A 69-year-old man, with a history of moderate aortic regurgitation, was admitted to our hospital with symptoms of congestive heart failure. He had undergone implantation of a permanent DDD-pacemaker 2 years previously. On physical examination, his blood pressure was 150/50 mmHg and cardiac auscultation revealed a 3/6 diastolic murmur along the left sternal border. Transthoracic echocardiography showed a left ventricular hypertrophy with an enlarged cavity (left ventricular end-diastolic diameter, 70 mm), a normal left ventricular ejection fraction (55%), a pulmonary artery systolic pressure of 50 mmHg and a central severe aortic regurgitation. Sigmoid aortic valves were mildly thickened, without the usual Y-aspect closure, leading to suspicion of an abnormality of the aortic valve.

Transoesophageal echocardiography showed a quadricuspid aortic valve with three equal thin cusps and one minor, which presented as an 'X-shaped' commissural aspect in diastole. Central coaptation defect due to an incomplete juxtaposition of the four cusps resulted in a central aortic regurgitation (Fig. 1).

A dual-source computed tomography examination was performed before intervention to exclude coronary stenosis and aortic root dilation, and to eliminate any associated malformations of the aorta, such as coarctation of the aorta. In addition, using a volume rendering technique, it was possible to display the abnormal anterior additional minor cusp (Fig. 2, Panels A [white arrow] and B).

The patient underwent elective aortic valve replacement with a biological prosthesis; at surgery, the valve was confirmed to be quadricuspid (Fig. 3), which is a very unusual cause of aortic regurgitation.
Figure 1. Central coaptation defect due to an incomplete juxta-position of the four cusps resulted in a central aortic regurgitation.

Figure 2. Abnormal anterior additional minor cusp (white arrow in Panel A).

Figure 3. Quadricuspid valve.