

Clinical Case

Noninfective endocarditis with atypical presentation, resembling infiltrative tumor with coronary vascularization

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ARTICLE INFORMATION

Received on January 18, 2025.

Accepted after review on July 7, 2025.

www.revistafac.org.ar

There are no conflicts of interest to disclose.

Keywords:

Noninfective endocarditis,
intracardiac tumor,
coronary vascularization.

ABSTRACT

Noninfective endocarditis is a rare pathologic process, primarily associated with autoimmune diseases and neoplasms. Diagnosis is established by ruling out infective endocarditis and cardiac tumors, such as myxoma or sarcomas. However, it is unusual to find highly vascularized vegetations, and even more unusual for this vascularization to originate from the main coronary arteries. We present the clinical case of a patient with a mass found in the left atrium, which was highly vascularized by the circumflex artery, leading to suspicion of intracardiac tumor. The patient underwent tumor resection, and histopathological analysis revealed findings consistent with endocarditis.

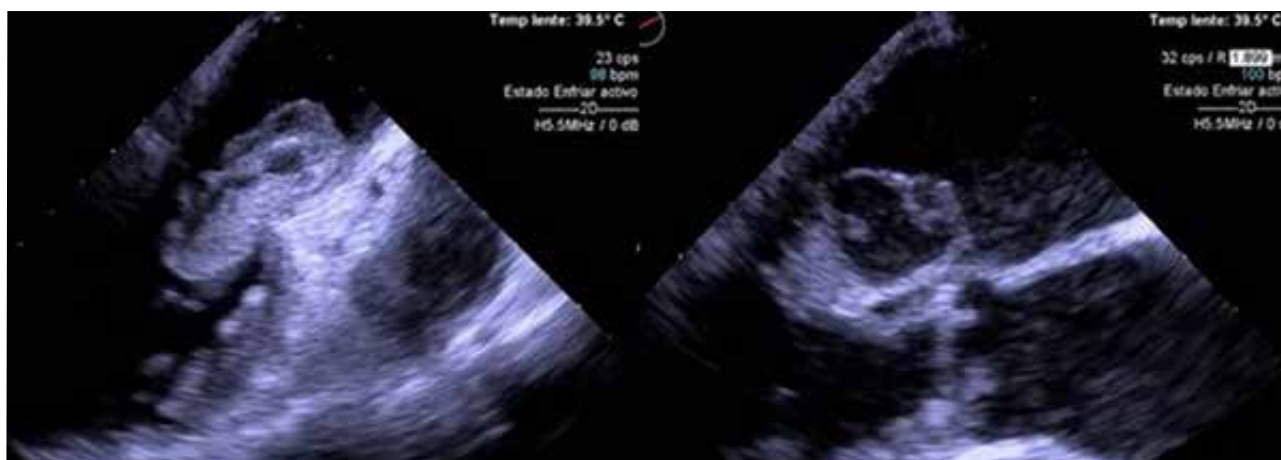
INTRODUCTION

Noninfective endocarditis is a rare pathological process that is mainly associated to autoimmune diseases and neoplasms. Diagnosis is made by ruling out infective endocarditis, and subsequently, cardiac tumors (for instance, myxomas or sarcomas). However, it is rare to find highly vascularized vegetations, and it is even more rare for said vascularization to emerge from the main coronary arteries. Next, we present the clinical case of a female patient with a finding of left atrial mass, that was highly vascularized by the circumflex artery. Because of this, there was suspicion of intracardiac tumor, and tumor resection was conducted. In the histopathological analysis, endocarditis findings were reported.

CLINICAL CASE PRESENTATION

Female, 51-year-old patient, with overweight and dyslipidemia in treatment with non-pharmacological measures and statins. The patient presented at the hospital with clinical symptoms characterized by fever of up to 39°C of afternoon-night predominance, dry cough fits and pleuritic pain in physical exploration with presence of diastolic murmur in mitral focus. As part of the approach, three blood tests were carried out, which were negative. Procalcitonin was within the normal range, and ECG was in sinus rhythm. On the other hand, chest X-rays showed cardiomegaly grade I, with no other significant findings. Transthoracic echocardiogram was also carried out, where left intra-atrial mass

