

# Apical Occupation by Endomyocardiofibrosis Associated with Thrombus Diagnosed with Contrast Echocardiography and Resolved with Clinical Treatment

*Ocupação Apical por Endomiocardiofibrose Associada a Trombo Diagnosticada com Ecocardiografia de Contraste e Resolvida com Tratamento Clínico*

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## Abstract

Endomyocardial fibrosis (EMF) is a neglected but prevalent disease in underdeveloped countries. Despite its frequency, it is still considered a rare disease. It is marked by the following findings: architectural distortion, ventricular filling changes, and segmental mobility changes affecting not only diastolic and systolic dynamics but also heart valve function. Apical thrombi can be formed in more advanced disease. In this case report, we revisit EMF and present the challenging case of a 52-year-old woman with New York Heart Association (NYHA) functional classification (FC) IV heart failure. Resting electrocardiography revealed sinus rhythm with left ventricular (LV) overload, high voltage vectors, rectified ST-segment depression, and a negative T-wave in the anterolateral region compatible with the strain pattern. The evaluation of echocardiographic images showed marked left atrial dilation, no ventricular dilatation, and hyper-refringent apical occupation within the LV suggestive of a large sessile thrombus superimposed on the endocardial fibrous layer. Although surgery is the most supported approach in the literature in such situations, this patient was treated with oral anticoagulants (OAC) for 1 month and 24 days. The patient progressed well with an improved NYHA FC. Endocavitary contrast echocardiography performed 2 months after OAC initiation showed regression of the apical thrombus image.

## Introduction

Endomyocardial fibrosis (EMF) is a neglected but prevalent disease in underdeveloped countries. Global estimates show that approximately 10–12 million people have EMF, mainly in sub-Saharan African and some Latin America regions.<sup>1</sup> In Brazil, the prevalence of the disease is 2%, with a higher incidence in women and a female-to-male ratio of 5:1.<sup>2</sup>

## Keywords

Thrombus, Contrast Echocardiography, Oral Anticoagulation.

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The etiology of EMF remains uncertain, but its main morphological characteristic is endocardial layer fibrosis, predominantly in the apical region, that causes architectural distortion, ventricular filling changes, and segmental mobility changes affecting not only diastolic and systolic dynamics but also heart valve functions. In addition, apical occupation by thrombi is found in more advanced disease stages.<sup>3</sup>

Thus, the non-consolidation of the pathophysiological and etiological mechanisms involved in EMF exacerbation complicates therapeutic conduct decisions. In most cases, treatment is based on symptomatic control of heart failure and other cardiomyopathies; in more severe situations, surgical intervention and even heart transplantation are recommended.<sup>4</sup>

## Case report

Here we describe the clinical case of a 52-year-old woman with a history of systemic arterial hypertension, hypothyroidism, hepatic steatosis, and gouty arthritis. She presented with short-duration atypical stabbing chest pain (10–15 minutes) of moderate intensity unrelated to physical exertion and without triggering or improving factors at 2 months after the initial appointment.

Resting electrocardiography (ECG) revealed sinus rhythm with left ventricular (LV) overload, high voltage vectors, rectified ST-segment depression, and a negative T-wave in the anterolateral region that may be compatible with the strain pattern (Figure 1).

Transthoracic echocardiography (TTE) at the time of the initial diagnosis revealed a predominantly apical increased myocardial thickness suggestive of differential diagnoses such as apical hypertrophic cardiomyopathy or EMF. Cardiac magnetic resonance imaging (CMR) suggested endomyocardial fibrosis as a cause of the apical occupancy (Figure 2).

A few months later, the patient demonstrated clinical worsening with restrictive syndrome for which she was hospitalized. A new TTE with sulfur hexafluoride contrast (SonoVue) revealed marked left atrial (LA) dilation with an indexed volume of 102 mL of 56 mL/m<sup>2</sup> (greater than in the previous test), and a hyper-refringent apical LV occupancy image suggested a large sessile thrombus superimposed on the endocardial fibrous tissue measuring 24 × 21 mm that was not present on a previous test (Figure 3).

