Unicuspid aortic valve prolapse with severe regurgitation

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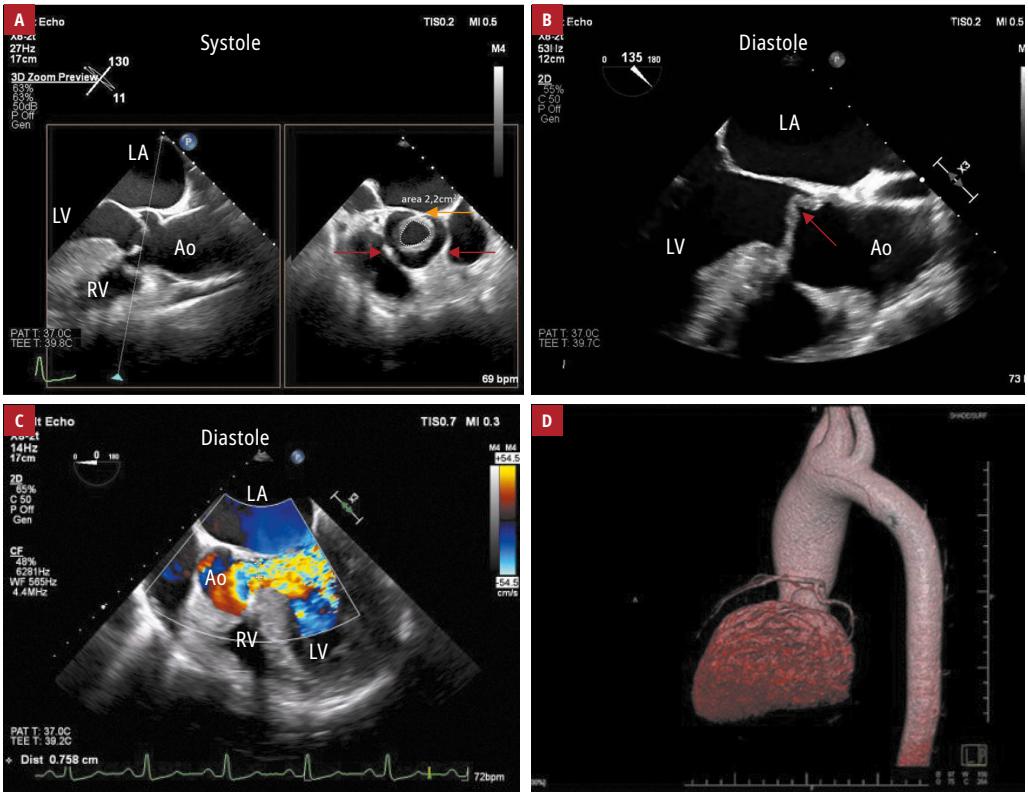

FIGURE 1 **A** – The xPlane imaging with transesophageal echocardiography in midesophageal long and short axes of the aortic root showing the true orifice of the aortic valve. Orange arrow indicates free commissure and red arrows indicate fused commissures. Raphes are not visible. **B** – diastolic image showing the partial prolapse of the unique cusp of the valve (arrow); **C** – a transesophageal echocardiographic 4-chamber view demonstrating severe aortic regurgitation; **D** – computed tomography of coronary vessels and the dilated ascending aorta. Abbreviations: Ao, aorta; LV, left ventricle; LA, left atrium; RV, right ventricle

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A 42-year-old man was admitted to the hospital because of worsening exertional dyspnea and chest pain. Several years ago, he was diagnosed with bicuspid aortic valve with moderate regurgitation and stenosis. Laboratory findings revealed mildly elevated levels of N-terminal pro-B-type natriuretic peptide and borderline elevated levels of troponin. Transthoracic echocardiography and transesophageal echocardiography (TEE) demonstrated ascending

aorta aneurysm (5.2 cm), a noncalcified, unicuspid, unicommissural aortic valve, and severe, eccentric aortic regurgitation due to partial prolapse of the cusp (FIGURE 1A-1C; Supplementary material, *Videos S1–S3*). Although the transaortic gradient was elevated (mean pressure gradient, 34 mm Hg), xPlane and 3-dimensional TEE planimetry revealed the aortic valve area of 2.2 to 2.3 cm^2 (FIGURE 1A; Supplementary material, *Figure S1*). Thus, the elevated gradient was mainly caused by increased flow rather than valve stenosis. Although left ventricular ejection fraction was normal (67%), the longitudinal strain analysis revealed subclinical dysfunction of basal and mid segments of the left ventricle, while the apical segments were hyperkinetic (Supplementary material, *Videos S4–S6*).

Computed tomography excluded coronary lesions and confirmed aortic aneurysm (FIGURE 1D). Because of the low probability of successful valve repair, the patient was referred for the Bentall and de Bono surgery.

The unicuspid aortic valve is a very rare congenital anomaly (prevalence of 0.02% of the adult population). It usually presents in the third to fifth decade of life; however, it might happen earlier in patients with concomitant stenosis, which substantially burdens left ventricle and accelerates progression and hemodynamic consequences of an aortic valve defect.[1] We present a very rare case of regurgitant but not significantly stenotic unicommissural aortic valve. Due to similar clinical features, it was misdiagnosed as bicuspid aortic valve. A detailed 3-dimensional TEE revealed the true anatomy of the valve.

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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