Pulmonary Artery Stenosis due to External Compression by a Calcified Pericardial Band

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SUMMARY

A 60-year-old male with exertional dyspnea was referred to our hospital. Right pulmonary artery stenosis due to external compression by a calcified band was diagnosed by echocardiography, computed tomography, and magnetic resonance imaging. Percutaneous transluminal angioplasty was conducted in vain due to vascular recoil and failure of stent delivery. Pulmonary bypass grafting was performed successfully. The surgery indicated a probable etiology of chronic pericarditis. This is an extremely rare case of adult pulmonary artery stenosis without a known history of congenital disease, constrictive pericarditis, tuberculosis, or surgery. (Jpn Heart J 2004; 45: 527-533)

Key words: Pulmonary artery stenosis, Pericarditis, Calcification, Angioplasty, Bypass, Stent

PULMONARY artery stenosis in adults is uncommon and especially rare in cases without congenital heart diseases.1-4) Most of the cases were the result of incomplete pericardiectomy of chronic constrictive pericarditis.1,5-7) Some cases with congenital peripheral pulmonary artery stenosis were reported in adults.8,9) We experienced an adult pulmonary artery stenosis by an external calcified band. He had no history of constrictive pericarditis, cardiac surgery, or congenital heart diseases.

CASE REPORT

A 60-year-old Japanese man went to the nearby hospital with mild chest oppression experienced during jogging for the last several years. Echocardiography revealed abnormal blood flow in the left atrium in June 2002. Mild dyspnea on exertion gradually appeared. He was admitted to our hospital in order to
undergo further examination of the abnormal flow in the left atrium and dyspnea. He had a history of treatment for colon cancer, glaucoma, and hypertension, but not lung tuberculosis or pericarditis. He had no family members with a history of tuberculosis.

On examination the patient was a moderately nourished male with a blood pressure of 126/80 mmHg in both arms. All pulses were equal with a rate of 78/min. The first heart sound was normal. The second sound was physiologically split on inspiration and became a single sound on expiration. A grade 2/6 systolic ejection heart murmur was heard at the second intercostal space along the left sternal border. Chest radiography revealed a cardiothoracic ratio of 58% with mild protrusion of the left pulmonary artery without a calcified shadow. An electrocardiogram showed no significant changes. Complete blood cell count showed hemoglobin was 14.8 g/dL, white cell count 7840/µL, and a platelet count 37.8 x 10^4/µL. Serum chemistry revealed an alanine aminotransferase level of 24 IU/L, blood urea nitrogen of 10.4 mg/dL, C-reactive protein of 0.0 mg/dL, blood glucose of 113 mg/dL, and total cholesterol of 187 mg/dL. Blood gas analysis indicated a pH of 7.429, PaCO₂ of 42.6 Torr, PaO₂ of 66.0 Torr, and a base excess of 2.9 mmol/L. Other laboratory findings were normal. Transthoracic echocardiography demonstrated normal left ventricular contractility with a left ventricular diastolic diameter of 40 mm, left ventricular systolic diameter of 22 mm, and % fractional shortening of 46%. Color Doppler echocardiography detected abnormal flow receding from the lateral wall of the left atrium and grade 2/4 tricuspid regurgitation (Figure 1A). Qp/Qs calculated from left ventricular outflow and right ventricular outflow was 0.9, indicating no shunt abnormality. Transesophageal echocardiography detected increased flow in the left pulmonary veins compared to that in the right pulmonary veins (Figure 1B). The flow pattern was not turbulent but laminar, indicating an increase in blood volume or velocity on the left side. The cardiac catheterization data were as follows: pulmonary capillary wedge pressure (PC)(a/v/m) 13/10/8 mmHg, pulmonary artery (PA)(s/d/m) 48/11/23 mmHg, right ventricle (RV)(s/d/e) 45/0/6 mmHg, right atrium (RA)(a/v/m) 8/6/4 mmHg, left ventricle (s/d/e) 143/0/15 mmHg, aorta (s/d/m) 148/69/100 mmHg, cardiac output 5.29 L/min, cardiac index (CI) 3.07 L/min/m², and Qp/Qs, 1.03. A dip and plateau pattern was not observed. A coronary angiogram was normal. Right cardiac catheterization revealed mild pulmonary hypertension. We were unable to pass the Swan-Ganz catheter to the right pulmonary artery in the first cardiac catheterization. On pulmonary artery angiography, pulmonary artery stenosis was not obvious because of compression in the anteroposterior direction. Fascicular calcification overlapped the right main pulmonary artery. Chest-enhanced computed tomography and magnetic resonance imaging showed right pulmonary artery stenosis caused by a calcified band (Figures 2A, 2B, 3A, and
Figure 1. A: transthoracic aortic short axis color Doppler view shows an increased flow moving away from the left lateral wall in the left atrium. B: transesophageal transverse color Doppler view shows an increased flow from the left superior pulmonary vein. The arrows indicate the increased flow.

Figure 2. Contrast-enhanced computed tomography. A: Arrow indicates the calcified band compressing the right pulmonary artery. B: The calcified band is clearer one slice lower than that in A.
Lung perfusion scintigraphy showed decreased flow in the right lung. The perfusion ratio of right over left was 0.7. Lung ventilation scintigraphy was normal.