The patient's LV ejection fraction was preserved (58%





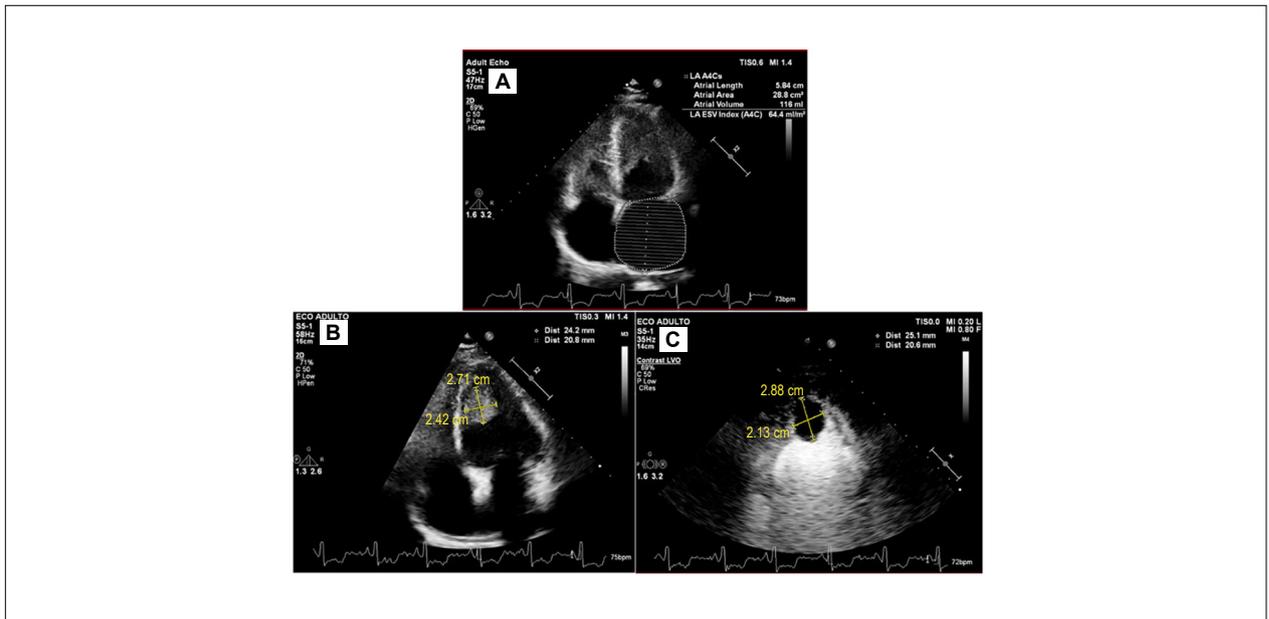


Figure 3 – (A) Measurement of left atrial volume using Simpson's method; (B) Intracavitary thrombus measurement; (C) Use of contrast to increase thrombus enhancement

