

Right Ventricular Endomyocardial Fibrosis: Case Report

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Introduction

Endomyocardial fibrosis (EMF), first described in 1948 in Uganda, is more prevalent in countries of tropical and subtropical climates. It is characterized by the deposition of fibrous tissue in the endomyocardium of both ventricles, with less frequent right ventricular (RV) involvement.¹⁻³ We report a case of EMF involving RV in a patient with symptomatic right heart failure.

Case Report

A 46-year-old male patient from Bahia, presenting lower limb edema and ascites refractory to clinical treatment since 2016, was admitted to the Hepatology Service for etiological investigation of ascites. Physical examination revealed lower limb edema and high volume ascites, with no other abnormalities. Diagnostic paracentesis produced ascitic fluid compatible with transudate by serum-ascites albumin gradient, pelvic ultrasound with signs of chronic liver disease and upper digestive endoscopy with esophageal varices of fine caliber, compatible with portal hypertension. The laboratory tests revealed the presence of significant eosinophilia (absolute value of 370 K/uL). The patient then underwent echocardiographic evaluation, which evidenced a significant increase of right chambers and presence of hyperrefracting mass, obliterating the middle and apical portion of the RV, highly suggestive of endomyocardial fibrosis (Figure 1 and Video 1). The right atrium, with a volume of 202 mL, presented massive spontaneous contrast inside (Figure 2 and Video 2), and the RV had contractile deficit (TAPSE 8.2 mm and S' by tissue Doppler of 3 cm/s). The tricuspid and pulmonary valves presented reduced mobility, without significant regurgitation. It presented preserved left ventricular

dysfunction. Electrocardiography revealed atrial fibrillation with right bundle branch block (Figure 3). Based on the findings, he was diagnosed with cardiogenic cirrhosis secondary to RV EMF and was maintained under clinical treatment.

Conclusion

EMF is a rare disease of uncertain etiology, poor prognosis and no specific treatment to date. Multiple factors are implicated. The main one is the presence of eosinophilia.⁴ Echocardiography is the first-line method for the diagnosis of EMF, because it is inexpensive and widely available.^{5,6} However, cardiac magnetic resonance imaging using late enhancement techniques, when available, allows early diagnosis and prognostic analysis of the disease, as well as a better evaluation of morphology, fibrosis grade and ventricular function, mainly when the RV is affected. In addition, it allows a better differential diagnosis with other cardiopathies, which leads to obliteration of the left ventricular apex.^{7,8} Treatment is based on relief of symptoms and rhythm control associated with anticoagulation, if necessary, in patients with tachyarrhythmia. Surgery can be considered to increase survival. However, it has high mortality rates in the immediate postoperative period and fibrosis may recur, although this is arguable in the literature.^{1,9} This report addressed a case of EMF affecting the RV, which is less common in a symptomatic patient with atrial fibrillation and right heart failure.

Authors' contributions

Data acquisition: Carneiro SS, Costa PV, Lirio LV, Batista DCFA; Data analysis and interpretation: Carneiro SS, Costa PV; Manuscript writing: Carneiro SS, Lima AV, Lirio LV; Critical revision of the manuscript as for important intellectual content: Carneiro SS, Costa PV, Lima AV, Gomes FLT.

Keywords

Endomyocardial Fibrosis/complications; Endomyocardial Fibrosis/diagnostic, imaging; Ventricular Dysfunction, Right/complications; Eosinophilia; Heart Failure; Echocardiography. Doppler/methods; Ultrasonography/methods.

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Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate programs.

