CASE REPORT

Bicuspid aortic valve endocarditis complicated by perivalvular abscess

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Abstract : A 37-year-old man presenting with fever and chest pain was admitted to our hospital. Electrocardiogram showed sinus tachycardia and complete left bundle branch block. Transthoracic echocardiogram showed infective endocarditis in the bicuspid aortic valve, complicated by multiple hyperechoic vegetations and severe aortic regurgitation. Blood cultures were negative and intravenous empiric antibiotic therapy was begun. However, fever lasted for 7 days and follow-up echocardiography revealed a newly emerged perivalvular abscess. The patient eventually underwent an urgent aortic root replacement that confirmed the echocardiographic findings. Our case report emphasizes that all patients with suspected aortic valve endocarditis should undergo early and follow-up echocardiographic studies. J. Med. Invest. 59 : 261-265, August, 2012

Keywords **:** *bicuspid aortic valve (BAV), infectious endocarditis (IE), perivalvular abscess (PVA), echocardiography*

INTRODUCTION

We present the case of a 37-year-old man with bicuspid aortic valve endocarditis complicated by a perivalvular abscess. He had a history of untreated heart murmur in childhood and untreated heart enlargement in recent years. We briefly discuss the main features of perivalvular abscess complicating infective endocarditis (IE).

Myocardial abscess formation is a severe complication of IE that is associated with high mortality. The aortic valve and its adjacent ring are more susceptible to abscess formation and complications of perivalvular extension of the infection than the mitral valve. Bicuspid aortic valve (BAV) endocarditis is also more frequently associated with these types of complications than the tricuspid aortic valve (1).

We report a case of BAV endocarditis complicated by perivalvular extension of the infection with abscess formation.

CASE PRESENTATION

A 37-year-old man presented to our hospital with episodes of persistent fever episodes (>38°C) and chest pain over the past 2 weeks. In addition, he had a medical history of untreated heart murmur 30 years previously, and untreated heart enlargement on chest X-ray for the past 5 years.

On physical examination he had an arterial blood pressure of 100/60 mmHg, regular tachycardia of 100-110 beats/min, and arterial saturation of 96%

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(room air). He had a systolic murmur, Levine 3/6, at the right cardiac base and a diastolic murmur at the left parasternal border, Levine 2/6. No crackles were audible in the pulmonary fields.

Chest X-ray showed significant cardiovascular enlargement, and 12-lead electrocardiogram (ECG) revealed sinus tachycardia with a mean rate of 100 beats/min and complete left bundle-branch block. He immediately underwent transthoracic echocardiography, which revealed mild near-circumferential pericardial effusion, a normal-sized left ventricle (LV) with concentric hypertrophy but normal systolic function, and a BAV with multiple hyperechoic vegetations. There was also severe aortic stenosis with a peak gradient of 80 mmHg and severe aortic regurgitation (2-3/4).

Laboratory data showed an increased white blood cell count (17,900 /μL) along with elevated C-reactive protein (11.84 mg/dL) and erythrocyte sedimentation rate (68 mm/hr). All eight blood cultures taken were negative (prior to hospitalization, levofloxacin 500 mg/day and ceftriaxone 1 g/day had been administered). Based on the clinical, laboratory, and echocardiographic findings, we made the diagnosis of culture-negative IE.

The patient received antibiotic treatment with gentamycin 1 mg/kg every 8 hours and ampicillin 2 g every 4 hours, according to the current guidelines of the Japanese Circulation Society. Seven days later, fever and C-reactive protein level had improved, but still remained moderately high (approximately 37°C and 5-6 mg/dL, respectively).

During this time, multiple follow-up echocardiographic studies were performed, 1, 4, and 8 days after the initial antibiotic administration (Fig. 1). The echocardiographic study on day 1 revealed a diffusely thickened aortic root wall and intramural multiple hypoechoic cavities near the aortic annulus. Follow-up studies on days 4 and 8 revealed that these cavities were generally enlarged and had fused with each other. Furthermore, we detected color Doppler signals inside of these cavities, which looked connected to the aortic lumen. Unfortunately, transesophageal echocardiography had difficulty detecting these cavities. To examine the cavities in greater detail, a multi-detector computed tomography (MDCT) scan was performed ; this showed a large abscess cavity that extended from the posterolateral aortic wall to the anterolateral side of aorta (Fig. 2).

