Quadricuspid aortic valve: a case study

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A 58-year-old man was admitted to our hospital for severe symptomatic aortic regurgitation revealed by symptoms of progressive dyspnoea (NYHA class III) and congestive heart failure. History included a long-standing hypertension, diabetes mellitus, and smoking. On physical examination, blood pressure was 139/74 mmHg, heart rate 74 b.p.m., cardiac auscultation showed a 2/6e systolic murmur at the aortic area associated with a 2/6e diastolic murmur along the left sternal border. There was no sign of congestive heart failure but the patient was on diuretics.

Transthoracic echocardiography showed a severe aortic regurgitation (left ventricular cardiac output 9.3 L/min, diastolic flow reversal 20 cm/s, and vena contracta 6 mm, no PISA performed) with no clear mechanism, a severe left ventricular enlargement and dysfunction (ejection fraction 35%) and a normal aortic root (38 mm). Two- and three-dimensional transoesophageal echocardiography revealed a quadricuspid asymmetric aortic valve (Panel A) and a severe aortic regurgitation (Panel C) due to the absence of coaptation (Panels A and B). Pre-operative coronary angiography showed a right coronary artery occlusion and a severe stenosis of the left ventricular anterior descending artery. The patient underwent an aortic valve replacement with an associated coronary artery bypass graft. Surgery confirmed the diagnosis of quadricuspid asymmetric aortic valve (Panels D and E).

Quadricuspid aortic valve is a very uncommon congenital malformation, occurring separately or associated with other congenital disorders. It is responsible for regurgitation more often than stenosis. Transoesophageal echocardiography is helpful for the diagnosis and the precise description of the mechanism of the regurgitation.

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