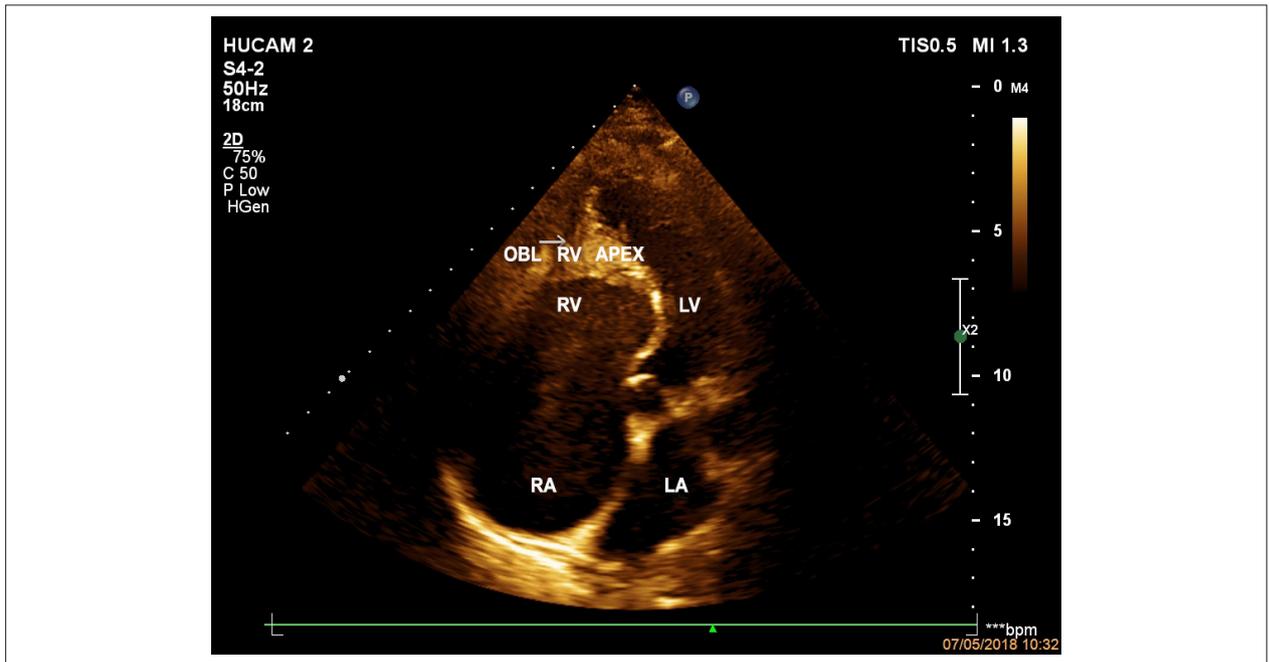
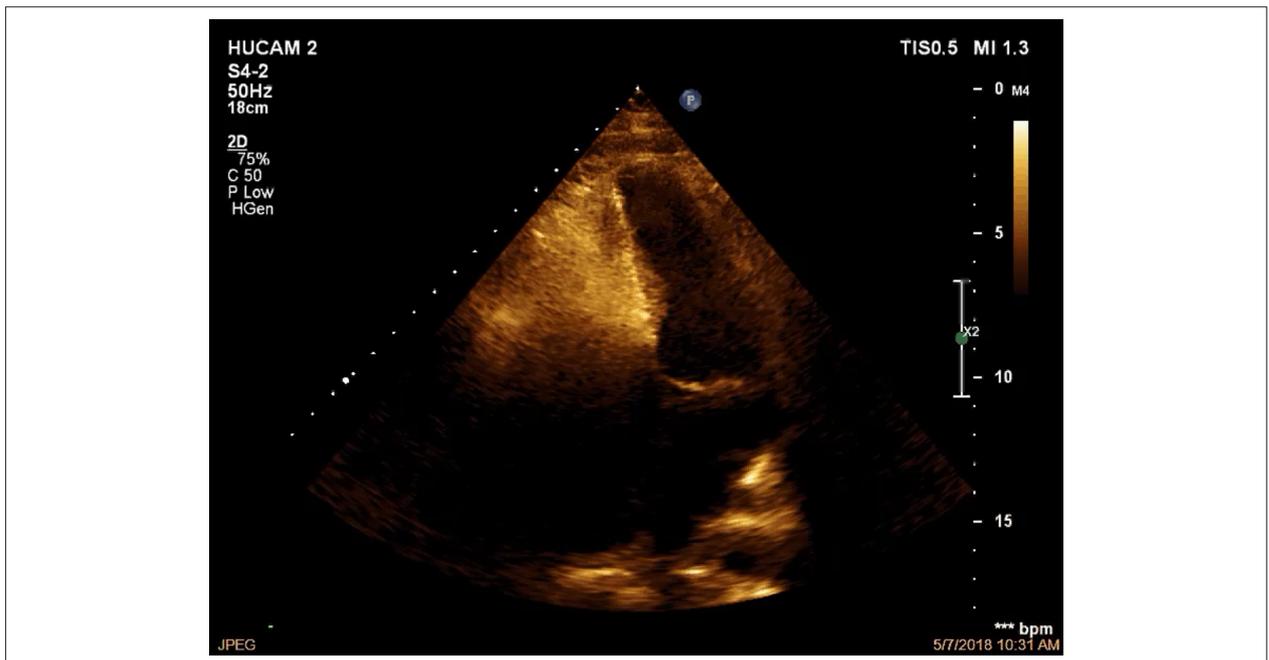


Figure 1 – Hyperrefringent image in the middle portion and at the right ventricular (RV) apex. OBL: obliteration; LV: left ventricle; LA: left atrium; RA: right atrium.



