## Atypical presentation of a primary ovarian carcinoid tumour with carcinoid heart disease: diagnosis by transthoracic echocardiography

## **CASE SUMMARY**

A 64-year-old Tanzanian female was admitted to the medical ward with large ascites and pedal oedema. An abdominal ultrasound detected a large right-ovarian mass without metastasis that was confirmed by CT scan. Her cancer antigen 125 was elevated at 766 IU/ml (normal, <35 IU/ml). An echocardiogram showed a dilated right side, along with severely thickened fibrotic tricuspid-valve leaflets which were tethered, retracted, shortened and immobile with lack of coaptation, and were fixed in the diastolic position (figure 1 A–C/movie). There were severe tricuspid regurgitation and stenosis (figure 2 A–C), which were typical of carcinoid heart disease. However, the urinary 5-hydroxyindoleacetic acid (5-HIAA) level was normal (19.1  $\mu$ mol/

24 h (normal, <47  $\mu mol/24$  h)). The patient underwent ovarian mass resection (15×15 cm) which on histopathology proved to be carcinoid. Subsequently, she also underwent a successful tricuspid valve replacement.

Primary ovarian carcinoid tumour causing carcinoid heart disease is very rare; approximately 30 cases are reported in the literature. It has been reported that large carcinoid tumours metastasise more frequently and present with carcinoid syndrome more often.<sup>1</sup> This was not seen in this patient, even though she had an ovarian carcinoid tumour measuring >15 cm. Her 5-HIAA levels were normal. This could be due to an atypical tumour that secretes serotonin which is not metabolised to 5-HIAA or secretes serotonin intermittently or due to secretion of non-serotonin vasoactive substances (eg, amines).<sup>1</sup> An increase in cancer antigen 125 is extremely rare in a patient with a carcinoid tumour, and this is explained by inflammation of mesentery and peritoneal irritation caused by mediators released from the carcinoid.<sup>2</sup>

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**Figure 1** (A) Transthoracic echocardiogram in a four-chamber view showing thickened, rigid, retracted and fixed tricuspid valve leaflets (arrowheads). (B) Transthoracic echocardiogram in a right-ventricular inflow view showing thickened, tricuspid valve (TV) leaflets fixed in mid-position causing failure of coaptation in systole resulting in a large regurgitant valve orifice. (C) Dilated right ventricle (RV) and right atrium (RA) with thickened and fused chordae tendinae (arrowheads) causing constant semiopen 'funnel shaped' position, in a patient with carcinoid heart disease.



**Figure 2** (A) Colour Doppler demonstrating severe tricuspid regurgitation through a wide regurgitant orifice into a dilated right atrium (RA). (B) Colour Doppler demonstrating a high-velocity turbulent diastolic tricuspid flow of severe tricuspid stenosis into the right ventricle (RV) in a patient with carcinoid heart disease. (C) Continuous-wave Doppler showing severe tricuspid stenosis and 'dagger-shaped' profile of tricuspid regurgitation (arrowheads) due to early peak pressure and rapid decline representing equalisation of right atrial and ventricular pressures.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; internally peer reviewed.

Heart Asia 2011;3:51-52. doi:10.1136/heartasia-2011-010022

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