

## Cardiac Imaging

# Carcinoid Heart Disease in a Patient with Primary Ovarian Carcinoid Tumour

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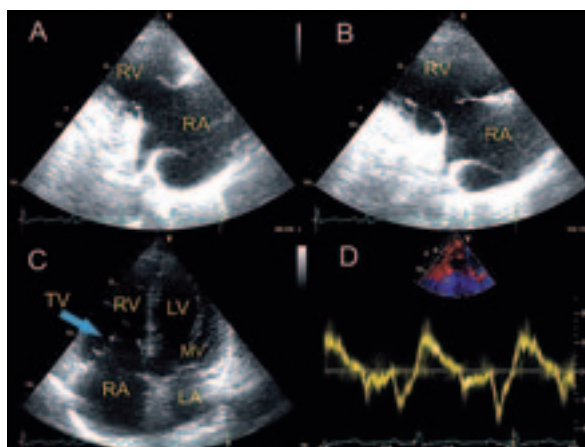
A 66-year-old woman was referred to our echocardiography lab with debilitating symptoms of right heart failure with New York Heart Association functional class III. She reported abdominal fullness, lower extremity oedema and easy fatigability during the last year.

Six months prior to referral she had undergone hysterectomy with bilateral salpingo-oophorectomy because of ovarian tumours. The histopathological study of the right ovary showed a teratoma predominantly consisting of carcinoid tissue. Chest and abdomen computed tomography at that time showed no evidence of disease on either side, throughout the body. During her routine follow-up, the patient progressively developed symptoms of right heart failure which at the beginning were managed medically with diuretics.

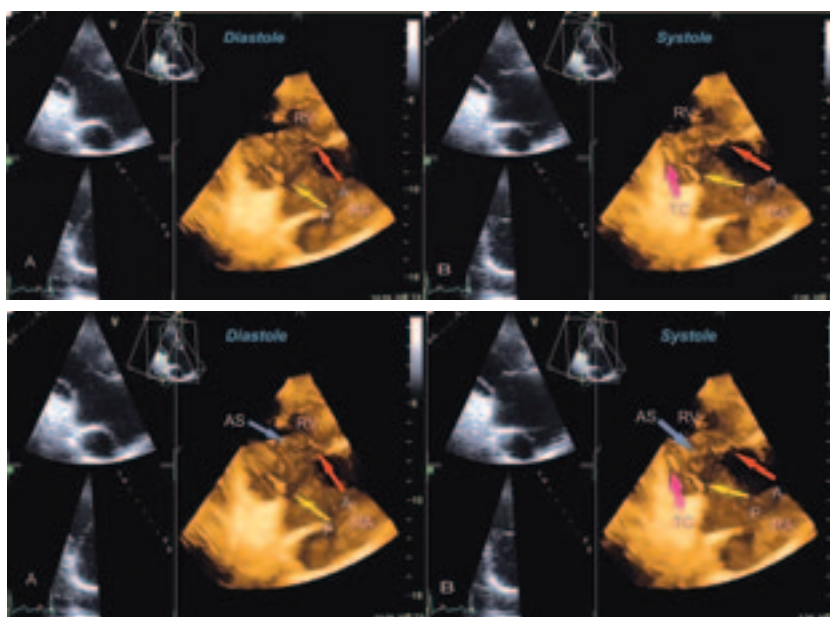
Echocardiographic examination revealed a dilated right ventricle, but with good function (Figures 1, 2), and a thickened, retracted and fixed tricuspid valve (Figure 2), which remained in a semi-open position, thus leading to severe tricuspid regurgitation (Figure 3). The pulmonary cusps also appeared fibrosed and thickened with subsequent severe pulmonic regurgitation and mild stenosis (Figure 4).

The diagnosis of carcinoid heart disease was proposed and the patient underwent tricuspid valve replacement. Microscopic examination of the tricuspid valve demonstrated the typical fibrous tissue deposition (Figure 5).

