Case Report

Non-Hodgkin's Lymphoma with Heart Disease as a Rare Cause of Acute Heart Failure: A Case Report

Linfoma Não Hodgkin com Acometimento Cardíaco: Uma Causa Rara de Insuficiência Cardíaca Aguda – Relato de Caso

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Introduction

Lymphoma is the third most common cause of cardiac metastases, following only lung and breast cancers. Cardiac involvement represents 13.6% of metastatic tumors of the heart.[1] Diffuse large B-cell lymphoma (DLBCL) is the most common non-Hodgkin's lymphoma (31%) and rapidly fatal when left untreated.[2] Cardiac involvement can occur through three paths: continuity from intrathoracic lesions, retrograde lymphatic dissemination, or the blood.[3] It is most commonly found in the interatrial septum of the right chambers extending to the pericardium, with a lower prevalence in the left atrium and ventricle.[4-6]

The spectrum of cardiac manifestations is wide and ranges from an absence of symptoms to heart failure (HF), pericardial effusion, or arrhythmias.[4,5] Cardiac involvement is rarely the initial manifestation of lymphoma.[5] However, it generally presents a poor prognosis due to delayed clinical diagnosis and high invasive potential. Thus, an early identification of the tumor allows for timely treatment in an attempt to improve prognosis.

This article presents a rare clinical case of lymphoproliferative disease with cardiac involvement of the pericardium extending to the left atrium and ventricle.

Case report

A 67-year-old female patient born in Pernambuco and living in São Paulo was admitted to a tertiary hospital with a dry cough, progressive dyspnea, orthopnea, and paroxysmal nocturnal dyspnea for 2 weeks that had worsened in the previous 3 days. The patient was hypertensive, a 20 pack-years ex-smoker, and used acetylsalicylic acid, losartan, spironolactone, and carvedilol.

A physical examination showed tachypnea, tachycardia, slight jugular stasis, and palpable bilateral cervical lymph nodes with a hardened consistency. No murmurs were found on cardiac auscultation, and the cardiac rhythm was regular. Pulmonary auscultation identified diffuse wheezing and bibasilar crackling rales. Palpation of the abdomen revealed painful hepatomegaly up to 4 cm from the right costal margin with peripheral edema (2+/4+). Chest radiography showed an enlarged cardiac area, slight pulmonary congestion, and mediastinal enlargement.

Transthoracic echocardiography showed an intracardiac lobulated heterogeneous mass approximately 5 cm in its longest axis adherent to the lateral and inferior walls of the left ventricle with a movable component at the extremity.

Chest computed tomography (CT) was performed to assess the intracardiac mass and its relationship with the surrounding structures and determine its limits. Chest CT showed contact with the adjacent pericardium and a central area of necrotic aspect measuring approximately 11.0 × 3.2 cm. Lymph node enlargement was also identified in the right upper and lower paratracheal, subcarinal, left pre-vascular, right supraclavicular, and left internal thoracic chains (Figure 1). Staging was performed using CT scans of the skull, abdomen, and pelvis, but no extracardiac tumors were identified.

Cardiac magnetic resonance imaging (CMR) for the morphological and functional evaluation of the heart and better characterization of the intracardiac mass showed the presence of a large pericardial mass located predominantly in the lower and lateral portions of the left atrium and basal inferolateral segment of the left ventricle. The mass infiltrated the myocardium and the interatrial septum, was hyperintense in T2-weighted images, and demonstrated perfusion after contrast injection with areas of heterogeneous impregnation (Figure 2).

Considering the morphological and hemodynamic characteristics of this intracardiac mass of undetermined nature and the presence of lymph node enlargement in the right upper and lower paratracheal chains, a right cervical lymph node biopsy was performed. The immunohistochemical study was positive for CD20, MUM1, BCL2, BCL6, c-myc, Ki-67 (90% of neoplastic cells), CD3, and CD5 (positive in small mature lymphocytes), negative for the other markers, and Epstein-Barr virus–negative. The association with histopathological findings led to the diagnosis of DLBCL, activated B-cell immunophenotype (Hans algorithm), with the double immunohistochemical expression of myc and bcl-2.

The patient underwent pulse therapy with prednisone, followed by a rituximab regimen of chemotherapy associated with miniCHOP (cyclophosphamide, adriamycin, vincristine,

Keywords

Heart Failure; Lymphoma, Non-Hodgkin; Neoplasm Metastasis.

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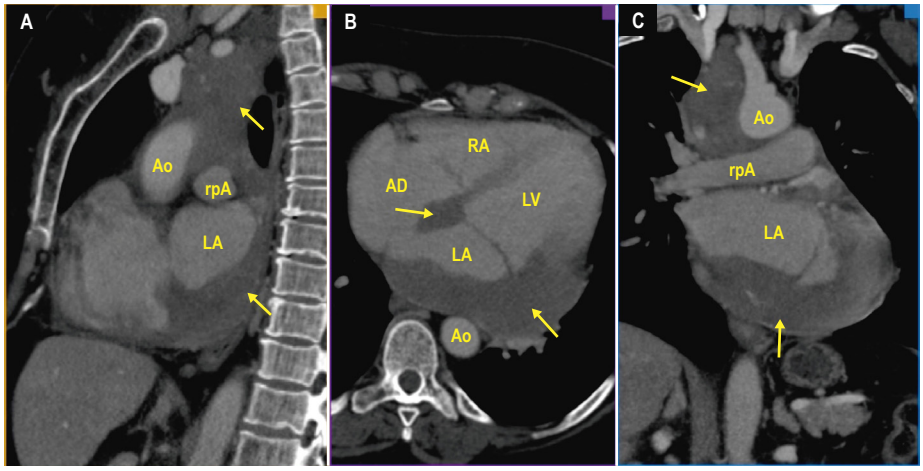

Ao, aorta; LA, left atrium; LV, left ventricle; RA, right atrium; rpA, right pulmonary artery; RV, right ventricle.

