Surgical Case of Isolated Pulmonary Takayasu’s Arteritis

Ichiya Yamazaki, MD; Yukio Ichikawa, MD; Masanori Ishii, MD; Toshiyuki Hamada, MD; Hirokazu Kajiwara, MD

Involvement of the pulmonary arteries is common in systemic Takayasu’s arteritis, but pulmonary arteritis alone is very rare. A 67-year-old woman had severe right pulmonary artery stenosis and left pulmonary arterial obstruction caused by Takayasu’s arteritis. Surgical treatment was successfully performed with extracorporeal circulation, without transection of the ascending aorta and the right pulmonary artery was enlarged with an autologous pericardial patch. (Circ J 2005; 69: 500–502)

Key Words: Isolated pulmonary lesion; Pulmonary artery; Surgical treatment; Takayasu’s arteritis

Takayasu’s arteritis is a primary, chronic, progressive, vascular inflammatory disease that causes stenosis and/or aneurysmal dilatation of the aorta and aortic arch branches, and in 50–80% of cases the pulmonary arteries as well! However, isolated pulmonary Takayasu’s arteritis is very uncommon. We present a case of isolated pulmonary Takayasu’s arteritis and describe the successful surgical treatment of this rare condition.

Case Report

A 67-year-old woman with a 6-month history of progressive exertional dyspnea was referred in January 2003. Laboratory examination showed an erythrocyte sedimentation rate (ESR) of 36 mm at 1 h and 70 mm at 2 h; C-reactive protein, 0.8 mg/dl; white blood cells, 9,800/μl; platelets, 42×10^4/μl. Rheumatoid factor, antinuclear antibody and Wasserman reaction were all negative. Human leucocyte antigens (HLA) were B52, A24 and DR2.

Pulmonary perfusion scintigraphy revealed complete absence of perfusate in the left lung and no defects in the right lung. Magnetic resonance imaging revealed remarkable wall thickness of the pulmonary trunk and proximal right pulmonary artery (Fig 1A). Magnetic resonance angiography (MRA) demonstrated severe stenosis of the right main pulmonary artery and obstruction of the left main pulmonary artery (Fig 1B). The aortic arch and its primary branches were normal. Cardiac catheterization showed that the systolic pressure of the main pulmonary artery was 85 mmHg and 25 mmHg in the right pulmonary artery, and the arterial pressure gradient was 60 mmHg. There was no stenosis of the coronary arteries and no incompetence of the aortic valves. A diagnosis of Takayasu’s arteritis based on the angiographic findings was confirmed by the laboratory, although the inflammatory response was mild.

In March 2003, the patient underwent surgical treatment. Exposure was obtained through a median sternotomy and total cardiopulmonary bypass, involving cannulation of the superior and inferior vena cava through the right atrium and of the ascending aorta, was instituted. Inflamed tissues were found in close contact with both the aorta and right pulmonary artery. Under cardioplegic cardiac arrest, the main trunk and bifurcation of the pulmonary artery were

![Fig 1](image-url)
Isolated Pulmonary Takayasu's Arteritis

Discussion

Isolated pulmonary Takayasu's arteritis is a very rare condition, comprising only 4% of all cases of Takayasu's arteritis. Surgical correction of pulmonary arterial lesions is also rare and only 7 reports of this procedure have been published.2-7

The present patient was 67 years old at onset of symptoms, which is not typical according to previous reports, but the laboratory data, HLA typing and angiographic findings supported the diagnosis of Takayasu's arteritis. Other differential diagnoses (e.g., Bechet disease, giant cell arteritis and syphilitic arteritis) were not supported by the history, physical findings and laboratory data.

We selected surgical correction rather than corticosteroid therapy. Steroid treatment relieves the symptoms, but requires high doses and long-term administration (at least several months). The present patient had a mild inflammatory response and symptomatic, severe lesions of the pulmonary artery and because we could neither predict the efficacy of high-dose steroid therapy nor consider long-term administration, we considered that surgical correction would be faster and more reliable in this case.

We were able to complete all procedures without transection of the ascending aorta to expose the proximal pulmonary artery.2,5,7 We used an autologous pericardial patch to enlarge the pulmonary arterial stenosis, but various methods have been described in previous reports. Moore et al performed both patch angioplasty of the left pulmonary artery and bypass grafting between the right and left pulmonary arteries using woven Dacron materials.3 Chauvaud et al did not use a conduit because of the risk of endoluminal fibrous proliferation within it and instead they used patch angioplasty with glutaraldehyde-preserved autologous pericardium. Dietl et al also used autologous pericardium for patch angioplasty, whereas Jakob et al used Dacron patch angioplasty to treat stenosis of the pulmonary artery bifurcation.6 Sugimoto et al resected the stenotic pulmonary artery bifurcation and replaced it with a tailored autologous pericardium roll.7 Although Lie et al describe 5 patients with isolated pulmonary Takayasu's arteritis who underwent surgery for either lung resection (3 patients) or reconstruction/bypass of the obstructed pulmonary arteries (2 patients), they do not give a detailed description of the materials and procedures used for the reconstruction and bypass.8

We selected autologous pericardium as the patch material for the following reasons. It was used in 3 of the 5 previous cases reports and has been used as cardiovascular patches, and in the reconstruction of the pulmonary artery and superior vena cava.9 It was easy to harvest. We favored the use of autologous pericardium over prosthetic materials such as Gore-Tex or Dacron primarily because of potential thrombogenicity. We also believed that prosthetic materials were inadequate for repairing pulmonary stenosis caused by inflammatory disease. We will follow this patient closely in subsequent years, paying particular attention to both the appearance of aortic lesions and the recurrence of pulmonary arterial lesions.
References