmonary Artery

1. Congenital
   A. Left to right shunts: atrial septal defect, ventricular septal defect, patent ductus arteriosus, Lutembacher’s aortopulmonary fistula, etc.
   B. Unequal division of truncus arteriosus.
   C. Congenital malformation of arterial wall (with/without Marfan’s syndrome).
   D. Pulmonary valve anomaly: stenosis, bicuspid or quadricuspid valve, absence of pulmonary valve (with/without tetralogy of Fallot).

2. Acquired
   A. Pulmonary hypertension
      1) Extravascular
         a. Pulmonary parenchymal diseases: benign, malignant
         b. Mediastinal disease: benign, malignant
      2) Intravascular
         a. Left heart disease: mitral stenosis, etc.
         b. Primary pulmonary hypertension
         c. Pulmonary embolism
         d. Rupture of aortic aneurysm into pulmonary artery
   B. Destructive process of arterial wall
      1) Infectious: syphilis, bacterial, mycotic
      2) Atheromatous
      3) Trauma
      4) Cystic medionecrosis
   C. Traction of pulmonary artery
   D. Idiopathic