Infective Endocarditis Associated with Quadricuspid Aortic Valve

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SUMMARY

A 26-year old Japanese woman experienced new aortic valve regurgitation associated with a preceding high fever of unknown cause. During the fever episode, although bacteremia or fungemia was not evident despite frequent blood cultures, intravenous panipenem/betamipron (PAPM/BP) gradually resulted in decline of the fever. Echocardiography and operative procedures revealed a quadricuspid aortic valve (QAV), which was composed of two equal larger cusps and two unequal smaller cusps (type f). A smaller accessory cusp was damaged but showed no active vegetation. A Medtronic Freestyle bioprosthesis was implanted using a subcoronary technique. Although the risk of endocarditis for this rare valve abnormality is not well documented, the present case may support the conventional assumption that patients with unequal small cusps are prone to endocarditis. (Jpn Heart J 2003; 44: 441-445)

Key words: Heart failure, Aortic valve regurgitation, Echocardiography, Freestyle bioprosthesis

ISOLATED four-cusped (quadricuspid) aortic valve (QAV) is a rare congenital anomaly and the potential clinical course has remained unclear because of difficulty in their detection before necropsy or aortic valve replacement. QAV is classified into seven types (type a to g) according to the size variation of the four cusps,1 which might be associated with the susceptibility to infective endocarditis.2 To the best of our knowledge, however, few cases of endocarditis involving QAV have been reported.1,3,4 We report a case of unequally distributed QAV associated with endocarditis, resulting in an abrupt onset of aortic valve regurgitation.

CASE REPORT

A 26-year old Japanese woman took a trip to Southeast Asia at the end of August 2001. Three weeks later she was admitted to a local hospital because of...
persistent high fever of unknown cause. She had a history of atopic dermatitis and bronchial asthma, and a female sibling had congenital ventricular septal defect. Physical examination showed clear heart sounds without audible murmurs. On admission, the electrocardiogram was normal. The cardiothoracic ratio on chest x-ray was 38% and the lungs were clear. Her temperature rose daily to as high as 39°C with shaking chills, and the cause remained unknown despite intensive investigations. Blood smears for malaria parasites were negative and C-reactive protein increased to 17.5 mg/dL. Although blood and cerebrospinal fluid cultures for bacteria were negative, intravenous panipenem/betamipron (PAPM/BP) administration resulted in a gradual decline of fever over a week, during which time she developed exertional and nocturnal dyspnea.

She was referred to a previous public (or community) hospital in the middle of November. On physical examination, a grade 4/6 cardiac diastolic blowing murmur was heard together with a gallop rhythm. Inspiratory rales were heard at both lung bases. Her blood pressure was 122/35 mmHg and brain natriuretic peptide was 936 pg/mL. An electrocardiogram revealed sinus tachycardia of 104 beats/min, with mitral P wave in the chest leads and ST-T segment depression in leads V5 and V6. Chest X-ray films showed a marked increase in the cardiothoracic ratio of 54% and pulmonary congestion. Echocardiography showed severe aortic regurgitation with a dilated left ventricle (end-diastolic diameter; 59 mm) and lowered systolic function (ejection fraction; 49%). The acute onset of aortic regurgitation together with a preceding high fever indicated a probable diagnosis of infective endocarditis.

Her New York Heart Association functional class IV symptoms remained even after her temperature had decreased, and she was transferred to our hospital for aortic valve replacement at the end of December. Transesophageal echocardiography showed an intracardiac ‘string-shaped’ piece of tissue six mm in length that appeared to be attached to a smaller accessory cusp between the noncoronary and right cusps (Figure 1A-C). The tissue was fluttering into the left ventricle during diastole, with a severe regurgitant jet originating from the fourth space arising from the small accessory cusp (Figure 1D). A quadricuspid aortic valve with a ruptured accessory cusp was strongly suspected, but the presence or absence of vegetation could not be confirmed. During surgery, the aortic valve had 4 definite cusps, two equal larger cusps and two unequal smaller cusps (type f). A smaller accessory cusp was located between the larger noncoronary cusp and a smaller right-sided cusp, and its free margin was almost completely ruptured. Fluttering of the free margin of the ruptured accessory cusp probably gave rise to the appearance of the “string-shaped” tissue on the echocardiogram. The structure was assumed to be the result of extended perforation. Active valvular vegetation and other local tissue destruction were not seen. Aortic valve replace-
ment was performed successfully with a 21-mm Medtronic Freestyle bioprosthesis, which we selected in consideration of her desire to have children in the future.

**DISCUSSION**

Isolated quadricuspid aortic valve (QAV) is a rare congenital abnormality with an estimated incidence of between 0.003% and 0.013%. Most cases of QAV were detected as an incidental finding at necropsy or during aortic valve replacement. Recently, the usefulness of noninvasive echocardiography in demonstrating QAV has been reported, however, the typical natural history and the exact risk of endocarditis, which might be associated with the size variation of the four cusps, have remained unclear.

**Figure 1.** Transesophageal Echocardiography. **A:** Diastolic frame shows four unequally distributed aortic cusps. A damaged smaller cusp (arrow) is located at the position of the conventional right aortic cusp. **B:** Systolic frame shows the four cusps in the open state. **C:** Ruptured cusp (arrow) is fluttering into the left ventricle (LV) during diastole. **D:** Color Doppler examination shows a severe aortic regurgitation (AR) jet originating from the area. A and B were imaged at a plane angle of 47°. C and D were imaged at a plane angle of 139°. LA = left atrium; RA = right atrium; RV = right ventricle; PA = pulmonary artery.
Prophylaxis against infective endocarditis might be required in patients with a small accessory cusp because of an unequal distribution of stress and abnormal leaflet coaptation that may contribute to the progression of aortic valve regurgitation. In contrast, patients with cusps of nearly equal areas had trivial or mild aortic valve regurgitation and might have no increased risk of endocarditis. On the other hand, Matsukawa, et al reported the case of a 75-year-old senile man with four equal-sized aortic cusps (type a), who developed bacterial endocarditis caused by \( \beta \)-streptococcus. Suda, et al reviewed 17 Japanese patients with QA V and predicted that they might have progressive sclerocalcified changes in later life, which usually result in increasing aortic valve regurgitation and sometimes aortic valve stenosis. Although these degenerative changes might be associated with conditions predisposing infective endocarditis, in the present case the 26-year-old young woman with two unequal smaller cusps had endocarditis, and histopathological examination of the cusps showed myxoid degeneration without calcification. This observation might support the hypothesis that patients with a small accessory cusp are prone to develop bacterial endocarditis.

Preoperative transesophageal echocardiography successfully depicted that the left and right coronary arteries arose from each of the two conventionally located cusps. In the past, coronary angiography was required to detect other possible cardiac abnormalities such as anomalies of the coronary ostium and coronary arteries, but now color flow Doppler echocardiographic images can be used as a substitute to assess the anatomical variation of such a valve. In the present case, the pulmonary valve could not be fully evaluated, although a quadricuspid pulmonary valve is ten-fold as frequent as the QA V.

We report a case of infective endocarditis associated with unequally distributed QA V (type f). Whether the four-cusped morphological variety contributes to susceptibility to endocarditis in the natural history should be further investigated. Cross-sectional echocardiography and color flow Doppler images are useful for such investigations.

REFERENCES