The patient was referred for an emergency operation and underwent replacement of the aortic

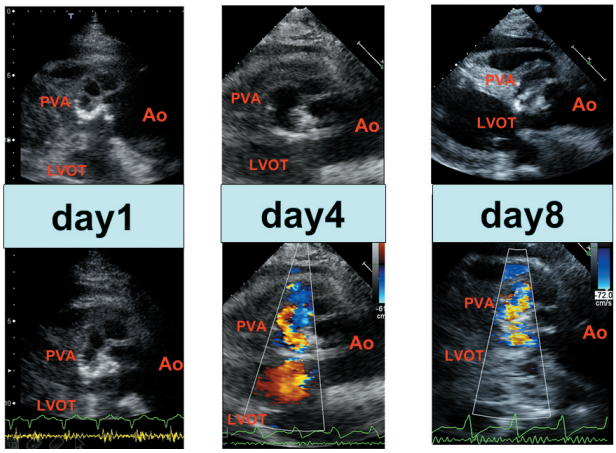

Figure 1. Follow-up transthoracic echocardiography with long-axis view focused on the region from left ventricular outflow tract to root of aorta, performed 1, 4, and 7 days after administration of empiric antibiotics. Upper figures show a bicuspid aortic valve with large echolucent perivalvular abscess formation ; cavities tended to be enlarged and fused with each other. Lower figures showing color Doppler signals inside these cavities, which appeared to be connected to the aortic lumen. Ao, root of aorta ; LVOT, left ventricular outflow tract ; PVA, perivalvular abscess cavity.

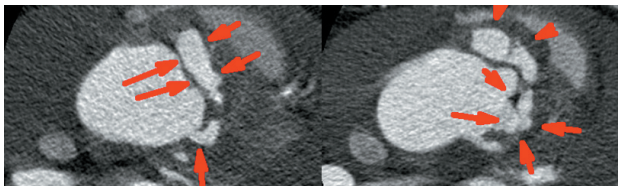

Figure 2. Multi-detector computed tomography (MDCT) axial view at the level of the aortic root. The figure shows a large abscess cavity extending from the posterolateral aortic wall to the anterolateral side of the aorta (red arrows).

root. The BAV showed severe degeneration, calcification, and vegetations. Almost half the circumference of the valve ring was completely absent, and both old and new thrombi-filled large abscess cavities were identified (Fig. 3). At the subaortic valve lesion, a small ventricular septal defect (VSD) (Kirklin type I) was found (it remains to be determined whether the VSD was congenital or an iatrogenic formation associated with operative procedure). We performed direct closure of the VSD, replaced the aortic valve (#27 Plima-Plus stentless bioprosthesis), and reconstructed the left ventricular outflow tract (with removal of abscess tissues, interposition of a vascular prosthesis (28 mm J-Graft Shield Neo), reconstruction of the coronary ostium with the Carrel patch technique).

Surgical pathology examination of the excised

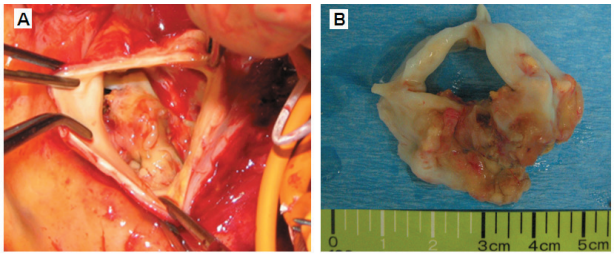

Figure 3A : Intraoperative photograph demonstrating bicuspid aortic valve involvement viewed through the aorta. After resection of the aortic valve, the abscess was seen involving the aortic root. 3B : Surgically excised bicuspid aortic valve. Gross view of bicuspid aortic valve showing coarse vegetations and calcific deposits.

valve showed two severely damaged semilunar leaflets with diffuse severe calcific deposits. On histologic examination, only severe calcification and clusters of Gram-positive cocci were detected. Cultures resulted in no bacterial growth and the causative microorganism was not identified. The patient had an uncomplicated postoperative course and there was no reactivation of infection.

DISCUSSION

There is increasing awareness that patients with BAVs are at high risk for cardiovascular complications such as aortic valve dysfunction (2-6), IE (5), ascending aortic aneurysm (6-8) and aortic dissection (9, 10). As BAV is one of the most common congenital cardiac abnormalities (11-13), the frequency of cardiac and vascular events associated with BAV might be greater than the combined cardiovascular morbidity of all other congenital heart diseases (2). High rates of cardiovascular adverse events in adults with BAV have been recently reported by two large-scale studies (14, 15).

