Case report

Severe pulmonary artery stenosis by a calcified pericardial ring

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Abstract

We present a rare case of a 44-year-old man with acquired pulmonary stenosis caused by a calcified pericardial ring. Chest computer tomography and cardiac angiography showed external compression of the main pulmonary artery by a calcified pericardial ring. A calcified pericardial ring was removed and the pulmonary artery angioplasty was performed for the stenotic pulmonary artery. The patient’s condition improved dramatically thereafter.

Keywords: Calcified pericardial ring; Pulmonary artery stenosis

1. Introduction

Acquired pulmonary artery stenosis (PS) is rare, and that caused by a calcified pericardial band even less common [1]. In most cases, this condition results from an incomplete pericardiectomy for chronic constrictive pericarditis. In this paper, we report an extremely rare case of acquired PS due to external compression by a calcified pericardial ring without constrictive pericarditis in a patient with a history of pulmonary tuberculosis.

2. Case report

A 44-year-old man was admitted to our hospital for dyspnea and swollen lower legs. He complained of weight gain and abdominal discomfort, and had been treated for pulmonary tuberculosis about 15 years ago. Blood pressure was 100/60 mmHg with regular heartbeat. A grade 4/6 systolic heart murmur was heard at the second left intercostal space. Bilateral pretibial pitting edema was present.

Electrocardiogram showed T wave inversion in II, III, aVF, and V1 ~ 6 leads. Chest roentgenogram revealed fibrotic consolidation of the right upper lung secondary to pulmonary tuberculosis, right ventricular (RV) hypertrophy, a calcified ring around the base of both great arteries, and a prominent left pulmonary artery (Fig. 1A). Echocardiography demonstrated supravalvular PS due to external compression, an elevated RV pressure of 98 mmHg, peak pressure gradient of 72 mmHg between the RV and the pulmonary trunk, and 3/4 grade tricuspid regurgitation. The ejection fraction was 38% and the systolic function of both ventricles was impaired. Chest computed tomography (CT) showed a calcified pericardial ring encompassing the aorta and the pulmonary trunk at the level of main pulmonary artery resulting in severe stenosis, and poststenotic dilatation of left pulmonary artery (Fig. 1B). There was no evidence of a thickened or calcified pericardium overlying the ventricular mass.

On cardiac catheterization, RA pressure (a/v/m) were 3/3/2 mmHg, RV (s/d/e) 95/0/3 mmHg, LV (s/d/e) 106/1/10 mmHg, and aorta (s/d/m) 126/68/85 mmHg. There was no pressure gradient between the aorta and the left ventricle, and no square root sign was observed on the pressure tracing. The catheter could not be passed through the main pulmonary artery due to severe stenosis at the calcified ring level (Fig. 1C). Coronary angiography was normal.

The operation was performed through a median sternotomy. The pericardium showed loose adhesions in the lower pericardium, while the upper pericardial reflection area covering the great arteries was firmly thickened, calcified, and adhered tightly to adjacent tissues. The pericardial calcification formed a complete ring surrounding the great vessels at their origins and adhered severely to the left main coronary artery. The main pulmonary artery was very tightly narrowed, and the distal main and left pulmonary arteries showed severe poststenotic dilatation. The pulmonary valve was intact. The calcified ring was removed with an on-pump beating heart (Fig. 2A). Most of the calcified ring was removed except for an area at the left main coronary artery, in fear of arterial injury. After excision of the calcified ring,
the stenotic main pulmonary artery was enlarged by angioplasty using glutaldehyde-fixed autopericardium. A portion of the dilated left pulmonary artery was excised and tricuspid annulus was repaired by De Vega’s method. The systolic pressure gradient between the pulmonary trunk and the RV by direct needle puncture was zero postoperatively.

The patient had an uneventful postoperative course. Histopathologic examination confirmed fibrosis and calcification without the presence of acid-fast bacilli. Postoperative echocardiography showed complete disappearance of pressure gradient without any residual tricuspid regurgitation. Postoperative chest CT also showed no calcification and no main PS (Fig. 2B). The patient was discharged on his eighth supravalvular pulmonary artery stenosis (*) due to compression by a calcified ring (arrow). The catheter could not be passed through the pulmonary artery.
postoperative day in a satisfactory clinical condition and had a good activity of NYHA I at his 12-month follow-up.

3. Discussion

Tuberculous pericarditis occurs in about 2% of patients with pulmonary tuberculosis [2], and is the most common cause of constrictive pericarditis in Korea. It may evolve from a hematogenous lesion formed at the time of primary infection or may result from lymphatic, contiguous, or even miliary spread, during the later stages of tuberculosis. Approximately 50% of patients with tuberculous pericarditis develop constriction, irrespective of treatment [2]. Although our patient had a history of pulmonary tuberculosis, there was no evidence of constrictive pericarditis. The constriction involved only a limited area of pericardium overlying both great vessels due to calcified ring formation.

Acquired PS, especially due to external compression by a calcified pericardial band or ring, is extremely rare, and was first reported by McGaff and colleagues in 1963 [1]. In most cases, a pericardial band was complicated by an incomplete pericardiectomy for constrictive pericarditis. It constricted the right ventricular outflow tract (RVOT) and resulted in the RVOT obstruction and RV failure [3—5].

Rarely, the constriction becomes localized, probably as a consequence of uneven pericardial involvement and fibrosis. In such cases, the annular calcified lesion shows a variety of compressive symptoms depending on the sites involved such as RVOT, pulmonary artery, and coronary artery [5—7]. In our case, the annular calcified ring uniquely encompassed both aorta and pulmonary artery.

Chest CT could help us to determine the obstruction site and differentiate between the extrinsic compression and the intrinsic stenosis. Although cardiac catheterization and angiography are less frequently used in this era of three-dimensional scan, they can be required to determine the site and severity of obstruction by quantifying pressures, and to identify concomitant cardiac disease such as coronary artery obstruction. Therefore, it is advisable to evaluate the lesion indubitably with echocardiography, cardiac catheterization, angiography, chest CT and MRI in patients who had the evidence of RVOT obstruction.

A calcified pericardial ring, once developed with symptoms, should be removed surgically irrespective of the presence of pericarditis. Surgical resection usually eliminates pressure gradients between the RV and the pulmonary trunk, and RV overload. Additional procedures such as coronary artery bypass graft, pulmonary artery angioplasty, or pulmonary artery bypass graft surgery, may be needed depending on the site and severity of obstruction.

Pericardial lesions must be removed completely because the inflammatory constriction may recur even after complete pericardiectomy, in cases of constrictive pericarditis [7]. In these cases, patients should be followed for many years to assure that the constrictive pericarditis does not develop.

References