Video 1 – Apical 4-chamber view (left chambers to the right of the screen) showing dilation of the right chambers and hyperrefringent structure occupying the middle portion and the apex of the right ventricle, with spontaneous contrast in the right ventricle and anomalous movement of the interventricular septum. Poorly seen tricuspid valve. Access the video here: http://departamentos.cardiol.br/dic/publicacoes/revistadic/2019/v32_1/video_v32_1_262_ingles.asp

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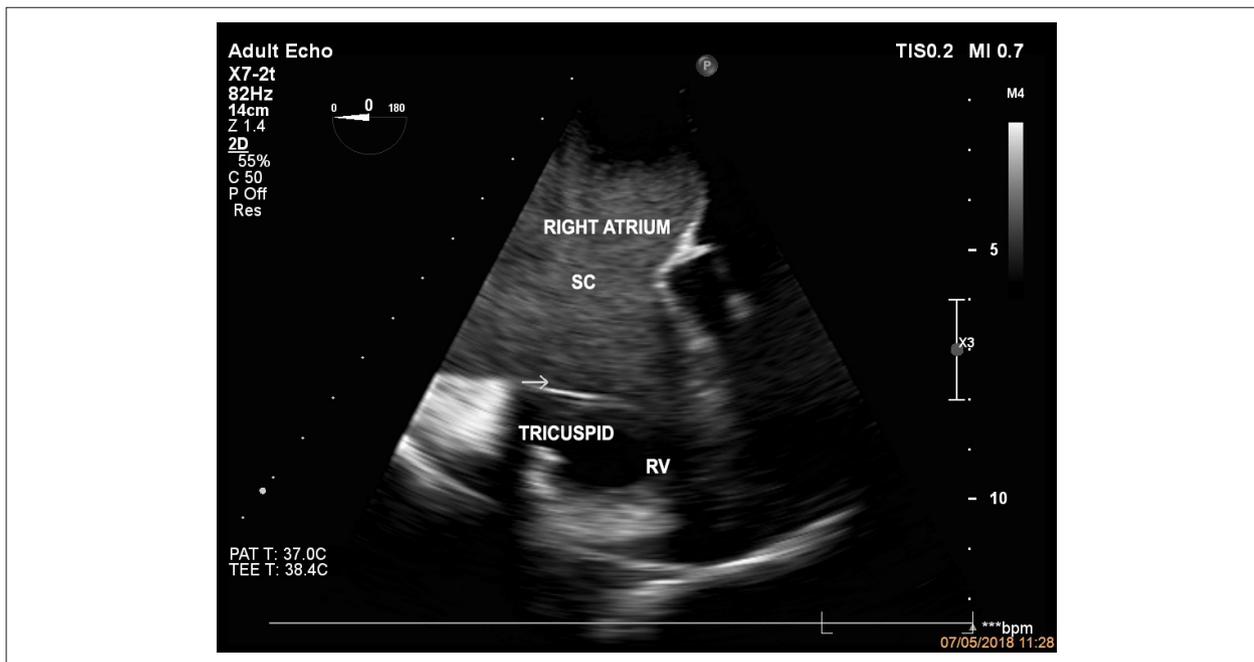
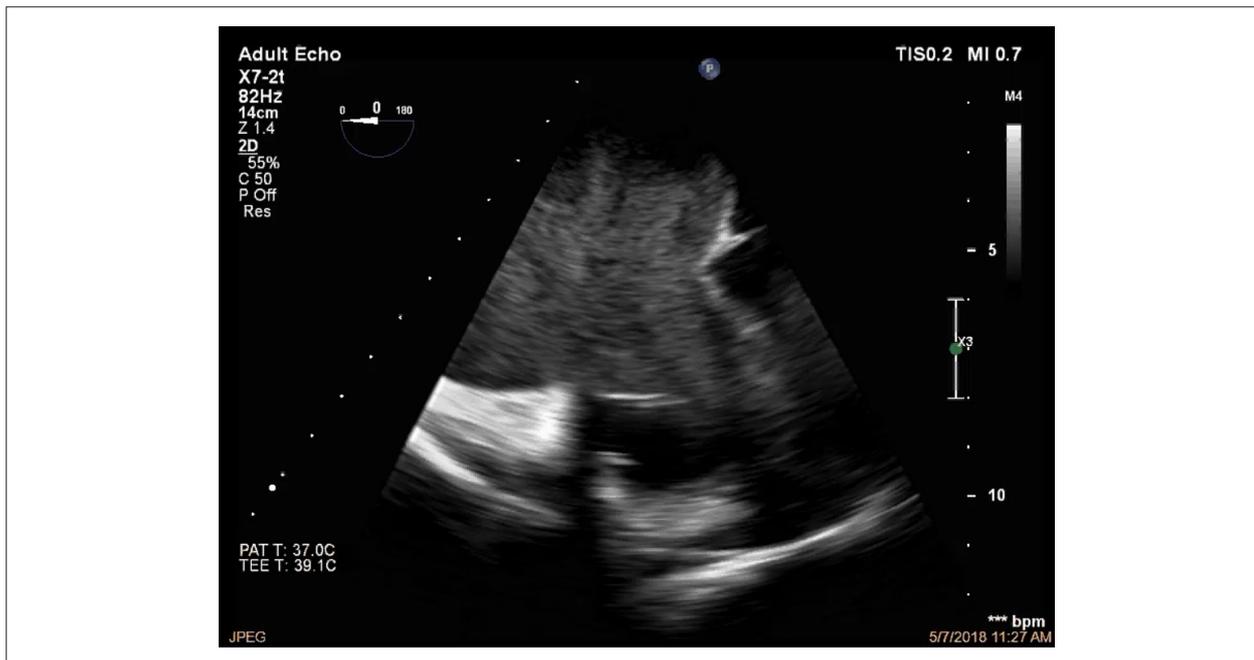


