A 45-year-old man was referred to our hospital after a prolonged history of exertional dyspnea. Clinical examination revealed systolic and diastolic heart murmurs without any systolic click in the second right sternal border. Chest x-ray showed pulmonary artery dilation (Figure 1). Multidetector computed tomography showed a huge pulmonary artery aneurysm ~70 mm in diameter (Figure 2). Transthoracic echocardiography showed a huge aneurysm of the pulmonary artery by 2D echocardiography (Figure 3). There was no significant tricuspid regurgitation in either the 4-chamber view or the short-axis view, and pulmonary regurgitation due to dilatation of pulmonary annulus was seen by color-Doppler echocardiography (Movies I through III in the online-only Data Supplement). Cardiac catheterization did not show any pressure gradient between pulmonary artery and right ventricle or any sign of pulmonary hypertension. We diagnosed an idiopathic pulmonary artery aneurysm in this patient.

Aneurysmectomy of the pulmonary main trunk with a 24-mm synthetic graft and valvuloplasty of an enlarged pulmonary annulus were performed (Figure 4). The pathological examination did not show any sign of cystic medial degeneration (Figure 5).

Pulmonary artery aneurysm is a rare anomaly found in ~1 of every 14 000 autopsies; most of these anomalies are present in the main pulmonary artery. The cause of pulmonary artery aneurysm may be idiopathic; however, other causes include congenital shunt disease, syphilis, atherosclerosis, trauma, and pulmonary hypertension. In >50% of the postmortem cases, pulmonary aneurysms were associated with congenital heart disease, most frequently patent ductus arteriosus. In the remaining reported cases, such aneurysms were associated mainly with secondary acquired lesions, such as syphilis and cystic medial degeneration. In our case, however, no pathological abnormality was shown by the histological sections of aneurysmal wall.

Idiopathic pulmonary arterial aneurysm is considered to be a possible cause of rupture, dissection of pulmonary artery, or cardiac sudden death. Most of the pulmonary artery dissections are diagnosed only at necropsy, because of their high mortality rate. The natural history of an untreated idiopathic pulmonary artery aneurysm has not been well elucidated. A case report suggested that the long-term outcome of pulmonary artery aneurysm was favorable without surgical treatment. It is unclear whether there is a size hinge point similar to aortic aneurysms in which the risk of complication increases. Although some recommend medical treatment for such patients, others recommend surgical intervention for those with an aneurysm that has a diameter of 60 mm or greater.
Figure 2. Three-dimensional reconstruction of chest computed tomography showing the main pulmonary artery aneurysm (arrows).

Figure 3. Huge aneurysm of pulmonary artery observed by 2D echocardiography. Pulmonary regurgitation due to dilatation of pulmonary annulus is seen by color-Doppler echocardiography.

Figure 4. Surgical view of the pulmonary artery aneurysm (arrows).

Figure 5. Histology of pulmonary artery aneurysm wall with elastic-van Gieson stain. There was no remarkable pathological change.
Idiopathic Pulmonary Artery Aneurysm
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