An Adolescent With Aortic Regurgitation Caused by Behçet's Disease Mimicking Endocarditis

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Aortic regurgitation is a serious complication of Behçet's disease. We report a 17-year-old male with severe aortic regurgitation caused by Behçet's disease. An early diagnosis led to the immediate start of immunosuppressants followed by successful valvuloplasty with autologous pericardium.

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Behçet's disease (BD) is a chronic multisystemic inflammatory disorder of unknown etiology with high incidences in Asian, Middle Eastern, and Mediterranean populations [1]. Aortic regurgitation (AR) in BD is a rare complication and standard surgical techniques have resulted in recurrent valve dehiscence due to fragile aortic structures and inflamed tissue caused by systemic vasculitis [2]. A timely diagnosis is essential for proper treatment to reduce postoperative morbidity and mortality [2, 3]. Herein, we report an adolescent male with severe AR treated with immunosuppressants before receiving surgical valvuloplasty with autologous pericardium under the suspicion of BD.

A 17-year-old male was referred for infective endocarditis in the aortic valve (AV). Other than his right great toe-second toe syndactyly, he had previously been healthy.

Nine days prior, the patient was admitted to another hospital for syndactyly repair. Preoperative electrocardiogram showed left ventricular hypertrophy; therefore, an echocardiography was done revealing suspicious vegetation on the aortic and mitral valve with severe AR. Suspicious for infective endocarditis, intravenous antibiotics were immediately administered. Both initial and follow-up blood cultures reported no bacterial growth.

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For further treatment, this patient was transferred to our hospital. Upon arrival, the patient presented without symptoms. His blood pressure was 113/49 mm Hg, heart rate 101 beats per minute, and body temperature 36.9°C. A grade 2/6 diastolic murmur was auscultated. Labs showed no leukocytosis, a C-reactive protein of 0.68 mg/dL, and elevated erythrocyte sedimentation rate (ESR) of 77 mm/hour. Echocardiographic findings revealed oscillating redundant tissue with shaggy surface on the left coronary cusp, prominent marginal thickening of all aortic cusps, and coaptation failure (Fig 1). Redundant tissue on the mid-portion of the anterior mitral leaflet was seen with severe AR and markedly dilated left ventricle (end-diastolic dimension: 68.2 mm). Considering the characteristic vegetation-mimicking oscillating mass on the AV and elevated ESR without evidence of infection, we suspected cardiac involvement of a systemic inflammatory disease.

Although the patient had recurrent oral ulcers, there was no history of skin lesions, genital ulcers, or uveitis. Pathergy test was negative, but human leukocyte antigen-B51 was positive. Despite failure to fulfill the diagnostic criteria of BD, clinical manifestations and echocardiographic findings strongly suggested cardiac involvement of BD. His elevated ESR indicated active disease state; therefore, we prescribed oral prednisolone 60 mg/day starting the 8th hospital day. With decreased ESR of 29 mm/hour on the 21st hospital day, we performed aortic valvuloplasty on the 29th hospital day (Fig 2).

Under conventional cardiopulmonary bypass, a hockey stick incision of the ascending aorta was made. Ascending aorta and root were preserved and aortic annulus was measured to be 24 mm. There was a small ulcer-like lesion just above the left main coronary os. The left coronary cusp was perforated and showed thickening with big fragile redundant fibrous tissue tags. Other cusps had thickened enrolling leaflet coaptation margins with huge central regurgitant gaps (Fig 3A) partially resected for cusp extension. The leaflets were designed with glutaraldehyde-fixed autologous pericardium for each cusp extension. The designed leaflets were sutured from the base of the leaflet, 3-mm higher than the usual commissure. Then reinforce sutures were done. On the mitral valve, there were 2 pouch-like leaflet defects without perforation and a fibrous tissue tag on the anterior leaflet. The defects were repaired with fixed autologous pericardium through the AV. The aortic clamping and cardiopulmonary bypass time was 110 and 162 minutes, respectively.

