

IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Carcinoid Heart Disease From a Primary Liver Carcinoid Tumor



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ABSTRACT

Carcinoid heart disease (CaHD) is part of the carcinoid syndrome. Cardiac involvement is present in 20% to 60% of patients with carcinoid syndrome and is normally from liver metastasis. We report the case of a patient who presented with CaHD disease with an undiagnosed primary tumor or a possible primary liver carcinoid tumor. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:1360-1362) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 31-year-old woman presented with a 2-year history of evolving facial flushing, palpitations, abdominal pain, and chronic diarrhea, to which progressive dyspnea and lower extremity edema was added 12 months before admission. The patient arrived at our hospital because of exacerbation of dyspnea at rest, nocturnal paroxysmal dyspnea, orthopnea, and worsening edema in her lower limbs. Physical examination showed stable vital signs, facial flushing, grade III jugular plethora, a pulmonary systolic murmur II/III and a tricuspid III/IV murmur, hepatalgia, and bilateral hard edema in lower limbs. A clinical diagnosis of right-sided heart failure was made.

A transthoracic echocardiogram was requested, which showed dilation of the right atrial and right ventricular cavities, with severe tricuspid and pulmonary insufficiency (**Figure 1A**, **Videos 1 and 2**). The diagnosis of cardiac carcinoid syndrome was suspected because of the transthoracic echocardiogram findings in combination with the patient's clinical characteristics. Her 5-hydroxyindoleacetic acid level was requested in a 24-hour urine collection and was reported to be 50 mg (normal range, 2-8 mg/d). In addition, a total body scan was performed using scintigraphy (Infinia Hawkeye 4 High Resolution, GE Healthcare) with the somatostatin analogue indium-111 octreotide. The scan showed focal areas of abnormal concentration of the radiotracer in the upper right hepatic lobe, a finding corroborated by single-photon emission computed tomography (CT)-CT hybrid images (**Figure 1B**).

The patient underwent bioprosthetic valve replacement with an Epic #27 tricuspid valve (Abbott) and an Epic #25 pulmonary valve (Abbott). Later, the patient was discharged with orders for 30-mL intramuscular octreotide monthly. The removed valves were sent for pathologic examination, which determined the presence of stromal myxoid degeneration, mixed with areas of fibrosis; some necrotic foci and areas with some inflammatory processes related to mononuclear activity were also identified. The patient continues to have

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abdominal pain, and chemoembolization of the hepatic tumor was performed. In later follow-up, she is recovering adequately and remains asymptomatic.