Figure 2 – Transesophageal study showing severe right atrial increase and spontaneous contrast. SC: spontaneous contrast; RV: right ventricle.



Video 2 – Transesophageal study with 0° section, focused on the right chambers, showing intense spontaneous contrast in the right atrium and right ventricular inflow tract, with thickened tricuspid valve and restricted mobility. Access the video here: http://departamentos.cardiol.br/dic/publicacoes/revistadic/2019/v32_1/video_v32_1_262_ingles.asp

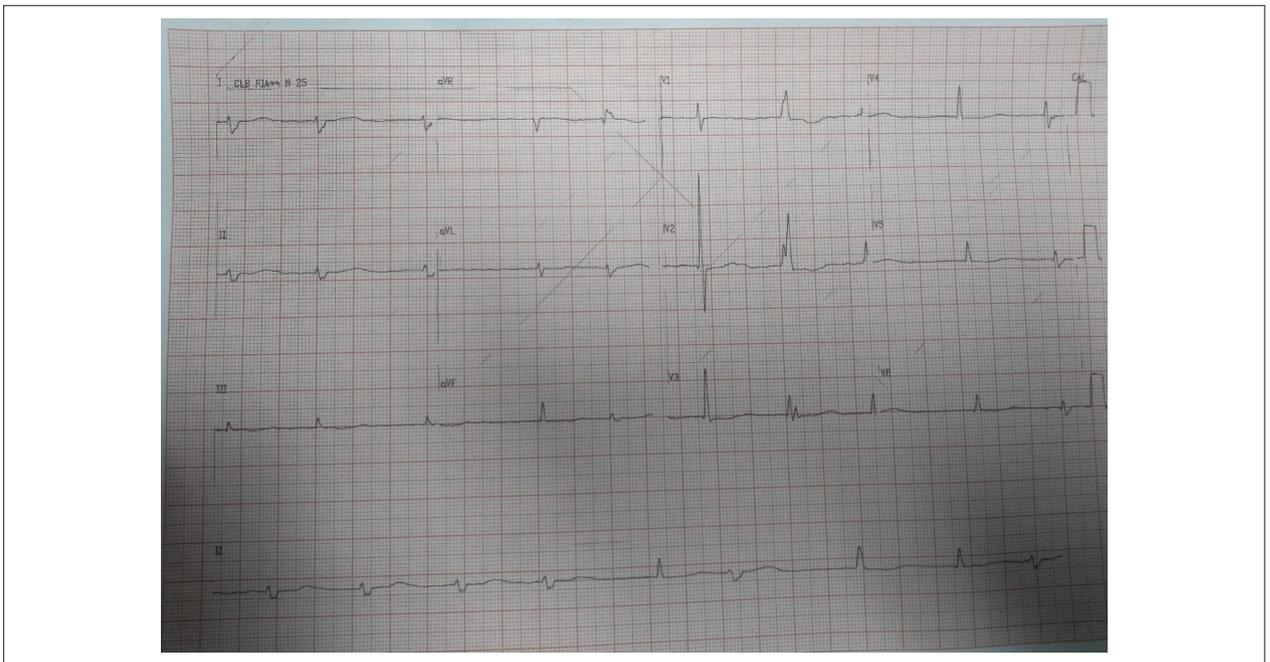


Figure 3 – Electrocardiography.

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