CASE REPORT

# Main Pulmonary Artery Aneurysm

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Pulmonary artery aneurysm is a rare lesion of the thoracic cavity. Different etiologies have been reviewed, but idiopathic lesions without other symptoms are seldom reported. Usually, surgical interventions are suggested, but the long-term outcomes are not well established. Here, we report a 24-year-old man with main pulmonary artery aneurysm who successfully underwent aneurysmectomy and polytetrafluoroethylene vascular graft replacement. The postoperative course was uneventful, and the following image study revealed normal size of the great vessels. [*J Chin Med Assoc* 2007;70(10):453–455]

**Key Words:** aneurysm, graft replacement, main pulmonary artery aneurysm, surgical repair

## Introduction

Main pulmonary artery aneurysm (PAA) is an unusual lesion of the thoracic cavity.[1] Many underlying diseases have been described as etiologies of PAA;[2,3] however, isolated or idiopathic etiologies are relatively rare.[4,5]

Currently, there are no clear guidelines for surgical intervention in PAA because the clinical manifestations of PAA are not specific and treatments remain controversial. Generally, early surgical intervention is recommended for individuals with an underlying origin or for PAA of large size to avoid possible fatal rupture of the aneurysm.[6] Here, we report an unusual case where the main PAA was treated successfully with aneurysmectomy and polytetrafluoroethylene (PTFE) graft replacement.

## Case Report

A 24-year-old male patient was referred to our institution for further evaluation because abnormal grade II/VI systolic murmur over the pulmonary area was heard on routine health examination before his military service. He reported that he has had unusual heart murmurs since childhood. In our institution, transesophageal echocardiography, chest computed tomography, and angiography showed a lesion with 2-cm fusiform dilatation of the main pulmonary artery above the pulmonary valve (Figure 1). The maximal diameter was 5.2 cm, at which mean pressure measured by angiographic catheterization was 12 mmHg.

The operation was carried out through a median sternotomy. After pericardiotomy, the dilated main PAA was presented (Figure 2). Under mild hypothermia, cardiopulmonary bypass was performed with aortic and bicaval cannulations. The main pulmonary artery and bilateral pulmonary arteries were dissected carefully under cardiopulmonary bypass. Diseased

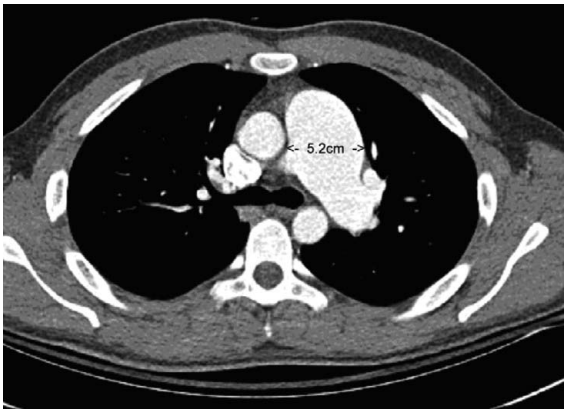


**Figure 1.** Preoperative chest computed tomography shows a main pulmonary artery aneurysm with a maximal size of 5.2 cm.

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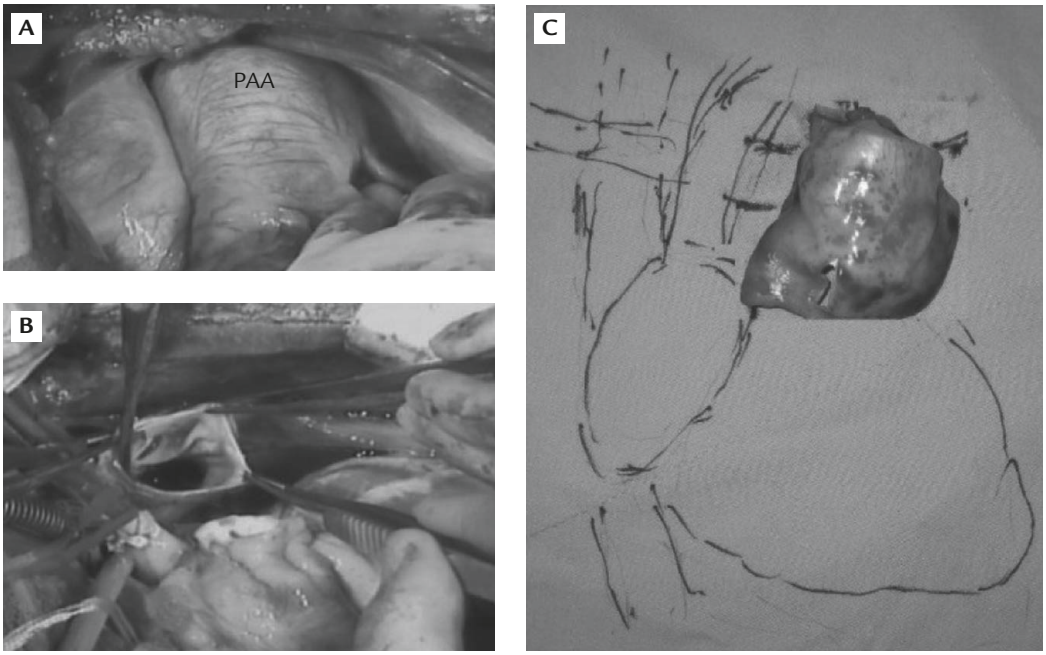


**Figure 2.** (A, B) Intraoperative photograph of the main pulmonary aneurysm, and (C) the resected pulmonary artery aneurysm.