The histopathology of the resected AV confirmed no evidence of infection. Microscopic findings showed fibromyxoid valvulopathy without leukocyte infiltration (Fig

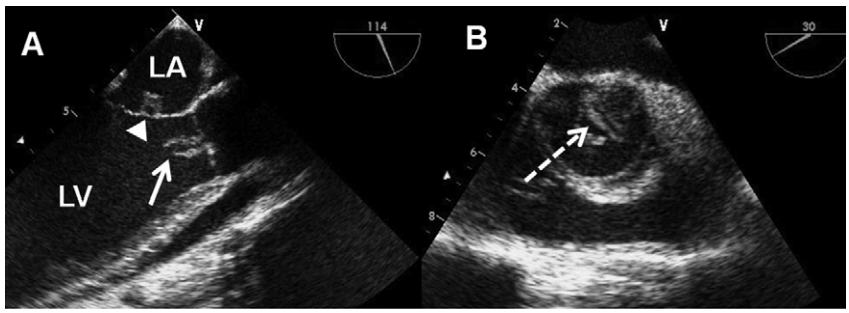

Fig 1. (A, solid arrow) Preoperative transesophageal echocardiography. Oscillating redundant tissue with shaggy left coronary cusp surface and prominent marginal thickening on all aortic cusps. (B, dotted arrow) Coaptation failure. (A, arrowhead) Redundant tissue at the mid portion of anterior mitral leaflet. (LA = left atrium; LV = left ventricle.)

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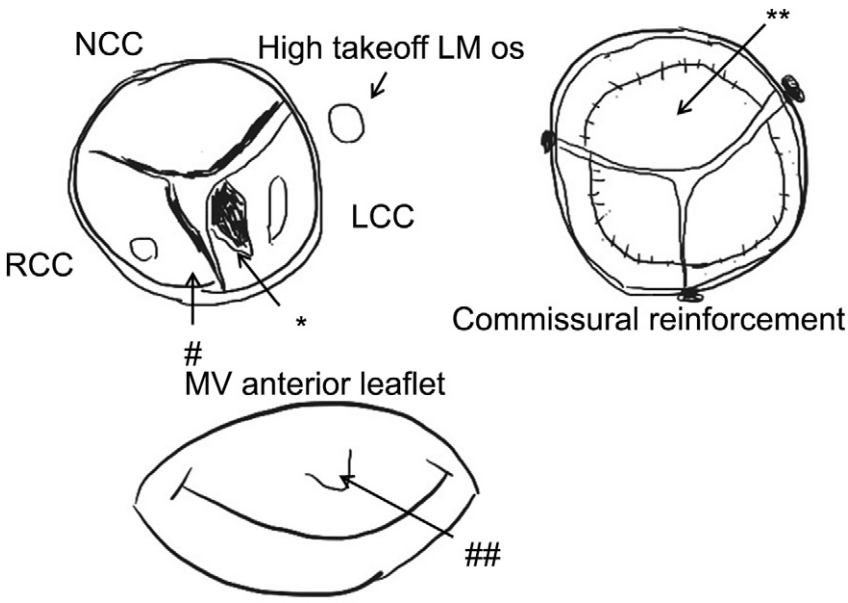

*Fig 2. Operative findings and procedures. The thickened aortic left coronary cusp (LCC) was perforated. Right and noncoronary cusps (RCC, NCC) had thickened enrolling leaflet coaptation margins and huge central regurgitant gaps, partially resected for extension. Cusp extensions for all 3 leaflets were designed with autologous pericardium. (LM = left main; LV = left ventricle; MV = mitral valve; * = perforation with big fragile redundant fibrous tissue tags; ** = leaflet extension with autologous pericardium; # = thickening enrolling leaflet coaptation margin; ## = defect with fibrous tissue tag.)*

3B). Two months post operation, transthoracic echocardiography (Fig 4) showed trivial AR with good leaflet motion and decreased left ventricular end-diastolic dimension (50 mm). Low-dose prednisolone was maintained with follow-up ESR of 2 mm/hour.

Comment

Cardiovascular complications related to aortic root dilatation and AR are the main causes of death in patients with BD [4], and early diagnosis is crucial for initiating immunosuppressive therapy to improve treatment outcome [2, 5]. Han and colleagues [3] showed that patients required more frequent surgical treatment for AR before a proper diagnosis of BD was reached.