Percutaneous transluminal angioplasty (PTA) for pulmonary artery stenosis was attempted guided by intravascular ultrasound (IVUS). The stenosis was elliptic with a diameter of 4.5 x 1.3 mm. The systolic pressure gradient between the pulmonary trunk and the right pulmonary artery was 33 mmHg. The right pulmonary artery was dilated with an 8 mm-sized balloon but it was not dilated completely. Delivery of a Palmaz stent failed because of kinking of the guiding catheter. Surgical treatment was performed via a median sternotomy using cardiopulmonary bypass. Direct removal of the calcified band overlapping the right pulmonary artery was impossible because of severe adhesion and thickening of the whole anterior part of the pericardium to the heart. Pulmonary artery trunk-right pulmonary artery 10 mm-diameter polytetrafluoroethylene (PTFE) bypass graft surgery was performed to remove the pulmonary pressure gradient (Figure 4). Biopsy was performed from the pericardium just next to the calcified pericardial band. Biopsy specimens showed a thick fibrous calcified pericardium without inflammatory cells indicating old pericarditis. Cardiac catheterization revealed a right pulmonary artery pressure of 22/5/15 mmHg and a pulmonary trunk pressure of 40/10/20 mmHg (s/d/m) after the surgery. The left atrial abnormal flow had disappeared on the echocardiogram. The patient recovered uneventfully and was discharged with relief of the chest oppression and dyspnea on effort.
DISCUSSION

Pulmonary artery stenosis due to a calcified pericardial band is rare and was first described by McGaff, et al.1) Almost all previous cases had constrictive pericarditis.1,5,6,10) In several cases, after an incomplete pericardiectomy, a pericardial band constricted the main pulmonary artery just above its origin or the right ventricular outflow tract, producing a supravalvular, valvular, or infundibular pulmonary artery stenosis with right ventricular failure.1,5-7) Surgical resection of the fibrocalcified band usually abolished the pressure gradients and the right ventricular overload. Almost all of the cases of pulmonary artery stenosis due to calcification revealed the symptom accompanying constrictive pericarditis even without a past history of pericarditis. There is a report of right ventricular calcification causing pulmonic stenosis.11) The authors subdivided the cardiac calcifications into three etiologic categories, metastatic calcification, dystrophic calcification, and idiopathic calcification. Annular constrictive pericarditis with pulmonary artery stenosis was described in which an annular band formed a hoop around the pericardial sac at the level of the atrio-ventricular grooves.12) One with a family history of tuberculosis showed functional pulmonary, mitral, and aortic stenosis compressed by two bands of thickened pericardium that was relieved by
There are several reports of congenital peripheral pulmonary artery stenosis in children. However, congenital peripheral pulmonary artery stenosis in adults is especially rare. Kreutzer, et al reported 12 cases of peripheral pulmonary artery stenosis in adults. Their IVUS demonstrated marked medial thickening encroaching on the vessel lumen with maintenance of total vessel diameter in the stenotic site. There was no medial thickening in our IVUS finding. This suggests our case was not congenital. It is unusual that he has no history of constrictive pericarditis, tuberculosis, abnormal metabolism such as hyperparathyroidism, or surgery. The cause or origin of the calcified band was unknown until the surgery was performed. The site of the pulmonary artery stenosis is also rare because almost all of the pulmonary artery stenosis caused by a pericardial band was observed in the pulmonary trunk. Only one adult case with pulmonary artery branch stenosis due to pericarditis was reported. Our case may be the second adult case of pulmonary artery branch stenosis due to a calcified pericardial band. The pericardial band may be due to idiopathic pericarditis or an incomplete form of annular constrictive pericarditis.

The clue to the diagnosis in our case was an abnormal flow in the left atrium. The compensatory increase in blood flow in the left pulmonary vasculature induced abnormal jet flow in the left atrium, which was confirmed by a pulmonary blood flow scintigram. The calcification was not visible in the chest roentgenogram. Fluoroscopy during the cardiac catheterization showed abnormal calcification. A computed tomography scan revealed severe stenosis of the right pulmonary artery accompanied by a calcified band. In conjunction with magnetic resonance imaging, we concluded that the pulmonary artery stenosis was caused by the compression from outside the artery. IVUS was also useful to assess the characteristics and the vessel size of the lesion.

Concerning the treatment, PTA has been performed for congenital peripheral pulmonary artery stenosis. Recently, stents were implanted in some congenital pediatric cases although the long-term prognosis is not known. As for adult pulmonary artery stenosis, stent implantation is rare and a high restenosis rate was reported. Therefore, after the first attempt of PTA and stenting for the pulmonary artery failed, we referred the patient to surgical treatment. Complete angioplasty was also impossible even by surgery because of severe adhesion of the pericardium to the heart. The bypass operation relieved the pressure gradient and the patient's symptom although complete disappearance of the pressure gradient was not acquired, probably due to the relatively small-sized long graft.

This is a rare case of adult pulmonary artery stenosis due to a calcified pericardial band, which was diagnosed and treated by various imaging techniques and surgery.
REFERENCES