QUADRICUSPID AORTIC VALVE ASSOCIATED WITH SEVERE AORTIC REGURGITATION

MASAHIKO MATSUMOTO, M.D., SHIGEHICO MIKI, M.D., KENJI KUSUHARA, M.D.
YUICHI UEDA, M.D., YUTAKA OHKITA, M.D., TAKAFUMI TAHATA, M.D.
AND MASASHI KOMEDA, M.D.

A 57-year old man with severe aortic regurgitation was found to have a quadricuspid aortic valve. Aortic valve replacement was performed successfully with a St. Jude Medical prosthetic valve. On histological examination, the resected cusps showed fibrotic thickening with calcification.

AMONG the various congenital cardiac malformations of the semilunar valves, a bicuspid aortic valve is relatively common. However, a supernumerary aortic valve is extremely rare. We report a case of quadricuspid aortic valve with severe regurgitation corrected surgically with a prosthetic valve.

CASE REPORT

In October 1981 a 55-year old man was hospitalized with palpitation, nocturnal dyspnea and orthopnea. The patient had had increasing shortness of breath for eight years without a previous history of rheumatic fever. Cardiac catheterization was performed in November 1981. Right heart catheterization showed normal pressure. Left heart catheterization showed no aortic valve stenosis and a left ventricular end-diastolic pressure of 11 mmHg. Aortic root angiography revealed grade 4 aortic valve regurgitation (Fig. 1). Left ventricular angiography showed grade 2 mitral valve regurgitation. The left ventricular end-diastolic volume index was 316 ml/m², the end-systolic volume

index, 220 ml/m², and the ejection fraction, 0.30. An echocardiogram showed marked enlargement of the left ventricle, decreased ejection fraction and a coarse fluttering of the mitral valve. The coronary arteries appeared normal.

In October 1983 the patient was admitted for aortic valve replacement. Functionally he was in class III (NYHA). His blood pressure was 130/68 mmHg and the heart rate was regular, 67 per minute. On auscultation, there was a grade 3/6 blowing diastolic murmur along the left sternal border and a grade 3/6 blowing systolic murmur at the apex. The liver was not enlarged. Chest X-ray showed moderate cardiomegaly, with a cardiothoracic ratio of 0.64. His electrocardiogram showed sinus rhythm with ventricular premature contractions and left ventricular hypertrophy. Echocardiographic findings were the same as before. Catheterization was not repeated.

At operation, the aortic valve was found to have four cusps, three equal-sized cusps and one smaller cusp. The supernumerary cusp was situated between the right and noncoronary cusps. Two small fenestrations were seen in this cusp. All four cusps were moderately thickened and retracted (Fig. 2). The cusps were excised and replaced with a 27 mm St. Jude Medical

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Department of Cardiovascular Surgery, Tenri Hospital, Nara, Japan

Mailing address: Masahiko Matsumoto, M.D., Department of Cardiovascular Surgery, Tenri Hospital,
200 Mishima-cho, Tenri, Nara 632, Japan

Fig.1. Aortic root angiography showing severe aortic regurgitation (grade 4).

Fig.2. The resected four cusps of the aortic valve. Arrow indicates the supernumerary cusp. L = left coronary cusp; N = noncoronary cusp; R = right coronary cusp.

prosthesis. Histological examination of the resected cusps showed fibrotic thickening with calcification (Fig. 3).

His postoperative course was protracted with delayed cardiac tamponade and persisting left heart failure. He was discharged on the 60th postoperative day and is doing well six months postoperatively.

DISCUSSION

Quadricuspid aortic valves are rather common in semilunar valves of the truncus arteriosus. However, in patients with normally developed

Fig.3. Histopathological section of a cusp showing fibrotic thickening with calcification. (hematoxylin and eosin stain. $\times 40$).

great arteries, it is a rare congenital cardiac malformation. We found no case of quadricuspid aortic valves among 1941 necropsies at Tenri Hospital. Simonds reported an incidence of 0.008 percent in a review of the literature (2 cases in 25666 postmortem examinations¹). Our case was diagnosed incidentally at operation for aortic valve regurgitation. Most of the reported cases have been autopsy findings². Reviewing the literature, we found 9 cases in addition to our own case which were corrected surgically; eight of them received a prosthetic valve or homograft and one was treated with aortic valvuloplasty³. Although the quadricuspid aortic valve usually functions normally in infants or young children, valvular regurgitation occurs frequently in adults⁴. In our case, fibrotic thickening and retraction with calcification were observed. However, the exact cause of the regurgitation in the quadricuspid aortic valve is not known.

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