was documented; thus, transesophageal echo was conducted to better identify the lesion (*Figure 1*). In this test, irregular mobile cardiac mass was found, with solid and cystic characteristics and vascularized; likewise, the mass measured 5.1 cm by 9 cm, with an area of 9.2 cm²; adhered from the left atrial appendage to the aortomitral continuity, protruding through the mitral valve, conditioning severe mitral valve stenosis. For this reason, the patient was admitted in the cardiology area, where coronary angiography was performed, with multiple microfistulae being observed with origin in the proximal segment of the circumflex artery draining into the appendage, left atrium and an infiltrative tumor (*Figure 2*). An assessment was requested by the cardiothoracic surgery service, and tumor resection was indicated. The surgical intervention was carried out later, by extracorporeal circulation with aortic cross-clamping for 1 h and 25 min; an irregular tumor was found, of cystic appearance, of 3 cm by 3 cm, vascularized, which was adhered to the left atrial roof, 3 cm from the atrioventricular valve. During the surgery, mesenchymal neoplasm was found; the patient was extubated successfully in the OR, presenting hemodynamic instability, requiring vasopressor and inotropic support, which could be withdrawn immediately after the surgery. Later, the patient was referred to post-surgical care in the Intensive Care Unit, where she coursed favorably, and with no complications associated to the hospital stay. Finally, the patient was discharged 8 days later. The anatomopathological result was collected, with

**FIGURE 1.**

Transesophageal echocardiogram: mobile intra-atrial mass, conditioning severe mitral valve stenosis

report of connective tissue with reactive fibroblasts associated to sclerotic areas, with neoformation vessels with fibrin strands deposits, chronic inflammation with lymphocytes, compatible with chronic and acute endocarditis; furthermore, special staining was carried out that led to ruling out neoplastic process (Figure 3). Currently, the patient remains asymptomatic and in functional class I of the New York Heart Association.

DISCUSSION

Noninfective endocarditis, also known as nonbacterial thrombotic endocarditis (NBTE) or aseptic endocarditis, refers to a rare condition characterized by the formation of sterile vegetations.

Libman and Sacks published in 1924 a series of cases of four patients in whom these atypical lesions were associated to systemic lupus erythematosus. These vegetations are not associated to inflammatory response or bloodstream in-

fections, and in general occur as a result of fibrin deposition and platelet aggregation.¹

The etiology of NBTE is little known and several interaction mechanisms (including circulating immune complexes, hypercoagulable states and carcinomatosis) may play a role, which in the end, produce endothelial damage and exposition of subendothelial connective tissue to circulating platelets. It is well known that malignant neoplasms have impaired antithrombotic mechanisms, thus increasing the risk of thrombosis and subsequent valve lesions; on the other hand, the NBTE relationship with malignancy is well known.²

Macrophages interact with malignant cells to produce circulating cytokines (such as tumor necrosis factor and interleukin-1) that cause endothelial damage, with the subsequent promotion of sterile thrombi and platelet deposition in the endocardium. These macrophages also stimulate the production of coagulation factors to create a hypercoagulable state.³

It is imperative to differentiate NBTE from infective endocarditis (including culture-negative endocarditis); thus, the diagnostic approach is addressed to ruling out infective endocarditis and assessing the probable cause for NBTE.

Clinically, the patients with NBTE often appear asymptomatic, and occasionally, the only clinical manifestation are thrombotic events, because vegetations are more friable. Rarely, patients present murmurs or signs and symptoms of valve or heart failure. Those who do present them, valve regurgitation is more frequent than stenosis.

Diagnostic tests should be made for infective endocarditis. Equally, three blood tests should be made before starting the therapy with antibiotics, with the aim of differentiating infective endocarditis from culture-negative endocarditis. Also, typical and atypical organisms of infective endocarditis should be sought (including fungi, the HACEK group, *Coxiella burnetii* or chlamydia), using specialized techniques as necessary; furthermore, if there is a clinical suspicion of autoimmune disease, it should be verified: ANA,

**FIGURE 2.**

Coronary angiography: multiple microfistulae are observed, originating in the proximal segment of the circumflex artery, irrigating infiltrative tumor.

