Carcinoid heart disease: correlation of echocardiographic and histopathological findings

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DESCRIPTION

A 41-year-old man presented to an anaesthetic pre-assessment clinic with a metastatic neuroendocrine tumour and clinical features of carcinoid syndrome for consideration of hepatic resection. He gave a history of worsening exertional dyspnoea and peripheral oedema. A loud systolic murmur was audible over the right sternal edge. Transthoracic echocardiography revealed severe tricuspid and pulmonic regurgitation and right ventricular volume overload consistent with carcinoid heart disease, necessitating double valve replacement.

Intraoperative transoesophageal echocardiography (figure 1A) demonstrated thickening and retraction of the tricuspid valve leaflets. Colour flow Doppler (figure 1B) revealed torrential tricuspid regurgitation. Three-dimensional imaging in systole (figure 1C) and diastole (figure 1D) shows minimal motion of the tricuspid valve over the cardiac cycle, and a large regurgitant orifice area.

Histopathological analysis of the excised cardiac valves demonstrated thickening of valve leaflets due to carcinoid plaques, composed of a proliferation of myofibroblasts and the deposition of a myxoid extracellular matrix (figure 2A, B).

Valvular dysfunction in carcinoid heart disease is caused by proliferation of endocardial fibroblasts in response to chronic inflammation, or induced by circulating vasoactive mediators.¹ Deposition of plaques on the valve leaflets and subvalvular apparatus results in thickening, retraction, impaired leaflet motion and greatly enhanced echogenicity.²

Carcinoid heart disease is a challenge for anaesthetists, because of right ventricular failure and the risk of intraoperative carcinoid crisis. This manifests as severe bronchospasm, hypertension or hypotension, and is associated with mortality of up to 18%.³ With appropriate preventative measures, our patient encountered no serious intraoperative issues, was discharged from intensive care unit 24 h

Figure 1 Two-dimensional imaging demonstrates a tricuspid valve fixed open in systole (A). The red arrow points to the normal mitral valve. Colour flow Doppler, again during systole showing severe tricuspid regurgitation (B). Three-dimensional imaging in systole (C) and diastole (D) showing relatively little change in the orifice area during the cardiac cycle. The approximate positions of the septal (S), posterior (P) and anterior (A) tricuspid valve leaflets are shown.

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Figure 2 The pulmonic valve (stained with H&E) is shown at $\times 40$ (A) and $\times 200$ (B) magnification. The leaflet is grossly thickened by carcinoid plaque, composed of myofibroblasts (red arrow) surrounded by a myxoid matrix.

after his procedure, and eventually to home on postoperative day 5. Now 4 months postprocedure, he is asymptomatic, and has returned to work. Clinicians managing patients with carcinoid syndrome should be aware of the high incidence of carcinoid heart disease and the implications for anaesthesia and surgery.

Learning points

- Echocardiography imaging in carcinoid heart disease demonstrates the classical findings of thickening and retraction of the valve leaflets with coaptation failure due to plaque deposition.
- The right-sided valves are usually affected in isolation as the pulmonary circulation filters out vasoactive mediators released by the tumour.
- About 50% of patients with carcinoid syndrome will develop carcinoid heart disease. Clinicians involved in the management of patients with carcinoid syndrome must be aware of this high incidence and investigate appropriately.

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