Endocarditis complicating a congenital quadricuspid aortic valve

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The most common aortic valve congenital abnormality is observed in bicuspid aortic valve. Only a few cases of aortic valve quadricuspidy have been reported in the literature. We report a new case of endocarditis complicating a congenital quadricuspid aortic valve.

Case report

A 68-year-old male with a history of hypertension and angina was referred to our hospital for a septic spondylodiscitis after complaining of inflammatory back pain and chronic fever. Physical examination revealed a 3/6 diastolic murmur and no signs of heart failure. Numerous blood cultures were positive to *Streptococcus oralis*. Transesophageal echocardiography short-axis view demonstrated a quadricuspid aortic valve with fully opening four equal leaflets (Figure 1). Additionally, transesophageal echocardiography detected multiple vegetations involving all four leaflets (Figure 2). Destruction of the aortic valve was evidenced by a severe aortic regurgitation documented from the transesophageal echocardiography long-axis view (Figure 3). No other cardiac structural abnormalities were found. Anti-biotic therapy was instituted parenterally by ampicillin plus gentamycin based on susceptibility tests. After three weeks of treatment, the patient was operated because of significant aortic valve deterioration and left ventricular enlargement as demonstrated by transthoracic echocardiography. Intra-operatively, the surgical view corroborated the ultrasound findings (Figure 4). The quadricuspid aortic valve presented with multiple vegetations on each leaflet. A prosthetic aortic valve was employed to replace the defective valve. The patient was discharged after six weeks of additional antibiotic treatment with no evidence of active infectious or prosthetic aortic valve dysfunction.

Discussion

The quadricuspid aortic valve has been recognized as a rare congenital abnormality accounting for less than 0.001% of the population at autopsy. The aortic valve normally forms from the embryonic truncus arteriosus. Abnormalities in this area will lead to the development of a quadricuspid aortic valve and others coexisting congenital cardiac defects implicating the coronary arteries, the left ventricular outflow tract, the pulmonary valve or the inter-atrial septum. Aortic valve quadricuspidy is frequently misdiagnosed by transthoracic echocardiography necessitating the transesophageal approach. The quadricuspid aortic valve is usually composed of four unequal-sized leaflets with a fixed orientation of the commissures (anterior–posterior and horizontal commissures). The coronary arteries arise from the two opposite anterior-right and posterior-left leaflets. The quadricuspid aortic valve with four equal-sized leaflet found in our patient is less frequent and corresponds to type ‘a’ in the Hurwitz and Roberts’ classification. With degeneration of aging valve, sclerosis and calcification can occur. Incidence of hemodynamic abnormality is not documented but aortic regurgitation seems to be more prevalent than aortic stenosis.

The importance of quadricuspid aortic valve as a potential substrate for infective endocarditis is not known but could
be related to the morphology of the valve. Usually, patients with infective endocarditis have unequal-sized leaflets. The asymmetrical morphology of the cusps is a predisposing condition to turbulences, thus infective endocarditis as described in aortic bicuspid valve. Quadricuspid aortic valve with equal-sized leaflets is infrequently associated with infective endocarditis. We report the third case in the literature. The clinical presentation of quadricuspid aortic valve endocarditis does not differ from common acute infective endocarditis. Here, spondylodiscitis resulted from embolic complication. Multiple vegetations were the only echocardiographic predictor of systemic embolisation in our patient. We did not see large, pedunculated, noncalcified or prolapsing valvular vegetation.

Therefore, aortic valve quadricuspid is prone to infective endocarditis irrespective of its morphology. This rare congenital abnormality should deserve close attention, medical care, patient education for a good oral and dental hygiene, and appropriate antibiotic prophylaxis for procedures.

References