



## Case Report/Relato de Caso

### Endomyocardial fibrosis associated with mansoni schistosomiasis

#### Endomiocardiofibrose associada à esquistossomose mansoni

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#### ABSTRACT

Endomyocardial fibrosis (EMF) is a neglected tropical disease that affects millions of people worldwide. EMF is the most common cause of restrictive cardiomyopathy, caused by deposition of fibrous tissue on endocardial surfaces. EMF is a major cause of death in areas where it is endemic, but the pathogenesis of the disease is poorly understood. Schistosomiasis mansoni is a parasitic disease endemic in Brazil, where EMF has also been described. The association between EMF and schistosomiasis has been suggested in various publications, seeking a possible correlation between endocardial and periportal fibroses. This report describes a case of EMF associated with schistosomiasis.

**Keywords:** Endomyocardial fibrosis. Mansoni schistosomiasis. Restrictive cardiomyopathy.

#### RESUMO

A endomiocardiofibrose (EMF) é uma doença tropical, negligenciada, que afeta milhões de pessoas no mundo. É considerada a causa mais comum de cardiomiopatia restritiva, causada pela deposição de tecido fibroso em superfícies endocárdicas. Endomiocardiofibrose é uma das principais causas de morte em áreas onde é endêmica, e sua patogênese é pouco conhecida. A esquistossomose mansoni é uma doença parasitária endêmica no Brasil, onde a EMF também tem sido descrita. Tem sido apontada associação entre EMF e esquistossomose em várias publicações, buscando-se uma possível correlação entre a fibrose endocárdica e periportal. Neste relato, descreve-se um caso de EMF associado a esquistossomose.

**Palavras-chaves:** Endomiocardiofibrose. Esquistossomose mansoni. Cardiomiopatia restritiva.

#### INTRODUCTION

Endomyocardial fibrosis (EMF) is a neglected tropical disease, affecting about 10 million people worldwide<sup>1</sup>. The disease is the most common restrictive cardiomyopathy, with impaired filling of one or both ventricles caused by deposition of fibrous tissue on endocardial surfaces. Endomyocardial fibrosis is a leading cause of death in areas where it is endemic, and in its most severe form, it carries a poor prognosis, with an estimated survival of 2 years after diagnosis. The cause and pathogenesis of the disease are not understood, with several theories being considered<sup>2</sup>.

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Schistosomiasis mansoni is a parasitic disease endemic in certain regions of Brazil where EMF has also been described<sup>3</sup>. Cardiac involvement in schistosomiasis is classically described as secondary to pulmonary lesions<sup>4</sup>. The association between EMF and parasitic diseases, including schistosomiasis, has been suggested in several publications<sup>5-7</sup>, seeking a possible correlation between periportal and endocardial fibroses.

The following case report describes a case of EMF in a patient with schistosomiasis mansoni.

#### CASE REPORT

A 62-year-old woman with chronic hypertension, dyslipidemia, and hypothyroidism was admitted with heart failure to the *Hospital das Clínicas* of the *Universidade Federal de Minas Gerais*, Brazil. The patient had previous history of schistosomiasis, diagnosed by the presence of eggs of *Schistosoma mansoni* on stool testing.

On physical examination, the patient was dyspneic, with respiratory rate of 24bpm, blood pressure of 160/90mmHg, and heart rate of 68bpm. Irregular pulses and irregular heart rhythm were observed. There was lower extremity edema, and the external jugular veins were distended. Breath sounds were reduced at lung bases. The abdomen was slightly distended with the liver palpable at 3cm below the costal margin, and the spleen was not palpable.

Laboratory tests revealed blood glucose of 124mg/dL and leukocytosis with eosinophilia (584 cells/ $\mu$ L). Chest radiography showed an overall increase in the cardiac area, with signs of pulmonary congestion. The electrocardiogram showed atrial fibrillation, with right bundle branch block. Serological test for *Trypanosoma cruzi* was positive.

Echocardiogram showed an increase of the atrium with obliteration of the left ventricular apex and adjacent endocardial calcification (**Figure 1**). The left ventricular systolic function was preserved (ejection fraction of 79%), with high filling pressures (E/e' of 25). The right ventricle was slightly enlarged, with pulmonary hypertension. There was also mild mitral regurgitation, with small pericardial effusion (**Figure 2**). The patient presented progressive improvement of pulmonary congestion after conventional treatment for heart failure and was discharged after 5 days of hospitalization.

