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João Mendes Cravo Ana Rita Faria Francisco Capinha Rafael Cruz
Ana Spencer

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Frozen tricuspid valve – A case of a Carcinoid Syndrome

A válvula tricúspide congelada – Um caso de síndrome carcinoide

João Mendes Cravo[1*], Ana Rita Faria[2], Francisco Capinha[3], Rafael Cruz[4], Ana Spencer[5]

(1)Department of Cardiology, Unidade Local de Saúde Santa Maria, CAML, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal
(2)Department of Cardiology, Unidade Local de Saúde Santa Maria, CAML, CCUL@RISE, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal
(3)Department of Gastrenterology, Unidade Local de Saúde Santa Maria. University Clinic of Gastrenterology, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal
(4) Department of Pathology, Unidade Local de Saúde Santa Maria. Institute of Anatomic Pathology & Institute of Histology and Development Biology, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal
(5) Medical Oncology, Unidade Local de Saúde Santa Maria, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal

*E-mail address: jmcravo123@gmail.com (J. Mendes Cravo)

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 showed 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.
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-B; Video 1), thickening and restriction of the pulmonary valve with moderate regurgitation and mild stenosis (Figure 2: Panels C-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.

Ethics in publishing
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3. Does your study include a clinical trial?:
No
4. Are all data shown in the figures and tables also shown in the text of the Results section and
discussed in the Conclusions?:
Yes

Conflicts of Interest

The authors have no conflicts of interest to declare.

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Legends:

Figure 1: Chest Xray

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 e F).

Figure 1: Chest X-Ray

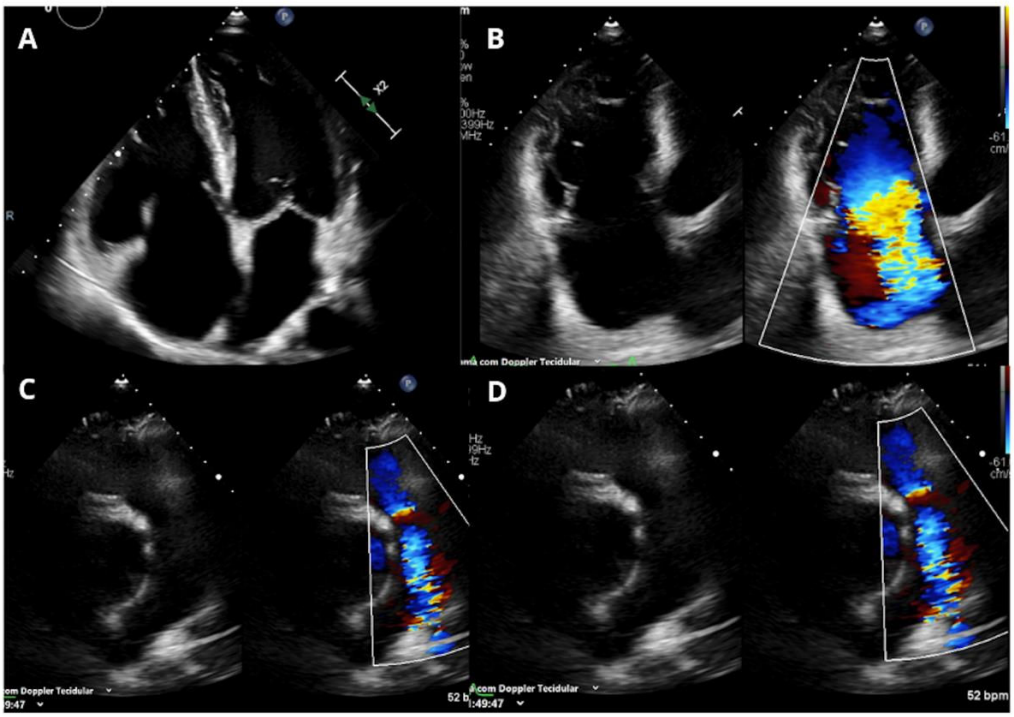

Figure 2: Transthoracic echocardiogram showing: Thickening and immobility of the tricuspid leaflets (A) and torrential regurgitation (B and C). Thickening and restriction of the pulmonary valve (D) with moderate regurgitation and mild stenosis (D e F).

Video 1: Torrential tricuspid regurgitation.

Video 2: Moderate regurgitation and mild stenosis of pulmonary valve.