Previous small-scale necropsy and surgical series have suggested that 25% to 50% of all infected aortic valves were bicuspid (16, 17). In a study by Lamas and Eykyn, BAV IE represented 12% of patients with native valve IE (5). In a cohort of 310 patients with aortic valve IE (1), a BAV was identified in 16% of cases. In a study including 642 adults with BAV, IE occurred in 13 patients over 9 years of follow-up (15), while in another study, 4 out of 212 patients with BAV developed IE during a follow-up of 20 years (14). A recent multicentre study (1) demonstrated that of 148 patients with BAV, 4(2.7%) were diagnosed with IE. All four patients had significant regurgitation and underwent aortic valve replacement. These observations suggest that the risk of IE in adults with BAV is relatively low, whereas BAV is common in cases with confirmed aortic valve IE.

Our case of BAV IE was complicated by perivalvular abscess and eventually required surgical intervention. A perivalvular abscess was identified in 50% of the BAV IE patients in the study by Tribouilloy *et al.* (1) compared with 30% in the study by Lamas and Eykyn (5). Periannular complications were observed in 14 of the 22 patients with BAV IE (64%) in the report by Kahveci *et al.* (18). Moreover, Tribouilloy *et al.* found that the presence of a BAV was independently associated with abscess formation in aortic valve IE (1). This finding is important as patients with aortic valve IE complicated by perivalvular abscesses are more likely to undergo surgery (19), which is consistent with the present case.

Early identification of abscesses is particularly important since it has been shown that surgery may improve outcomes by preventing widespread tissue destruction. Daniel *et al.* (20) studied 44 patients with proven abscesses ; 28.3% of abscesses could be identified using transthoracic echocardiography while 87% were diagnosed with the transesophageal approach. In a larger-scale series (21), investigators identified 36% of abscesses using the transthoracic technique and 80% using the transesophageal approach. Therefore, current European guidelines on the prevention, diagnosis, and treatment of infectious endocarditis from 2009 (22) recommend transesophageal echocardiography(TEE) in most adults with suspected IE, even in the presence of a positive transthoracic echocardiography(TTE) exam, due to its higher sensitivity and specificity for the diagnosis and assessment of intracardiac lesions (class IIa, level of evidence C). However, some previous reports have indicated that TTE is an accurate method for the diagnosis of abscesses associated with IE, especially in patients without prosthetic valves (23). In addition, recent technological innovation has rendered TTE more sensitive for the diagnosis of IE. In the present case, TTE very clearly revealed a diffusely thickened aortic root wall and intramural multiple hypoechoic cavities near the aortic annulus. We performed TTE frequently to monitor the progression of IE lesions and found that cavities tended to grow larger and fuse with each other. TTE is superior to TEE for frequent follow-up and detection of lesion progression so as to determine the appropriate time for surgery, in a

case that clear images can be acquired by the transthoracic echocardiography. Furthermore, TEE is not without risks. A previous report described a patient with impending rupture perivalvular abscess who died during a TEE study (24). Thus, TEE is not completely safe in these situations and is not suitable for frequent follow-up of perivalvular abscesses.

In the present case, MDCT was also useful for examining the cavities in detail. In a recent study (25) using a per-valve-based analysis, MDCT had a sensitivity of 96% and a specificity of 97% for detecting vegetations and abscesses/pseudoaneurysms, compared with 100% and 100%, respectively, for surgery. The accuracy of MDCT did not differ significantly from TEE, but did provide better anatomic information than TEE in terms of perivalvular involvement (i.e., myocardial, pericardial, and coronary sinus regions), which was helpful for planning surgery.

Patients with identified abscesses associated with IE are usually candidates for more aggressive therapy, including early surgery, than those without an abscess. It is important for the surgeon to know whether a perivalvular abscess is present because if it is, the operative procedure is more complicated and is also associated with higher morbidity and mortality than in operations for simple valve replacement or repair for active IE (26). It has been shown that endocarditis caused by *Staphylococcus aureus* and other virulent microorganisms on valves in the left side of the heart are best treated with early surgery (26-28). In a large merged database on native valve endocarditis, the overall mortality was higher in patients with *Staphylococcus aureus* endocarditis than in those with other bacteria (20% vs. 12%, $p=0.001$), but surprisingly, fewer patients infected with *Staphylococcus aureus* had surgery (26% vs. 39%, $p=0.001$) (27). In the present case, all the blood cultures were negative, probably due to empiric prehospital antibacterial treatment, and thus provided no useful information preoperatively.

CONFLICT OF INTEREST

No conflict exists for each author.

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