Carcinoid Complexity: A Syndrome Revealing Lung Tumor and Heart Disease

Complexidade Carcinoide: Uma Síndrome que Revela um Tumor Pulmonar e Doença Cardíaca

Keywords: Carcinoid Heart Disease; Carcinoid Tumor; Lung Neoplasms; Somatostatin Analogs
Palavras-chave: Doença Cardíaca Carcinoide; Neoplasias do Pulmão; Tumor Carcinoide

Dear Editor,

Lung carcinoid tumors (LC) account for 20% - 25% of all neuroendocrine tumors (NETs) and 1% - 2% of all cases of lung cancer.[1,2] About 20% to 30% of patients have disseminated disease at diagnosis, presenting with carcinoid syndrome (CS), and of these, about half have carcinoid heart disease (CHD).[2,3]

The authors present the case of a 79-year-old man, a former smoker, who was admitted to the emergency department due to shortness of breath and tiredness on minor exertion and productive cough starting one month before. The patient was admitted with a suspected respiratory infection and therefore started an empirical antibiotic regimen, but worsened over time, with dyspnea for progressively minor efforts and anasarca. Diagnostic tests were carried out detecting a progressive increase in NT-proBNP levels (up to 4300 pg/mL – reference value 0 – 125 pg/mL) and a transthoracic echocardiogram demonstrated dilated right cavities and left atrium, with decreased overall RV systolic function, mild to moderate tricuspid regurgitation, with an estimated pulmonary artery systolic pressure of about 82 mmHg – normal mPAP 14 ± 3 mmHg) (Fig. 1) and a minor pericardial effusion, predominantly posterior. Considering the hypothesis of CS as a presentation of a disseminated carcinoid tumor, a thoraco-abdominopelvic computed tomography (CT) revealed a mass in the right lower lobe, with approximately 50 mm in the longest axis, mediastinal and hilar adenopathy, and numerous hepatic nodules that captured contrast. Diagnostic tests included: measurement of serum chromogranin A (326 ng/mL) and urinary 5-HIAA (12.5 mg/24 hours), both high; bronchoscopy, with histology of the biopsies carried out suggestive of a typical lung carcinoid tumor, expressing chromogranin and Ki67 (about 1%); brain CT and PET/CT with gallium 68 (^{68}Ga-DOTATOC PET/CT) that showed a malignant tumor lesion (Fig. 2) with overex-pression of somatostatin receptors in the lymph nodes, liver, and bones. Given the findings, the patient was diagnosed with a primary bronchopulmonary carcinoid tumor with liver and bone metastasis and was proposed for therapy with octreotide every four weeks.

Although there was initial symptomatic improvement, the ^{68}Ga-DOTATOC PET/CT performed to assess the response showed extensive splenic metastasis and progression of lymph node metastasis, leading to initiation of second-line therapy with everolimus, which was suspended 15 days after the first dose due to adverse effects. At the time of writing, the patient was under surveillance, with symptomatic treatment with octreotide. This case highlights the diagnostic and therapeutic challenges of CS and the importance of an integrated approach using biochemical screening, imaging, and echocardiography to guide diagnosis and treatment.

AUTHOR CONTRIBUTIONS

SRL: Study design, writing of the manuscript.
JB, MIMVL: Data analysis and interpretation.
SV: Critical review of the manuscript.
All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki

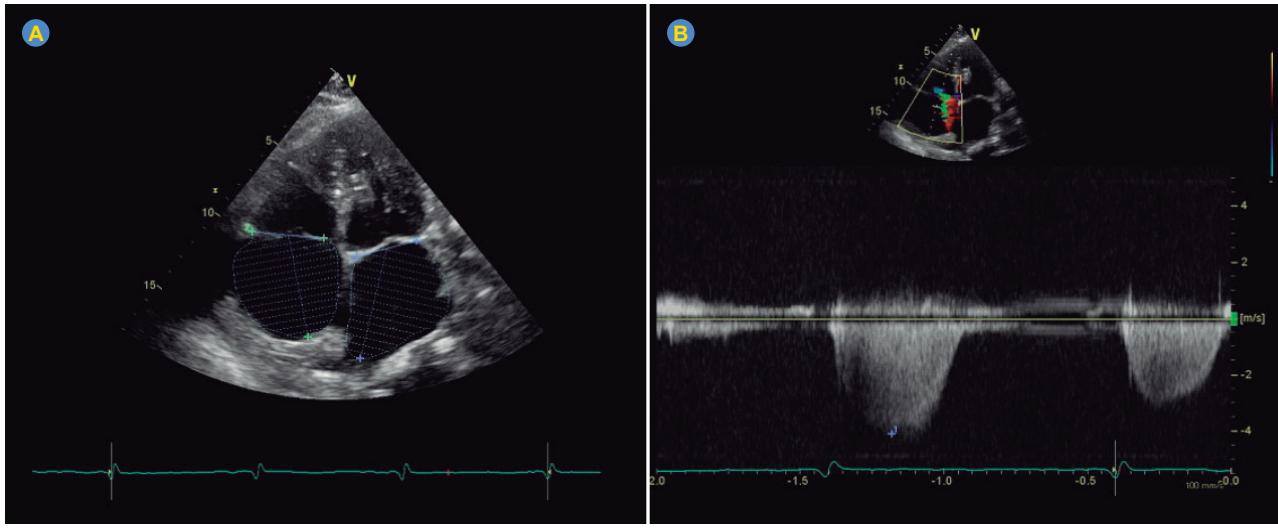

Figure 1 – Transthoracic echocardiogram demonstrating dilated right cavities and left atrium (A); mild to moderate tricuspid regurgitation, with an estimated pulmonary artery systolic pressure (ePASP) of about 82 mmHg (B).

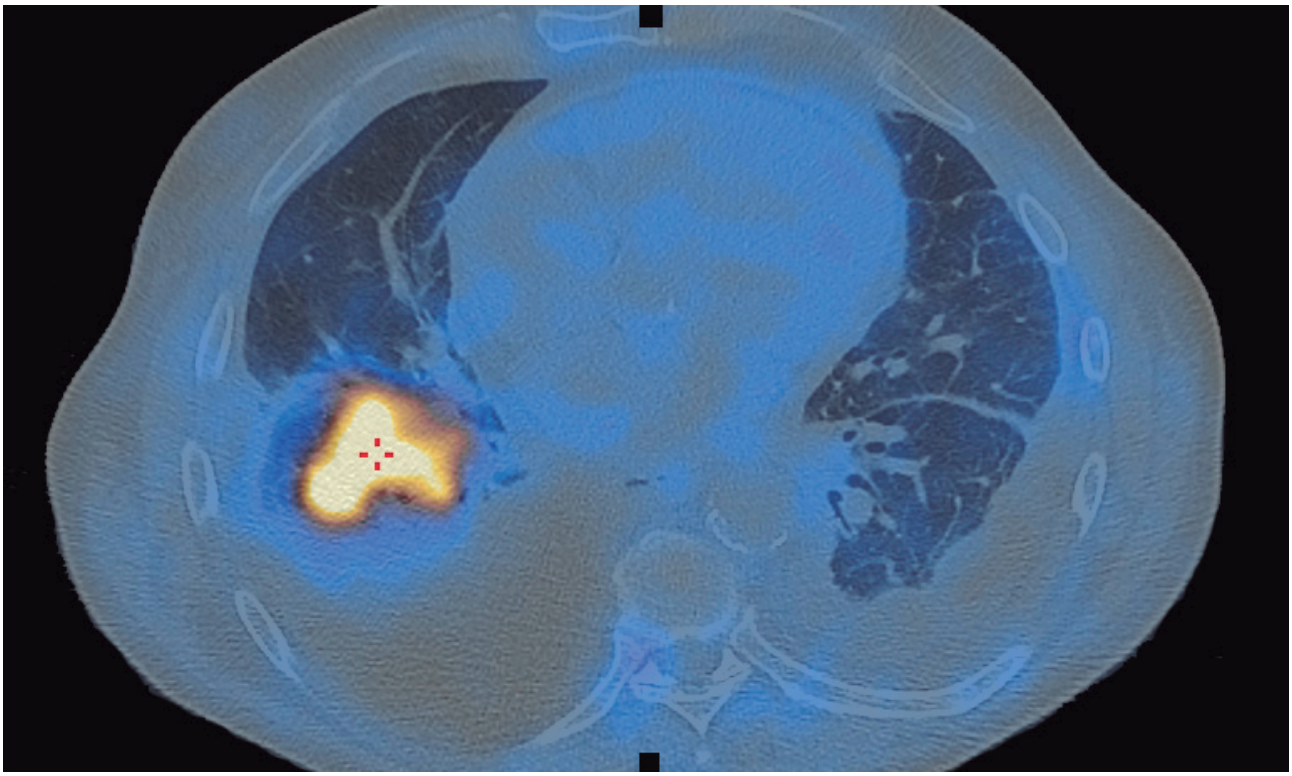

Figure 2 – PET-CT with gallium 68 showing uptake in the right lower lobe (SUVmax = 12.3)

Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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REFERENCES

1. Herde R, Kokeny K, Reddy C, Akerley W, Hu N, Boltax J, et al. Primary pulmonary carcinoid tumor: a long-term single institution experience/ Primary pulmonary carcinoid tumor. Am J Clin Oncol. 2018;41:24-9.
2. Baudin E, Caplin M, Garcia-Carbonero R, Fazia N, Ferolla P, Filosso P, et al. Lung and thymic carcinoids: ESMO clinical pratice guidelines for treatment and follow-up. Ann Oncol. 2021;32:439-51.
3. Kulke M, Mayer R. Carcinoid tumors. N Engl J Med. 1999;340:858-68.

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