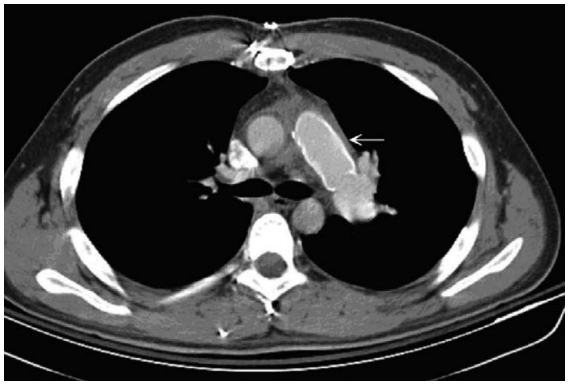

**Figure 3.** Chest computed tomography 1 month later, showing patent vascular graft with normal size (arrow).

pulmonary artery was excised, distal anastomosis from the left pulmonary artery to the pulmonary trunk root was started with a 22-mm PTFE graft, then the right pulmonary artery was connected to the PTFE graft with an end-to-side anastomosis. The postoperative course was uneventful; the patient was extubated 6 hours later, and discharged on the 6$^{th}$ postoperative day.

Pathology of the PAA specimen showed a picture of pulmonary aneurysm which consisted of focal fibrosis, myxoid degeneration of vascular wall, and congestion in adventitia with focal mild chronic inflammatory cell infiltration. The postoperative echocardiogram and chest computed tomography revealed normal heart systolic function without dilatation in the great vessels (Figure 3).

## Discussion

PAA is a rare lesion of the thoracic cavity, with an approximate incidence of 1 in 14,000 in autopsies.[1] Many etiologies have been discussed. Congenital structural heart disease, such as patent ductus arteriosus, and ventricular and atrial septal defects, accounts for half of the causes of PAA. Some connective tissue diseases, such as Marfan syndrome and Behcet syndrome, have been reported as well. Other conditions, i.e. pulmonary hypertension, idiopathic causes (isolated Hughes-Stovin syndrome), infections (mycotic aneurysms, syphilis, tuberculosis), and trauma have also been identified as possible causes.[2,3] Among all causes, idiopathic pulmonary trunk aneurysm is quite a rare clinical entity.[4,5]

The clinical manifestations are mostly nonspecific in patients with PAA, including hemoptysis, dyspnea on exertion, fever or cough, and chest pain. Severe hemoptysis may result in critical condition, and surgical intervention should be seriously considered in this group.[2,3] A loud systolic murmur may be heard over the left second intercostal space. The diagnostic tools include echocardiography, both transthoracic and transesophageal, and magnetic resonance imaging or computed tomography. Pulmonary angiography is suggested as the gold standard in diagnosis,[2,4] but other imaging studies may also be performed preoperatively.

According to previous literature, the management of PAA is controversial. Surgical intervention should be considered in some patients whose presentations

involve life-threatening symptoms such as massive hemoptysis or who have a large aneurysm size. Most reports of ruptured PAAs were associated with some underlying disease such as mycotic aneurysm, Behcet syndrome or other complications of some cardiac surgeries.[7–9] Veldtman et al reported that the timing of surgical intervention should be determined by changes in right ventricular size and function resulting from pulmonary regurgitation or pulmonary stenosis, rather than the size of the aneurysm.[10] Therefore, Imazio et al[3] and Kuwaki et al[11] believed that early surgical intervention for this main pulmonary artery lesion is indicated, because of the possible risk of aneurysm rupture, especially in mycotic lesions.[12] Fang and Tsai stated that idiopathic PAA is a benign condition with better survival than PAAs of other etiologies,[5] and conservative treatment is recommended for this group.

Several different surgical procedures are reported as treatments for PAAs, such as aneurysmorrhaphy, aneurysmectomy and repair with Dacron graft, or autologous pericardial replacement. Kuwaki et al considered that aneurysmorrhaphy may be relatively simple and not time-consuming.[11] However, for PAAs associated with connective tissue disorder or congenital structural heart disease, high recurrence is expected. Therefore, aneurysmectomy followed by graft replacement for these patients is advised.[11] Many choices of conduit for right ventricular outflow tract (RVOT) reconstruction have been described in pediatric cardiac surgery, including valve-less Goretex or Dacron tubes, homograft, porcine aortic xenograft and bovine jugular vein graft. Herijgers et al compared bovine jugular venous conduits with Dacron conduits for RVOT replacement in young sheep at 20 weeks' post-implantation; the bovine group showed good function, preserved structure and minimal calcification.[13] Boethig et al considered Contegras™ (Medtronic, Minneapolis, MN, USA) because it has the advantages of easy handling and availability for RVOT reconstruction, and compares well with homografts regarding freedom from explanation and reoperation.[14] Currently, authoritative guidelines for surgical intervention in PAA is still lacking, due to the nonspecific clinical manifestations of PAA and controversy over treatment. But early surgical intervention in individuals with an underlying origin or large aneurysm size is generally recommended.

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