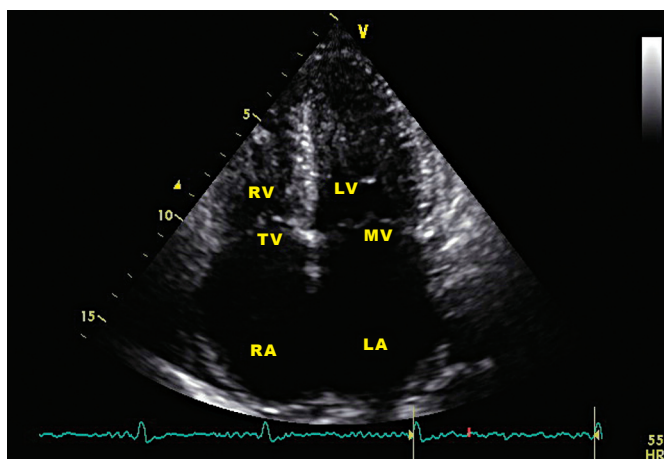


FIGURE 1 - Image from echocardiogram transthoracic, apical four-chamber view, showing endocardial thickening with obliteration of the apical region of both ventricles, especially the left ventricle.

RA: right atrium, LA: left atrium, RV: right ventricle, LV: left ventricle, MV: mitral valve, TV: tricuspid valve.

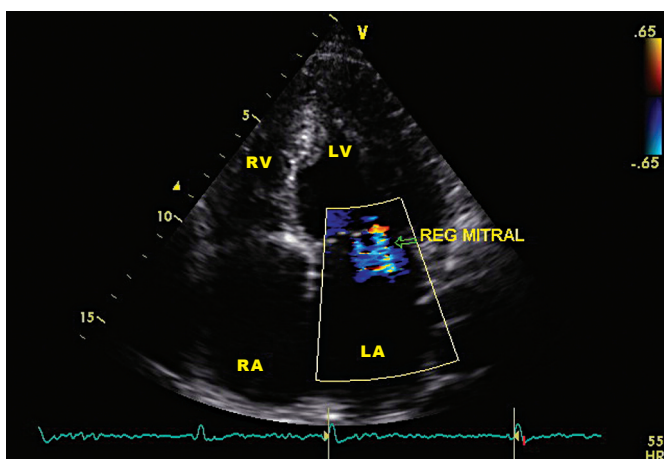


FIGURE 2 - Transthoracic echocardiogram with the use of color Doppler, apical four-chamber view, showing mitral regurgitation (arrow).

RA: right atrium, LA: left atrium, RV: right ventricle, LV: left ventricle.

## DISCUSSION

Endomyocardial fibrosis is characterized by fibrotic involvement of the endocardium and adjacent myocardium, determining a restrictive syndrome<sup>1-2, 8</sup>. Several theories are being considered to better understand the cause and pathogenesis of the disease. The accepted mechanisms are a pancarditis with eosinophilia, primarily by helminthiasis, or by immunological factors, genetic, nutritional deficiencies, or post-radiation treatment<sup>8</sup>. Endomyocardial fibrosis is an uncommon disease in Brazil, where the prevalence is higher among female than (among) male subjects. Until recently, EMF was diagnosed only in patients with advanced heart failure with a restrictive pattern of left ventricular filling. As a consequence of advances in imaging techniques, such as echocardiography, this disease can be identified earlier<sup>5</sup>. However, in the present case, the diagnosis was made only when the patient had clinical manifestation of heart failure.

The role of eosinophils as a cardiotoxic agent and precursor of fibrosis has been studied. The eosinophilic endomyocardial disease seems to be a result of prolonged release products of degranulating eosinophils<sup>8</sup>. Rasshwan et al., in an extensive echocardiographic investigation involving 10,000 patients with schistosomiasis,

had raised a supposed relationship between schistosomiasis and endomyocardial fibrosis in 15 patients<sup>9</sup>. The migration of eggs of *S. mansoni* in the human body has been described in different organs and systems, such as esophagus, spinal cord, brain, spleen, pericardium, and myocardium<sup>10</sup>.

Although the EMF is related to clinical states of hyper eosinophilia, including infestation by intestinal microorganisms, there is, usually, no evidence of parasites in lesions of the endocardium. It is assumed that endomyocardial fibrosis is a consequence of the organization of successive thrombosis, which ultimately leads to agglutination and contraction of the ventricular trabeculae. We can speculate that as a result of parasitism, eggs of *S. mansoni* can be retained in thrombotic formations, and they are involved in the repair process and may have a role in expansion of the lesion<sup>10</sup>. As it is known, the deviation of eggs of *S. mansoni* for circulation is a consequence of opening of superior porto-cava collateral circulation, accompanying portal hypertension with all its corollaries. The restriction of the disease on the right side of the heart in some cases suggests that toxins originating from the liver are released directly into the hepatic vein, inferior vena cava, right atrium, and right ventricle where they cause damage.

In conclusion, although the EMF occurs in tropical regions where there is high prevalence of parasitic diseases, there is no conclusive evidence that eosinophils induced by the parasite may explain the pathogenesis of EMF. The fact that many eosinophils are present in granulomatous reactions around eggs and worms suggests that the coexistence of schistosomiasis and EMF is not a mere coincidence. Thus, this case emphasizes the possibility of a relationship between EMF and periportal fibrosis.

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