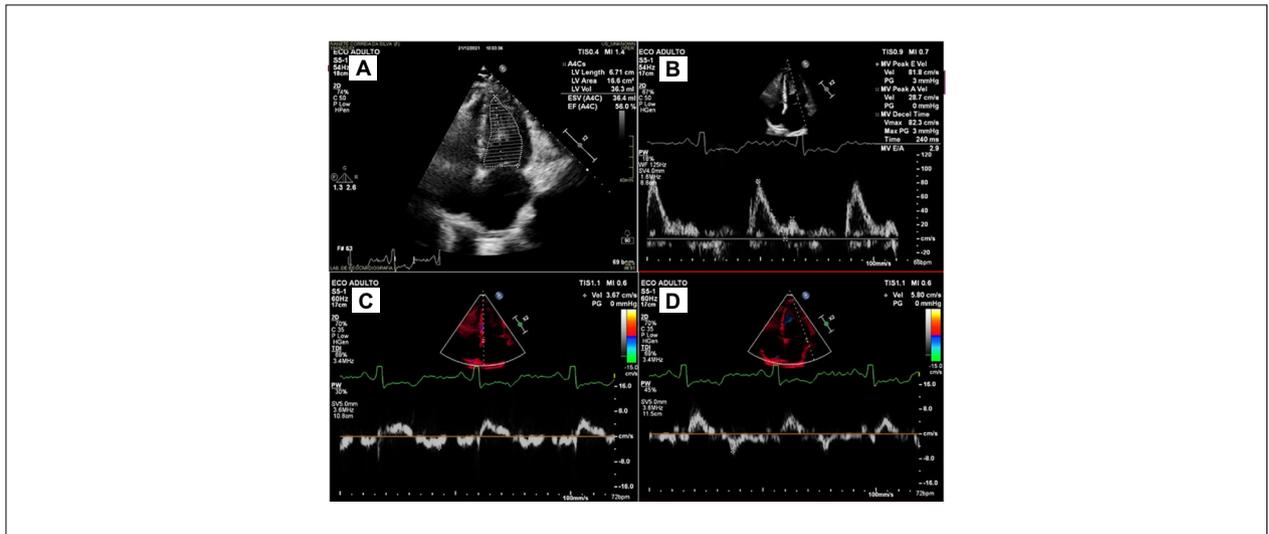


Figure 4 – (A) Ejection fraction measurement; (B) Doppler transmitral flow; (C) Movement of the septal mitral annular plane by tissue Doppler; (D) Movement of the lateral mitral annular plane.

treatment is based on the symptomatic control of heart failure. Surgical intervention for apical evacuation is beneficial in patients with signs of severe ventricular restriction and New York Heart Association functional class III or IV heart failure.<sup>8</sup>

In contrast, here we demonstrated a case of good clinical progression after symptomatic treatment with OAC that could not only be diagnosed with multimodality support, its follow-up and apical thrombus resolution was supported by TTE with cavity contrast.

### Authors' contributions

Manuscript Writing, Data Analysis and Interpretation: Sapalo AT; Data Collection: Cunha R, Gali LG; Critical Review of the Manuscript for Important Intellectual Content: Romano MMD.

### Conflict of interest

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

## Case Report

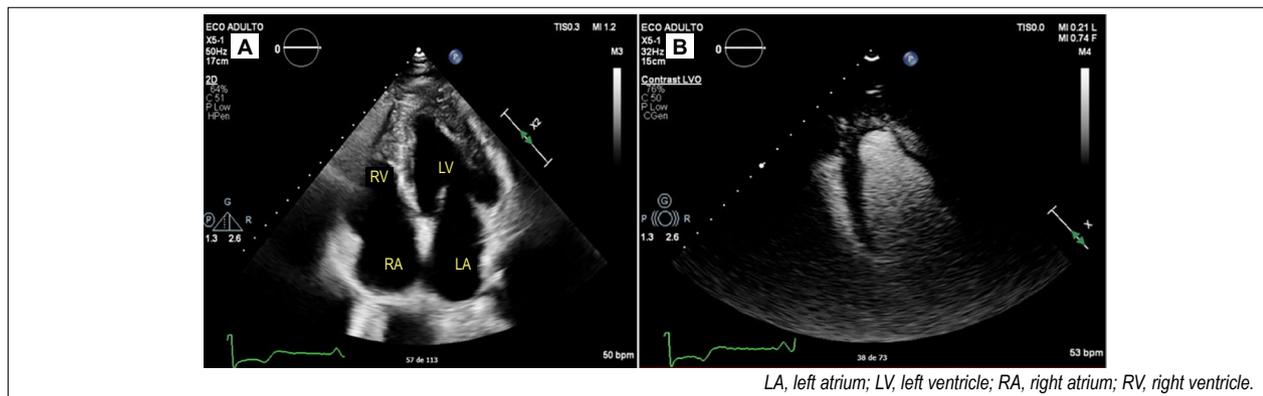


Figura 5 – (A) Apical window showing complete thrombus removal and LA diameter reduction; (B) Use of contrast to increase enhancement of the thrombus region.

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