The diagnosis of BD by the International Study Group for Behçet's Disease (ISG) criteria is based on common clinical symptoms [6], but because there are no pathognomonic symptoms or laboratory findings [1], concerns have been raised that the current diagnostic criteria may delay the diagnosis of BD [3]. In the study by Jeong and colleagues [2], only 8 of 19 patients fulfilled the ISG criteria, but with typical echocardiographic and surgical findings, patients were later diagnosed with BD. Although our patient lacked symptoms to fulfill the ISG criteria, his characteristic echocardiographic findings, together with elevated ESR and human leukocyte antigen-B51 positivity, led to the suspicion of BD [2, 3]. His surgical findings were consistent with previous reports of AR in BD; perforated left coronary cusp of the AV with fragile redundant fibrous tissue, thickened right and noncoronary cusp leaflet coaptation margin, and ulcer-like pouch above the left main os. Although methods such as aortic root or aortic valve replacement have been successfully employed in such patients [2], we specifically performed valvuloplasty of the AV with autopericardium. This was done to avoid the well-known valve dehiscence from AV replacement, to get better tissue healing, and to lower the risk of thromboembolism. The questionable long-term durability of the extensions and possibility of leaflet dysfunction are potential disadvantages of this technique over replacement [7].

To avoid repetitive operations, postsurgical inflammation is recommended to be kept low [2, 4, 8]. Therefore, it is important to maintain effective postoperative immuno-

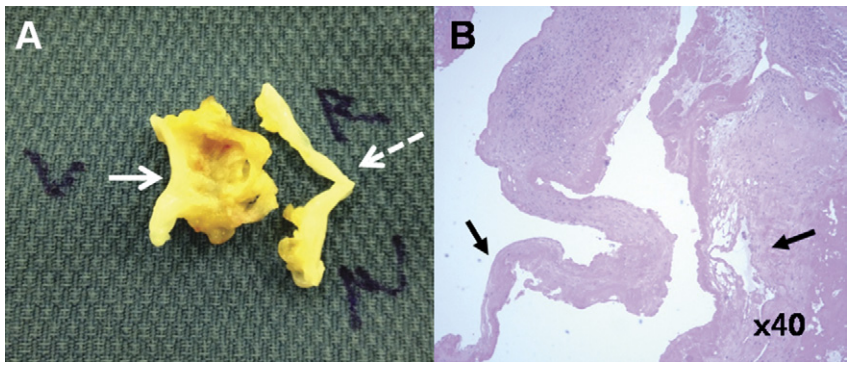

Fig 3. Valve tissue and pathologic findings. (A) Fragile redundant fibrous tissue tags on the thickened left coronary cusp (A, solid white arrow). Right and noncoronary cusps had thickened enrolling leaflet coaptation margins (dotted white arrow). (B) Pathologic findings revealed fibromyxoid change of the aortic valve (black arrows) (Hematoxylin-eosin, magnification ×40).

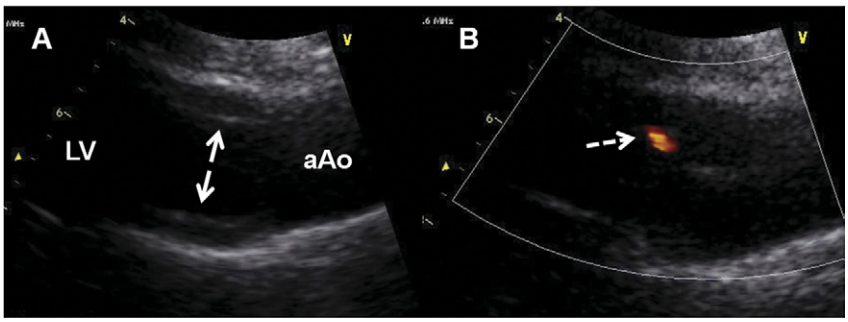

Fig 4. Postoperative transthoracic echocardiography. (A, solid arrow) Good leaflet motion and (B, dotted arrow) trivial aortic regurgitation 2 months after operation. (aAo = ascending aorta; LV = left ventricle.)

suppressant therapy while monitoring serum ESR and C-reactive protein levels [2].

In conclusion, severe AR from AV destruction associated with BD can occur even during adolescence. Early diagnosis with immediate initiation of immunosuppressants is important in the postoperative course of patients with valve problems from BD. We experienced good short-term results from successful AV valvuloplasty with autologous pericardium, although long-term follow-up is necessary.

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