CASE IMAGE

When right heart valves are open 24/7

7/24 açık sağ kalp kapakçıkları

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Department of Cardiology, King's College Hospital, London, United Kingdom A 76-year-old man presented with fatigue, facial flushing, and weight loss experienced over a period of 4 months. An examination revealed a palpable abdominal mass, elevated jugular venous

pressure, and a pansystolic heart murmur at the lower left sternal edge. A computed tomography scan was suggestive of a carcinoid tumor of the small bowel with mesenteric involvement and liver metastases. The level of urinary 5-hydroxyindole acetic acid was significantly



elevated and a magnetic resonance image of the liver showed extensive metastases. A transthoracic echocardiogram showed the typical findings of carcinoid heart disease. The tricuspid valve leaflets were thickened, retracted, and restricted, with a significant coaptation gap that remained open throughout the cardiac cycle (Fig. A, B; Fig. J; Video*) causing severe (free) tricuspid regurgitation (Fig. C, D) with systolic flow reversal in the hepatic veins (Fig. E). The continuous wave Doppler signal of the tricuspid regurgitation was dense, asymmetric, and V-shaped due to rapid equalization of right atrial and ventricular pressure (Fig. F). The pulmonary valve cusps were also thickened, retracted, and restricted, causing severe valvular regurgitation (Fig. G). As a result of the valvular lesions, the right heart chambers were severely dilated (Fig. H, I). Patients with neuroendocrine tumors and carcinoid syndrome are at risk of developing carcinoid heart disease. Pathological endocardial fibrosis of the tricuspid and pulmonary valves can cause valvular regurgitation and sometimes stenosis, leading to right heart dysfunction. Left-sided cardiac involvement is rare. Patients with carcinoid heart disease have a poor prognosis. Valve replacement improves symptoms and survival; however, it is indicated only in those with a stable carcinoid tumor and controlled symptoms.



Figures– Transthoracic echocardiogram. (A) Parasternal right ventricular (RV) inflow view in systole showing thickened and restricted tricuspid valve leaflets in open position; (B) Apical view focusing on RV in systole showing thickened and restricted tricuspid valve leaflets in open position; (C) Parasternal RV inflow view in systole with color Doppler showing severe (free) tricuspid regurgitation; (D) Apical 4-chamber view in systole with color Doppler (color comparison) showing severe (free) tricuspid regurgitation; (E) Pulse wave Doppler image of hepatic vein showing systolic flow reversal compatible with severe tricuspid regurgitation (white arrows); (F) Continuous-wave Doppler image of the tricuspid valve in parasternal RV inflow view showing dense systolic signal due to severe tricuspid regurgitation, with the characteristic asymmetric V-shape (red arrows) due to rapid equalization of pressure between the right atrium and right ventricle; (G) Parasternal RV outflow view in diastole with color Doppler (color comparison) showing severe pulmonic valve regurgitation; (H) Apical 4-chamber view in end-diastole showing the severely dilated right ventricle; (I) Apical 4-chamber view in end-systole showing the severely dilated right ventricle; (I) Apical 4-chamber view in end-systole showing the severely dilated right atrium. LA: Left atrium; LV: Left ventricle; PV: Pulmonic valve; RA: Right atrium; RV: Right ventricle; TV: Tricuspid valve. (J) Transthoracic echocardiogram. Three-dimensional, full volume in 4-chamber view in mid-systole. Right panel shows the atrioventricular valves from the ventricular side. The mitral valve is closed; however, the tricuspid valve remains open due to leaflet thickening and restriction. MV: Mitral valve; RA: Right atrium; RV: Right ventricle; TV: Tricuspid valve. **Supplementary video files associated with this presentation can be found in the online version of the journal*.