Figure 1 – *Computed tomography. (A) Sagittal section. (B) Axial section. (C) Coronal section. Mediastinal lymph node enlargement and expansive formation with heterogeneous content located in the lower and lateral regions of the left atrium. Infiltration is noted in the interatrial septum and the basal segment of the lower and lateral left ventricle walls.*

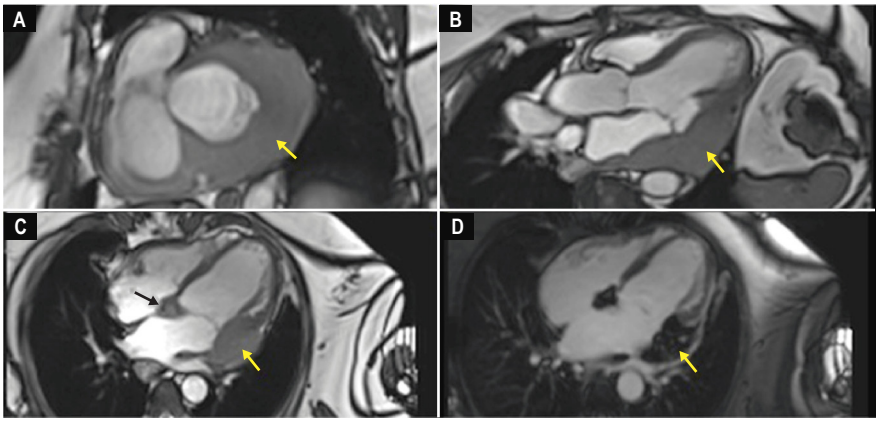

Figure 2 – *Cardiac magnetic resonance. (A) Short axis. (B) Three chambers. (C) Four chambers. (D) Late enhancement after administration of the contrast medium with heterogeneous impregnation suggestive of neoplasia. A mass is visible in the inferior and lateral regions of the left atrium. It also infiltrates the interatrial septum (black arrow) and the basal segment of the lower and lateral left ventricle walls.*

and prednisolone associated with rituximab), methotrexate, and intrathecal dexamethasone. The patient's clinical status improved, and she was discharged from the hospital while continuing the chemotherapy regimen. After 4 months of treatment, positron emission tomography–CT (PET-CT) showed total remission of the pericardial mass.

Discussion

This article presented a case of DLBCL involving the pericardium and extending to the posterior wall of the left cardiac chambers associated with cervical and mediastinal lymph node enlargement.

Lymphomas affecting the heart occur more commonly in the right chambers, with more than 80% located on the left ventricle wall. Of these, about half are associated with pericardial involvement causing a pericardial effusion. They can also occupy the right atrium, extending to the interatrial septum and left atrium.[8] Symptoms depend on tumor position and appear mostly in the final stage of the disease. The presence of HF is relatively common, as reported here.

Imaging exams can assist with the diagnosis. Chest radiography, despite its low sensitivity and specificity, can show changes such as mediastinal structure deviation or widening, cardiomegaly, cardiac silhouette changes, and HF signs. Transthoracic echocardiography is a sensitive method for identifying cardiac involvement by tumors.[7] Chest CT characterized the cardiac mass's morphology, location, and extension as well as the involvement of extracardiac structures, including lymph node enlargement. Magnetic resonance imaging enabled differentiation between the tumor mass and the myocardium due to better tissue characterization.

The diagnosis is usually confirmed by histopathological analysis of the pericardial or pleural fluid, when affected, or by endomyocardial biopsy. Adjuvant treatment includes several chemotherapy regimens such as CHOP, either alone or associated with radiotherapy. In this case, the patient underwent a cervical ganglion biopsy due to the risks inherent to myocardial biopsy as well as isolated chemotherapy due to her clinical conditions and tumor characteristics. The prognosis of these cases is generally poor, with a survival of less than 1 month without treatment. However, with an early diagnosis followed by appropriate treatment, survival can reach approximately 5 years.[8]

Conclusion

This case shows the early diagnosis of cardiac involvement by a lymphoproliferative malignant tumor in an elderly patient. Imaging examinations provided information about its location and involvement of the pericardium, left cardiac chambers, and lymph nodes. An early diagnosis and treatment change the natural history of the disease, improving the patient's prognosis and survival.

Authors' contributions

Research conception and design: Melo ESA, Oshiro FS, Veloso PM, and Terêncio AS; data collection: Melo ESA, Oshiro FS, Veloso PM, and Terêncio AS; data analysis and interpretation: Melo ESA, Oshiro FS, and Siqueira MEM; manuscript writing: Melo ESA, Oshiro FS, and Siqueira MEM; and critical review of the manuscript for important intellectual content: Siqueira MEM and Szarf G. Funding: This study received no funding.

Conflict of interest

The authors have declared that they have no conflict of interest.

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