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Surgical Correction of a Complex Aorto-Mitral Pathology

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Abstract

Pseudoaneurysm of mitral-aortic intervalvular fibrosa is a rare entity related to mostly infective endocarditis and surgical trauma of aortic valve. Its diagnosis may be missed following trans-thoracic echocardiographic assessment. Therefore, further imaging investigation such as transesophageal echocardiography and computed tomographic angiography may play a key diagnostic role. Here we present the successful surgical treatment of a 30-year-old male patient referred for surgical treatment of mixed severe calcific aortic valve disease and apparently without additional anatomical abnormalities.

Keywords: Pseudoaneurysm, Mitral aortic intervalvular fibrosa, Transesophageal echocardiography, Infective endocarditis

1. Introduction

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (P-MAIVF) is an anatomical abnormality of the fibrous body between the left coronary or the non coronary aortic cusp and the anterior leaflet of the mitral valve with concomitant communication into the left ventricular outflow tract. P-MAIVF is a rare entity related to mostly infective endocarditis and surgical trauma of aortic valve [1]. A few patients with P-MAIVF are asymptomatic in case of absence of complications (1). Diagnosis of P-MAIVF may be missed in trans-thoracic echocardiographic (TTE) assessment [2]. The aim is to present the successful surgical treatment of a patient referred for mixed aortic valve disease and apparently without additional anatomical abnormalities.

2. Case presentation

A 30-year-old male with severe symptomatic mixed aortic valve disease was referred to us for consideration of surgical treatment. He had been diagnosed three years earlier and kept under surveillance without history of infective endocarditis. Progressive deterioration of the clinical picture triggered surgical referral based on symptomatic and prognostic grounds. Trans-thoracic echocardiographic assessment showed a severe calcified aortic valve stenosis (mean pressure gradient 48 mmHg; aortic valve area 1.2 cm²), severe aortic regurgitation (PHT 190ms), aneurysmatic dilatation of the ascending aorta (50 mm) and relatively preserved left ventricular systolic function (55% ejection fraction). Therefore, a CT angiogram was performed for further anatomical evaluation. Unexpectedly, findings were consistent with a 18 × 31 × 29mm pseudo-aneurysmatic pouch arising from the aorto-mitral inter-annular zone and extending below the bifurcation of the left main coronary artery and the proximal part of the circumflex artery. The aortic root was dilated (45 mm) with significant calcification of the aortic annulus (Fig. 1) A conventional coronary angiography was performed which showed moderate
aortic insufficiency and with no evidence of any coronary artery disease.

2.1. Surgical intervention

Under general anesthesia, transesophageal echocardiography (TEE) examination was performed before the surgical intervention. A calcific cavity of P-MAIVF was identified as without any paradoxical movement during cardiac circle and there was a 20 mm diameter orifice of P-MAIVF towards the left ventricular outflow tract in the intraoperative TEE examination. In addition, the mild mitral annular calcification with extension of calcium into posterior leaflet without mitral regurgitation is seen in the TEE examination (Fig. 2). Surgical intervention was performed through median sternotomy and standard cannulation. The cross-clamp was applied and after achieving cardiac arrest with antegrade cold crystalloid cardioplegia and topical hypothermia, continuance of the arrest was ensured with intermittent antegrade cold blood cardioplegia. Operations were maintained under mild to moderate hypothermia (30°C–33°C) After aortotomy was performed, aorta and calcified aortic cusps were excised while preserving the coronary artery buttons. The orifice of the P-MAIVF was determined. The severe calcified pseudoaneurysm sac was observed during the surgical examination and this calcification spread towards the adjacent tissues. The orifice of the P-MAIVF closed using a Gore-Tex patch (WL Gore & Associates, USA) with continuous suture technique using 4-0 prolene suture (Figs. 3 and 4). After than, the Bentall procedure was performed with a mechanical valve prosthesis (27 mm St. Jude) and a 30 mm dacron graft. Cross clamping time was 172 min, and cardiopulmonary bypass time was 222 min. Completely closed sac orifice of P-MAIVF was seen the postoperative TEE examination (Fig. 5). The patient had an uneventful postoperative period and was discharged 10 days after the surgery.

