

Canadian Residents' Corner / Coin canadien des résidents en radiologie

Case of the Month #166: Carcinoid Heart Disease

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Clinical Presentation

A previously healthy 60-year-old woman presented with a 3-week history of fatigue and exertional dyspnea. Over the past year, she had experienced episodic wheezing, facial flushing, diarrhoea, and an unintentional 40-lb weight loss. On physical examination, she demonstrated venous telangiectasia in the malar region, mild peripheral cyanosis, and an elevated jugular venous pressure with a dominant V wave. On cardiac auscultation, there was a holosystolic murmur at the left lower sternal region of grade III/VI intensity that increased on inspiration. Transthoracic echocardiography was performed to confirm the valvular abnormalities detected on physical examination (Figure 1A). Cardiac magnetic resonance imaging (CMR) was performed to quantify right ventricular systolic function and computed tomography of the abdomen was performed as shown Figures 1B and C.

Diagnosis

Carcinoid syndrome.

Radiologic Findings

Transthoracic echocardiography identified severe tricuspid valve regurgitation with thickened immobile leaflets (Figure 1A). CMR imaging confirmed the echocardiographic

Key Words: Carcinoid syndrome; Tricuspid regurgitation; Echocardiography; Cardiac magnetic resonance imaging.

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findings of a dilated right ventricular end diastolic volume index (98 mL/m²) and mild right ventricular systolic dysfunction with an ejection fraction (EF) of 45% (Figure 1B). Computed tomography of the abdomen revealed multiple hypodensities in the liver and a mesenteric mass, with evidence of a primary small-bowel tumour (Figure 1C).

Discussion

Carcinoid tumours are relatively rare, with an incidence of 3–4 per 100,000 population per year [1]. The primary tumour is located in the gastrointestinal tract or the bronchopulmonary tree in more than 98% of cases [2]. Cardiac involvement is the presenting feature in fewer than 20% of cases but develops in more than 50% of cases [3]. The right-sided valves, with the tricuspid valve in all patients and the pulmonary valve in the majority of patients, are the major sites of cardiac involvement [3]. Left-sided heart disease is significantly less common and is usually associated with a patent foramen ovale [3]. Valvular lesions are characterized by shortened, thickened, and immobile leaflets. This leads to valvular regurgitation and can result in combined regurgitation and stenosis of the affected valve [3]. Right ventricular volume overload and dilatation may occur as a result of the tricuspid regurgitation. CMR offers additional information in terms of detailed evaluation of accurate right ventricular volumes and systolic function [3]. Our case supports the use of multimodality cardiac imaging for the diagnosis and anatomic characterization of carcinoid heart disease.

In our patient, markedly elevated levels of 5-hydroxyindolacetic acid, and chromogranin A confirmed the diagnosis of carcinoid syndrome. In the absence of surgical intervention, individuals with carcinoid heart disease may develop worsening symptomatic right-sided heart failure.

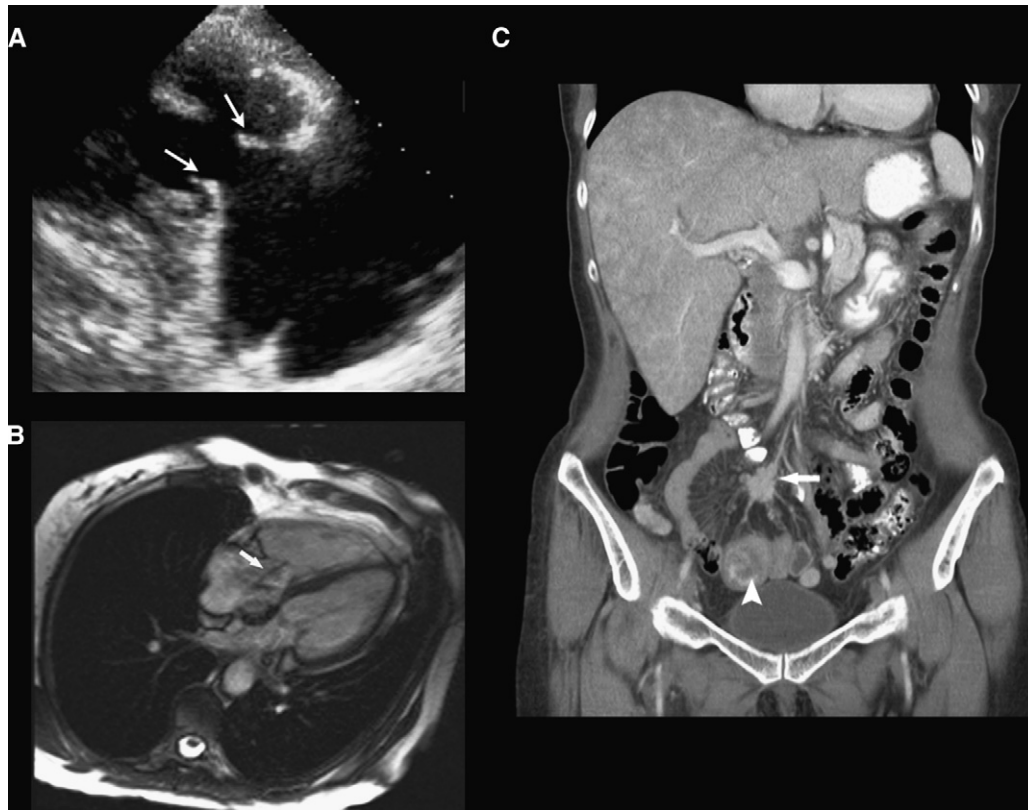


Figure 1. (A) Transthoracic echocardiographic right ventricular inflow view, demonstrating immobile tricuspid valve leaflets (arrows) consistent with carcinoid heart disease. (B) Four-chamber view obtained with balanced, steady-state, free precession magnetic resonance, demonstrating a linear signal void that resulted from tricuspid regurgitation (arrow) because of carcinoid heart disease. (C) Coronal contrast-enhanced computed tomography image, demonstrating a speculated nodal mass in the mesentery (arrow), which has metastasized from a more inferiorly located primary tumour in the small bowel (arrowhead).

The mean life expectancy of patients with carcinoid heart disease and with cardiac involvement is 1.6 years compared with 4.6 years for those without cardiac involvement [4]. Our patient underwent tricuspid valve replacement and closure of the patent foramen ovale. Before surgery, a continuous infusion of octreotide was started to prevent perioperative complications of carcinoid syndrome. After surgery, the patient initiated monthly octreotide injections and interferon alpha therapy, with significant improvement in her symptoms at 1 year of follow-up.

References

- [1] Modlin IM, Lye KD, Kidd MA. 5-decade analysis of 13,715 carcinoid tumors. *Cancer* 2003;97:934–59.
- [2] Modlin IM, Sandor A. An analysis of 8305 cases of carcinoid tumors. *Cancer* 1997;79:813–29.
- [3] Bhattacharyya S, Davar J, Dreyfus G, et al. Carcinoid heart disease. *Circulation* 2007;116:2860–5.
- [4] Pellika PA, Tajik AJ, Khandheria BK, et al. Carcinoid heart disease: clinical and echocardiographic spectrum in 74 patients. *Circulation* 1993;87:1188–96.