Revista Portuguesa de **Cardiologia**

Portuguese Journal of Cardiology

www.revportcardiol.org





Frozen tricuspid valve – A case of a carcinoid syndrome





A válvula tricúspide congelada – Um caso de síndrome carcinoide

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Received 15 September 2024; accepted 10 November 2024 Available online 3 April 2025

A 43-year-old male patient reported a one-year history of diarrhea, flushing, weight loss and jaundice. Physical examination showed bilateral lower limb edema. Cardiac auscultation revealed a holosystolic murmur irradiating to the axilla. He had no known medical conditions and took no medication. Electrocardiogram revealed sinus rhythm with a heart rate of 55 bpm. Chest xray was normal (Figure 1). Laboratory findings revealed cholestatic/hepatocellular liver injury with hyperbilirubinemia. Auto-immune markers and viral serologies were negative. Full body computed tomography showed hepatomegaly, no signs of chronic liver disease and three liver nodular lesions. Liver biopsy revealed metastasis of a neuroendocrine tumor. A 68Ga-DOTATATE positron emission tomography scan showed increased liver uptake and an uptake area in the small intestine.

Additional blood work revealed elevated serum chromogranin A, neuron-specific enolase and urinary 5-Hydroxyindoleacetic acid.

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Figure 1 Chest Xray.

https://doi.org/10.1016/j.repc.2024.11.017

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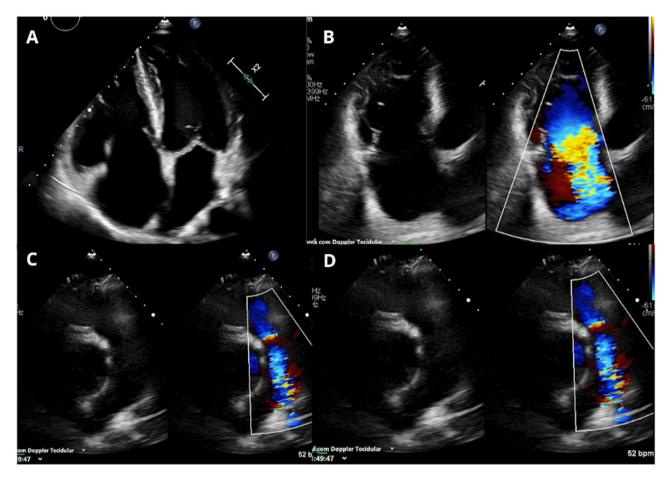


Figure 2 Transthoracic echocardiogram showing: thickening and immobility of the tricuspid leaflets (A) and torrential regurgitation (B and C). Thickening and restriction of the pulmonary valve (D) with moderate regurgitation and mild stenosis (D).

A multidisciplinary tumor board decision was to start systemic treatment with a somatostatin analog and consider liver surgery/transplant if stable disease or response took place after six months.

Pre-transplant transthoracic echocardiogram showed dilation of right heart chambers, thickening and immobility of the tricuspid leaflets causing torrential regurgitation (Figure 2: Panels A and B; Video 1), thickening and restriction of the pulmonary valve with moderate regurgitation and mild stenosis (Figure 2: Panels C and D; Video 2) suggestive of carcinoid syndrome. No left heart involvement was documented.

Carcinoid tumors are rare and have a systemic expression in up to 5% of cases with right heart involvement in 20–66% of cases^{1,2} Echocardiography is a useful imaging tool with diagnostic and prognostic value.^{1,2} This case highlights the importance of echocardiography showing pathognomonic cardiac manifestations of a rare subset of tumors.

Conflicts of interest

The authors have no conflicts of interest to declare.

Appendix A. Supplementary data

Supplementary material associated with this article can be found in the online version at https://doi.org/10. 1016/j.repc.2024.11.017.

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