Reconstruction of Bilateral Branch Pulmonary Artery Stenosis Caused by Takayasu’s Aortitis

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A 63-year-old female presented with dyspnea on exertion. Her chest X-ray showed cardiomegaly, and right ventricular overload and tricuspid regurgitation were detected. Her pulmonary ventilation and blood flow scintigraphy findings were suspicious of pulmonary vascular disease; the diagnosis was pulmonary hypertension and bilateral branch pulmonary artery stenosis. After the inflammation settled, the stenotic bilateral branch pulmonary artery was reconstructed with a prosthetic vessel and the pulmonary pressure normalized immediately. A resected specimen revealed that the stenotic changes were from Takayasu’s disease. The patient’s postoperative course was uneventful, and pulmonary ventilation and blood scintigraphy returned to an almost normal range. At follow-up 5 years and 6 months after the operation, there was no evidence of pulmonary artery disease (e.g., stenosis and/or ischemia) or of any change in the central vessels of the retina, the so-called Takayasu’s retinopathy. (Circ J 2004; 68: 791–794)

Key Words: Pulmonary artery stenosis; Pulmonary hypertension; Pulmonary scintigraphy; Takayasu’s aortitis

In 1908, Takayasu first reported a case of a 21-year-old female with retinal arteriovenous anastomoses and although numerous cases of Takayasu’s disease have been reported since, the etiology of this disease has not been identified. It does occur predominantly in young women. Shimizu and Sano in 1951 described a ‘pulseless disease’ caused by occlusion of the principal arteries arising from the aortic arch and it was thought to be a nosologic entity among the various diseases within the aortic arch syndrome.

Case Report

A 63-year-old woman presented at hospital with dyspnea on exertion. The cardiothoracic ratio on her chest X-ray was 62%. There was nothing remarkable in her own or her family history and she was a non-smoker. Her chief complaint increased gradually and in August 1996, she was admitted to hospital.

On auscultation of the heart, a pansystolic vascular murmur was heard, but no other abnormalities were found on physical examination. The chest X-ray showed hyperlucency of the left lung and decreased vascular markings. The laboratory data were normal except for C-reactive protein (CRP), which was extremely high (13.1 mg/dl). Chest computed tomography scan showed inflammatory change in the left upper lung. Echocardiography showed that the right atrium and ventricle were dilated; the regurgitation of the tricuspid valve was estimated to be at the III/IV level and the right ventricular systolic pressure was estimated to be 84 mmHg. Based on the echocardiographic data, her condition was diagnosed as pulmonary hypertension and right cardiac ventricular failure. Pulmonary blood scintigraphy showed that the distribution of the left:right ratio of the lung blood was 6:94 (Fig 1A). Pulmonary thrombus and embolus were ruled out on consideration of her clinical course. When the ventilation scintigram showed less difference between the right and left lungs (37.6% and 62.4%), we also excluded Swyer-James syndrome (Fig 1B). We considered whether it was a congenital or acquired pulmonary arterial disease. Pulmonary angiography revealed focal stenotic lesions of 75% and 99% in the right and the left branches of the pulmonary artery, respectively (Fig 2).

There were no stenotic lesions in the peripheral pulmonary artery. Pulmonary arterial pressure was 95/15 (mean, 45) mmHg, right atrial pressure was 11 mmHg, and right ventricular pressure was 100/10 (mean, 45) mmHg. After admission, antibiotic therapy was started and her symptoms improved gradually until she was able to function without the need for more oxygen. We diagnosed that her right cardiac failure from the pulmonary hypertension would become worse if there was an infection, so the pulmonary artery stenosis operation was delayed until any infection would have completely cleared up. Hematological and blood chemistry data were within normal limits except for CRP, which had decreased to 3.3 mg/dl. A lung function test results were normal (vital capacity (VC): 2.02 L; %VC: 85.2%; forced expiratory volume in one second (FEV1.0): 1.52 L; FEV1.0%: 88.9%). Analysis of her clinical course and laboratory data suggested that she had a stenosis of the pulmonary artery related to Takayasu’s aortitis.