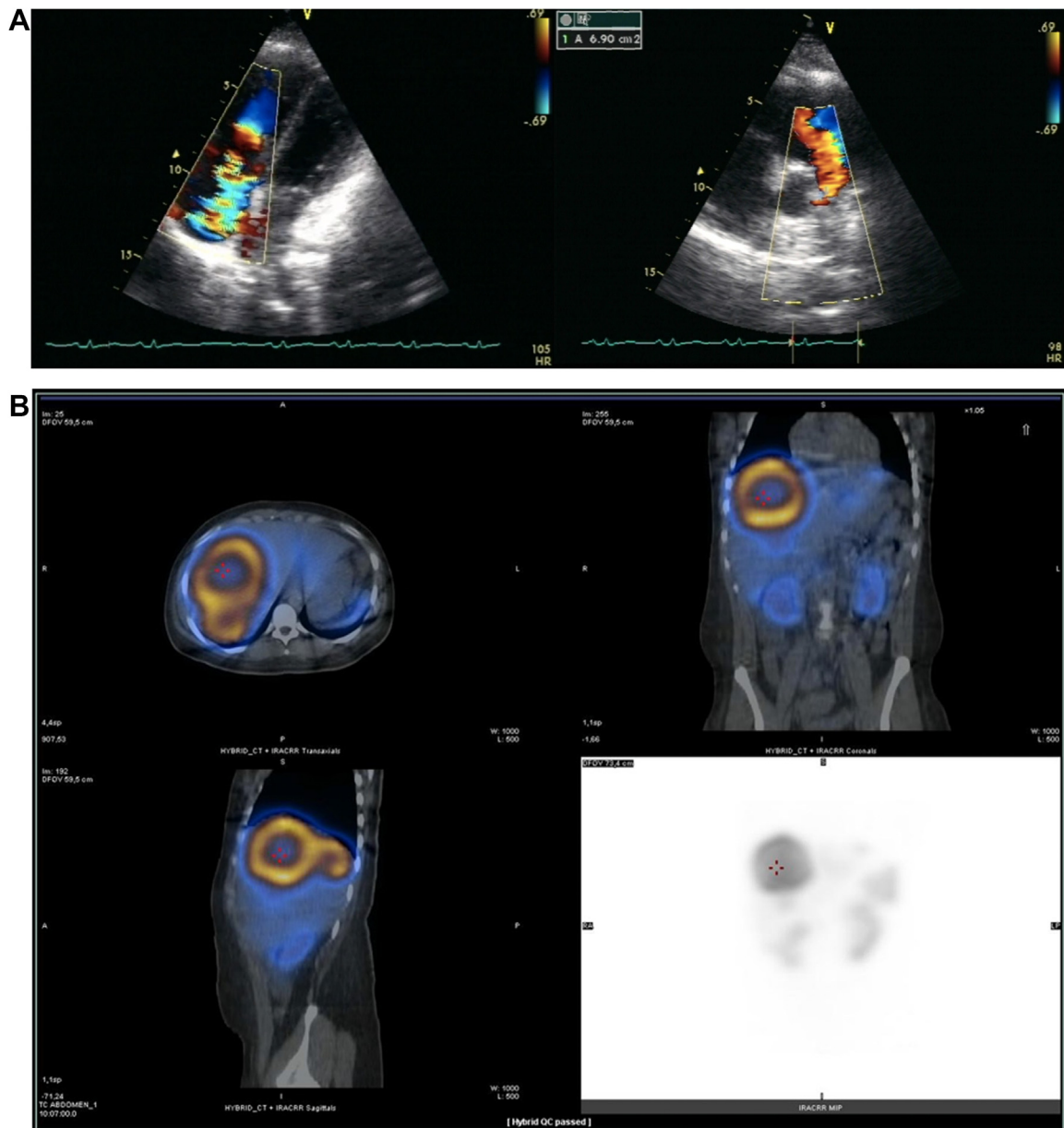
Carcinoid heart disease (CaHD) is an entity that is part of the carcinoid syndrome, which is characterized with the classic triad of facial flushing, intestinal hypermotility, and cardiac compromise, findings that were identified in our patient.

**ABBREVIATIONS
AND ACRONYMS**

CaHD = carcinoid heart disease

CT = computed tomography

FIGURE 1 Echocardiography and Scintigraphy



(A) Transthoracic echocardiogram. **(Left)** An apical 4-chamber view showed a dilated right ventricle and a fixed and thickened tricuspid valve. Once Doppler imaging was applied on this view, it showed severe tricuspid insufficiency jet that filled the entire right atrium. **(Right)** A parasternal short-axis view with Doppler imaging showed a severe pulmonary valve insufficiency jet. **(B)** A somatostatin analogue scintigraphy body scan showed focal areas of abnormal concentration of the radiotracer in the upper right hepatic lobe, corroborated by single-photon emission computed tomography-computed tomography hybrid images, with a probable primary carcinoid liver tumor, which can be seen on the **2 upper and left bottom** images.

Cardiac involvement can represent 20% to 60% of the carcinoid syndrome, mainly affecting the tricuspid and pulmonary valves, with insufficiency of these valves later leading to dilation of the right-sided cavities and finally right-sided heart failure. This phenomenon occurs when a carcinoid tumor involves the liver, thus allowing high levels of 5-hydroxytryptamine to reach the systemic circulation.¹ There are newer radionuclear modalities, such as gallium-68 dotatate positron emission tomography combined with CT or copper-64 dotatate scans, which are much more sensitive in revealing tumors with somatostatin, although in our hospital these modalities are not available. For this reason, the scan used in this case used a somatostatin analogue, and the results guided us toward the high probability of a primary hepatic tumor and CaHD, a rare presentation according to the available literature.² This case could represent liver metastasis from an undiagnosed primary tumor or, rarely, from a primary hepatic neuroendocrine tumor. Somatostatin analogues such as octreotide improve symptoms and survival in patients with CaHD by reducing the levels of vasoactive peptides that cause the symptoms.³

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KEY WORDS chronic heart failure, echocardiography, fibrosis, pulmonic valve, right ventricle, tricuspid valve

APPENDIX For supplemental videos, please see the online version of this article.