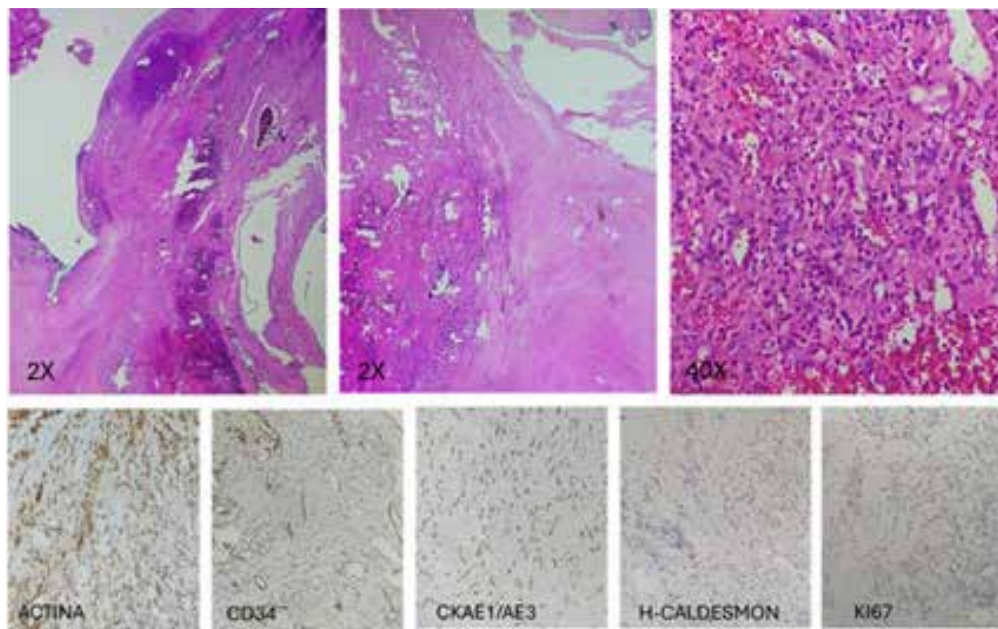


FIGURE 3. Histopathological slice with 10 and 40x images, where findings are observed, compatible with endocarditis. Besides, immunohistochemistry test was performed, where malignant neoplasm was ruled out.

anti-dsDNA, rheumatoid factor, anti-cardiolipin antibodies and anti- β 2-glycoprotein 1 antibodies.⁴

Likewise, transthoracic echocardiogram should be carried out to seek for vegetations in heart valves if NBTE is suspected. It is important to take into account that echocardiogram could be normal if the vegetations are already embolized. Transesophageal echocardiogram is more sensitive in comparison to transthoracic image; mainly in patients with small vegetations (<5 mm), and it should be done in those with normal transthoracic echocardiogram and a strong suspicion of NBTE.⁵ The vegetations of infectious and non-infectious etiology cannot be differentiated in echocardiography. Nevertheless, the vegetations observed in NBTE are usually small (<1 cm), wide-based and irregular.

Diagnosis can only be confirmed in histological evaluation. The vegetations vary in size and are usually located in areas of turbulent flow along the closure lines of the valve or in the valve themselves. Valve lesions present similar histological appearances, regardless of the underlying etiology, consisting of degenerative platelets intertwined with fibrin strands, granulomatous tissue and fibrotic foci. There are no inflammatory cells or infectious microorganisms present.⁶ The histopathology test also enables the diagnosis of primary malignant cardiac neoplasms, such as angiosarcoma and myxoma. In brief, the diagnosis of NBTE can only be confirmed by the demonstration of platelet thrombi in autopsy or in histology tests on surgical samples. Therefore, a high rate of clinical suspicion is required for diagnosis.

CONCLUSIONS

From what has been presented in this text, we can establish that this is a case of atypical presentation of endocarditis (based on histopathological diagnosis); blood tests in the patient were negative, and she had not received antibiotics previously; thus, we could determine that it was not culture-negative endocarditis, but rather noninfective endocarditis. In this regard, an association with autoimmune diseases is very usual; however, the patient did not know about any previous diseases, and had no clinical or biochemical suspicion of autoimmune diseases, so the neoplastic etiology became the second best choice. We suspected a malignant tumor, as during the surgery a neoplastic lesion was reported, together with it being highly visible due to the circumflex artery; however, in the end the lesion was reported as a highly vascularized vegetation, thus being a unique clinical case.

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