The operation was performed after the inflammation had subsided. Under a median sternotomy and standard cardiopulmonary bypass with ascending aortic perfusion and...
bicaval venous return, both branch pulmonary arteries were reconstructed with a vascular prosthesis without cardioplegic arrest. The vascular prosthesis was anastomosed to the peripheral native pulmonary arteries end-to-end, and anastomosed to the pulmonary artery truncus side-to-end (Fig 3). The stenotic lesions were confined to only 2 sections of the pulmonary artery and there were no more stenotic lesions in the peripheral pulmonary arteries.

Gross examination of the resected specimen showed remarkable thickening of the vessel wall (Fig 4). Mycobacterium tuberculosis was not found on Ziel-Neelsen staining of the resected specimen nor was the Mycobacterium gene detected in the DNA derived from the resected specimen. Microscopic examination showed that the necrotizing granulomatous angitis of the resected pulmonary stenotic lesions was incompatible with Takayasu’s aortitis (Fig 5). After the operation, her pulmonary artery pressure showed a dramatic return to normal levels (20 mmHg). No adverse event, such as life-threatening pulmonary edema, occurred after the pulmonary artery reconstruction. Postoperative angiography showed improvement of the bilateral branch pulmonary artery stenosis (Fig 6), and postoperative pulmonary ventilation and blood scintigraphy revealed no difference between the right and left lungs.

Her postoperative course was uneventful and she was discharged in January 1997, since when she has been followed up. Her pulmonary scintigraphy and retinal examination showed no abnormalities in February 2002.
In 1908, Takayasu, who was a Japanese ophthalmologist, first reported a 21-year-old woman with a peculiar condition involving an arteriovenous fistula of the central retinal artery. Subsequently, Shimizu and Sano in 1951 described ‘Pulseless disease’, which represented a syndrome caused by occlusion of the principal arteries arising from the aortic arch. This inflammatory disease of unknown etiology occurs predominantly in females in the second and third decades of life and has a worldwide distribution, although it is more common in the Orient. As it has been revealed that the arterial lesions are far more widespread and varied than originally expected, the disease is nowadays called by various names such as Takayasu’s disease, Takayasu’s arteritis, aortitis syndrome, Takayasu’s arteriopathy, and occlusive thromboaortopathy.

Pathologically, progressed Takayasu’s disease is characterized by chronic arteritis, which causes marked thickening and fibrosis of the intima, media and adventitia, with the most prominent changes occurring in the intima. However, the active phase of Takayasu’s arteritis is characterized by granulomatous vasculitis, leading to disruption of the media; a lymphocytic infiltrate is initially seen in the vasa vasorum of the adventitia and later stage in the media. Langhans’ and foreign body giant cells are often present in an autopsy case and the further reports that focused on pulmonary involvement in Takayasu’s disease alerted physicians and surgeons to the pulmonary artery lesions.

Lie reported that there are 3 distinct types of pulmonary...
vascular lesions in isolated pulmonary Takayasu’s arteritis;\(^8\) the classic large-vessel granulomatous giant cell arteritis, a peculiar type of organized thrombus with prominent recanalization and neangiogenesis, and plexogenic arteriopathy. Thus, the histopathologic findings of pulmonary Takayasu’s arteritis are distinctive and differ in many aspects from that of systemic Takayasu’s arteritis.

Many surgical treatments for occlusive lesions, other than pulmonary artery stenosis caused by Takayasu’s disease, have been reported\(^9\)–\(^11\) and recently non-surgical treatment by angioplasty with balloon and/or stent have become popular.\(^12\)–\(^15\) One study reported life-threatening pulmonary edema following stent implantation\(^16\) but only 1 case of angioplasty for a pulmonary arterial lesion caused by Takayasu’s disease has been reported by Sugiura et al in 1999.\(^17\) There are few reports of the surgical treatment of occlusive pulmonary arterial lesions related to Takayasu’s disease;\(^18\),\(^19\) the patient’s prognosis depends on the complications caused by aortitis and the cause of death is mainly congestive heart failure, acute myocardial infarction or cerebral vascular diseases.\(^20\),\(^21\) A long, careful post-operative follow up is essential for these patients.

References