A 4-cuspid (quadricuspid) aortic valve is an extremely rare congenital anomaly. Since the introduction of penicillin, it has been estimated that the incidence of pneumococcal endocarditis of all cases of native valve endocarditis in adults has decreased 1–3%.1 But penicillin-resistant strains of Streptococcus pneumoniae (PRSP) are now found worldwide. A rare case, which started with complete heart block by quadricuspid aortic valve PRSP endocarditis, is described.

Case Report

A 62-year-old woman was admitted to Hitachi General Hospital on December 22, 2000 because of syncope and fever. She had previously had high-grade fever with swelling of the left parotid gland and had been treated with antibiotics for several days at another hospital. She was referred to Hitachi General Hospital to be treated for complete heart block. She had no history of rheumatic fever or heart disease, and did not have a drinking habit. Her body temperature was 37.8°C and blood pressure was 140/60 mmHg with a regular pulse rate of 110 beats/min. A to-and-fro murmur of grade 2/6 was audible and moist rales were heard in both lower lung fields.

Routine laboratory tests showed normocytic anemia with hemoglobin and hematocrit levels of 9.9 g/dl and 29.6%, respectively. Her serum C-reactive protein level was elevated at 3.02 mg/dl. Blood culture showed PRSP that was sensitive only to imipenem and vancomycin. A chest X-ray revealed cardiomegaly with mild pulmonary congestion. The findings of an electrocardiography (ECG) performed 1 year earlier had been within normal limits (Fig 1), but the patient's ECG on admission revealed sinus tachycardia, first-degree atrioventricular block and a complete right bundle branch block without hemi-block of the left bundle (Fig 2). A transesophageal echocardiography (TEE) revealed...
prolapse of the quadricuspid aortic valve (Fig 3) with mild aortic regurgitation (AR), but vegetations could not be detected. The monitor ECG revealed a complete heart block (Fig 4), at which time the patient reported feeling dizzy. As temporary dual-chamber pacing was not performed, pulmonary congestion resulted in little improvement. Therefore, a permanent pacemaker (atrioventricular pacing; VDD type) was implanted on the fourth day of hospitalization, but heart failure was not improved because the patient’s hemodynamic changes worsened. The preoperative diagnosis was quadricuspid aortic valve endocarditis by PRSP.

The operation was done on the 26th day of hospitalization. The aortic valve comprised 4 equal-sized cusps. There was an extra cusp between the right and left coronary cusps (Fig 5). On the annulus of the extra cusp, there was a small vegetation, measuring 2–3 mm in diameter. A dissection, similar to a pocket (15 mm in depth), was made from the annulus of the right and extra cusps to the septum wall. It seemed that the AR was a result from prolapse of the dissected wall into the left ventricle with the right and extra cusps. Aortic valve replacement was performed using a prosthesis (Carbomedics Inc, Indiana, PA, USA). The patient recovered and was discharged.

Discussion

A quadricuspid aortic valve is extremely rare, occurring in only 0.008% of cases as reported by Simonds and colleagues. The anomaly is an important one because of the onset of spontaneous AR in later life and the possibility of infective endocarditis. According to the literature, including the case described, operations for 17 cases of quadricuspid aortic valve have been performed in Japan, and there have been 3 cases of endocarditis (18%).

Before the advent of antibiotics the incidence of pneumococcal infective endocarditis varied between 15% and 25%.

References