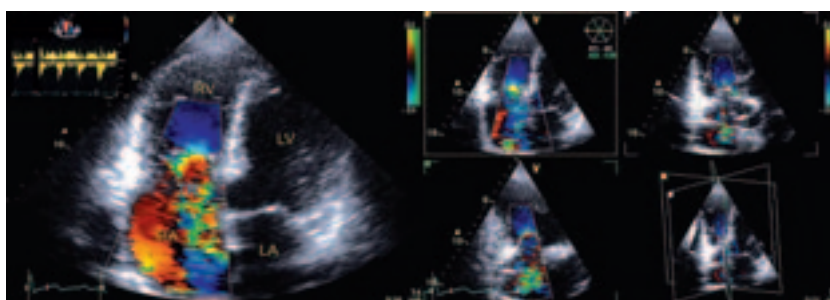
The most dreaded complication of carcinoid syndrome is carcinoid heart disease,<sup>1</sup> which in most cases is associated with the presence of hepatic metastases. However,



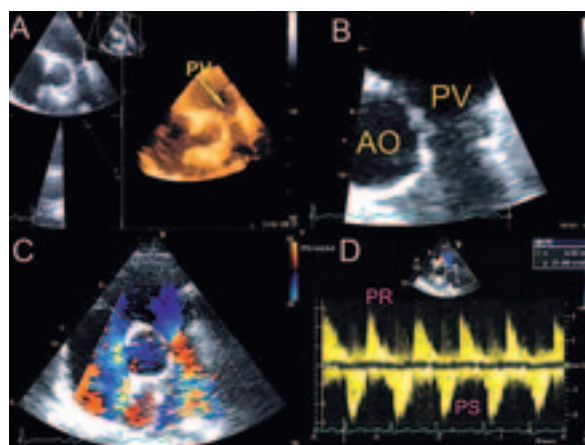
**Figure 1.** Long parasternal axis view of the right ventricular (RV) inflow tract. The anterior and posterior tricuspid leaflets are shown thickened and shortened in diastole (A) and systole (B), when they remain in a semi-open position. Apical 4-chamber view (C), showing the dilated RV compared to the left ventricle, the tricuspid leaflets that have become fixed and remain in a semi-open position (blue arrow) while the mitral valve is closed. Spectral tissue Doppler imaging (D) of the tricuspid annulus showing normal systolic wave, an index of preserved RV function. RV – right ventricle; RA – right atrium; LV – left ventricle; LA – left atrium; MV – mitral valve; TV – tricuspid valve.



**Figure 2.** Real time 3D-echocardiography showing the tricuspid annulus and tricuspid leaflets in diastole (A) and systole (B). The anterior tricuspid leaflet (A, red arrow) exhibits relatively preserved motion, while the posterior leaflet (P, yellow arrow) appears thickened, shortened, retracted and immobile. The *chorda tendinea* for the posterior leaflet (TC, pink arrow) also appears thickened, shortened and retracted.



**Figure 3.** Colour Doppler triplane echocardiography showing the severe tricuspid regurgitation (TR) as a result of the fixed tricuspid valve. In the left upper corner continuous wave Doppler shows the characteristic dagger-shaped profile of the TR, with an early peak velocity and a rapid decline, indicating rapid pressure equalisation between the right-sided cardiac chambers. RV – right ventricle; RA – right atrium; LV – left ventricle; LA – left atrium.



**Figure 4.** Short axis view of the great vessels illustrates the pulmonary valve (PV) cusps thickened with retraction (A: 3D-echocardiography, B: zoom of PV). Colour Doppler showing aliasing after the PV indicating high velocities (C) while continuous wave Doppler (D) shows severe pneumonic regurgitation with a rapid pressure half time and a mild pneumonic stenosis with a peak gradient of 21 mmHg. PV – pneumonic valve; AO – aorta; PR – pneumonic regurgitation; PS – pneumonic stenosis.



**Figure 5.** Microscopic examination of the tricuspid valve demonstrating typical fibrous tissue deposition (H&E stain, magnification x 40).

rarely, patients with a primary ovarian carcinoid, as in our case, develop carcinoid heart disease in the absence of hepatic metastases because the ovarian veins, which drain vasoactive substances secreted from the primary tumour, bypass the portal circulation.<sup>2</sup>

### References

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