3. Discussion

Pseudoaneurysm of mitral-aortic intervalvular fibrosa may lead to complications such as compression to vascular structures, thromboembolic events, fistula formation and rupture to pericardial cavity [2]. P-MAIVF Enlargement may lead to compression of adjacent structures including the left atrium, pulmonary artery and coronary arteries (most commonly circumflex artery). This situation may cause symptoms such as angina, pulmonary hypertension [1]. Perforation of the P-MAIVF may result in a fatal outcome. Therefore, timely surgical intervention should be performed in case diagnosis, especially in patient with high-risk for progression of P-MAIVF such as active endocarditis, diameter >3 cm, bicuspid aortic valve morphology, compression of adjacent structures (coronary or pulmonary artery) and presence of aortic regurgitation, fistula formation to cardiac chamber or aorta, thrombus formation in pseudoaneurysm [1–3]. In our patient, the pseudoaneurysm sac was existed under left main coronary artery bifurcation and the proximal part of the circumflex artery without any vascular compression in CT. However, the patient had a large P-MAIVF and mixed aortic valve disease.

Infective endocarditis, previous cardiac valve surgery, rheumatic fever or blunt chest trauma can
lead to MAIVF [1,2]. Calcific aortic stenosis and regurgitation was diagnosed three years ago. It has been previously hypothesized that patients with bicuspid aortic valves are more prone to this complication, likely because of congenital weakness in the area of the MAIVF due to avascular nature. In addition, it was suggested that aortic regurgitation may be an adding contributing factor for already compromised MAIVF region [4]. This pathology of the patient may be occurred due to previous infective endocarditis. However, the patient had no history of infective endocarditis. Therefore, we didn't find any predisposition factor for P-MAIVF.

Transthoracic echocardiography is the first imaging modality for evaluation of cardiac anatomy and function. However, further examination is suggested to determine the anatomic detail and precise relationship with the other cardiac structure [5]. It was found that while sensitivity of TTE was 43%, sensitivity of TEE was %90 in a study with small patient population [5]. There was only 90 cases were presented before 2009 in the literature. Interestingly, number of cases were doubled end of the 2012 [1]. This implies that the diagnosis of rare clinical conditions such as P-MAIF is more

Fig. 2. Intraoperative Transesophageal echocardiography (TEE).

Fig. 3. Intraoperative view of the mitral-aortic intervalvular fibrosa pseudoaneurysm after excision of calcific aortic valve.

Fig. 4. The pseudoaneurysm sac repaired with a Gore-Tex patch.
common with the technological developments in imaging methods. In an extended review showed missed diagnosis ratio of P-MAIVF was 42% in TTE examination despite technological advances [1]. The characteristic echocardiographic feature of a P-MAIVF is identified as a pulsatile echo-free space, expands in systole and collapses in diastole, at the level of MAIF [1,6]. In addition, blood flow is seen in to the cavity during systole, reverse blow flow is seen in to the left ventricular outflow track during diastole via color flow doppler interrogation [1,6]. First diagnosis of P-MAIVF of the patient was made via CTA examination. Intraoperative TEE confirmed the diagnosis and give further information about the pseudoaneurysm sac in real time imaging. Our interoperative TEE examination revealed that a calcifc posch of level of MAIVF without any paradoxical movement during cardiac circle. In addition, severe calcifc wall of the cavity is observed in intraoperative surgical examination. The patient may be misdiagnosed with trans-thoracic echocardiography due to the absence of paradoxical movement in the pseudoaneurysm and severe calcification.

In conclusion, P-MAIVF is a rare and potentially life-threatening condition. Paradoxic movement during cardiac circle of the pseudoaneurysm cavity is important in making the diagnosis. Severe calcification of the pseudoaneurysm pouch and its adjacent tissues may lead to misdiagnosis due to inability appearance the echocardiographic characteristics of the P-MAIVF as our case. Therefore, diagnosis of P-MAIVF should be kept in mind in severe calcification of the aortic valve and MAIVF region. Further evaluation with the use of other imaging techniques such as CT and TEE is required to identify P-MAIVF when severe calcification is seen on echocardiographic findings.

Author contributions
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