

# Biventricular Endomyocardial Fibrosis associated with Ulcerative Colitis: a case report

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**Abstract:** Endomyocardial Fibrosis (EMF) is a restrictive cardiomyopathy characterized by the fibrotic involvement of the endomyocardium. Describe an uncommon case and the general aspects of this entity. A 13-year-old female patient was admitted with a suspect of acute gastroenteritis. Cursed with intense protein depletion and deteriorating health conditions. During the investigation was diagnosed with Ulcerative Colitis. The Doppler echocardiogram showed hyperrefringent images and a tumoration-suggestive lesion in both ventricles. The cardiac magnetic resonance confirmed the presence of tumorations in the apical portion of right and left ventricles. The patient was submitted to a biventricular resection of the masses. After the intervention, clinical therapy was started. The diagnosis of EMF was later confirmed on the histopathologic examination. The clinical presentation of EMF is unspecific and difficult to recognize, demanding deep investigation skills. The correlation with imaging exams for diagnosis is essential, especially the echocardiogram and the magnetic resonance. In this case, it is observed the association between Ulcerative Colitis and EMF, as well as the potential of the imaging modalities in the confirmation and therapeutic guidance of this pathology.

**Keywords:** Endomyocardial Fibrosis; Cardiomyopathy, Restrictive; Colitis, Ulcerative.

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## 1. Introduction

Endomyocardial fibrosis is a cardiovascular condition that is characterized by the fibrotic involvement of the endocardium and adjacent myocardium associated with calcifications and thrombi. It is a restrictive cardiomyopathy of endemic distribution in equatorial Africa, India, southeast Asia, and Latin America. In these regions, it can be considered the main cause of restrictive disease [1]. Although epidemiologic large-scale registries are limited for this condition, a global prevalence of 10-12 million people with the disease is estimated [2]. It is typically more common in the childhood and adolescence with peak incidence between ages 10-14 [3].

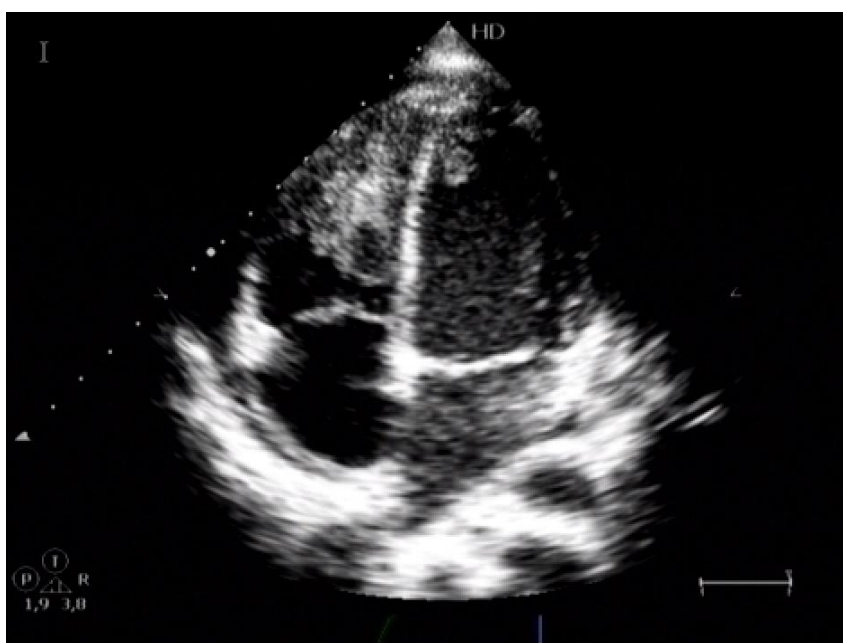
Ulcerative colitis is an inflammatory condition of the intestine localized especially in the colon. It is the most common form of intestinal inflammatory disease and is typically characterized by increased friability and erosions of the colonic wall. This inflammatory process is generally restricted to the submucosa and mucosa. There is a bimodal pattern of distribution associated with the disease: the first onset usually occurs between 15 and 30 years, whereas the later onset occurs between 50 and 70 years [3]. Ulcerative colitis has an incidence of 9 to 20 cases per 100,000 persons per year. Its prevalence is 156 to 291 cases

per 100,000 persons per year. There is no clear reference regarding prevalence and incidence of the simultaneous presentation of these conditions.

The etiology of both Endomyocardial fibrosis and Ulcerative colitis remains unknown, but some risk factors appear to have a relation to disease development and a possible connection between these pathologies: geographical location and socioeconomic status. These factors have a close relation to the three main proposed etiologies for Endomyocardial fibrosis: eosinophilia/parasitic diseases, diet/toxicity, and genetic susceptibility. It is believed that a toxic effect of the eosinophils at the myocardium is associated to immune hyperactivity and fibrosis development, especially in the early stages of the disease [4]. The association between a pro-inflammatory state generated by the inflammatory bowel disease and the development of immune hyperactivation and eosinophilia that may possibly lead to a cardiac pathology remains unknown.

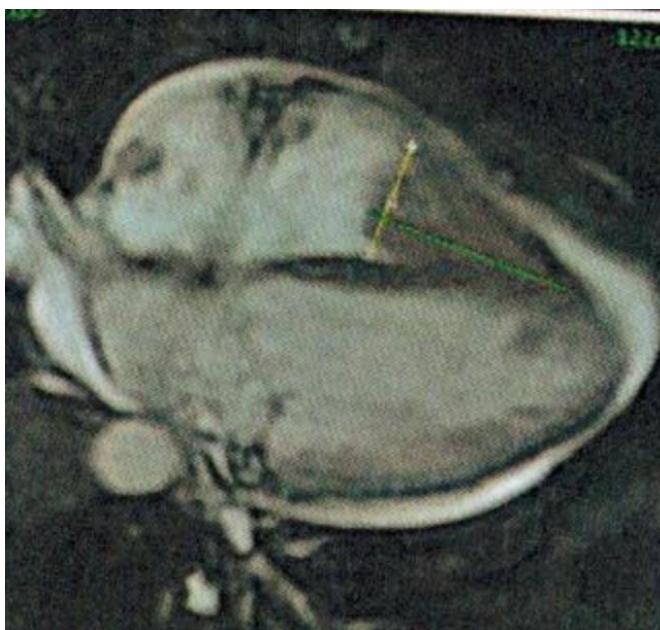
## 2. Case Report

A 13-year-old female patient reported diffuse abdominal pain associated to diarrhea, nausea, vomiting and dizziness that started 2 months prior to the current presentation. During the period, a 9kg weight loss was also reported. The patient was admitted at an inpatient facility with severe protein depletion and generalized edema. The initial investigation included a colonoscopy, which demonstrated diffuse chronic polyposis compatible with the Ulcerative Colitis diagnosis. At the time, there were no specific cardiovascular complaints. A transthoracic echocardiogram revealed important findings: mobile pediculate lesion in the left ventricle apex measuring 0,9 x 1,0 cm and another hyper refringent lesion at the right ventricle that occupied about 2/3 of its cavity (Figure 1). A mild pericardial effusion, tricuspid and mitral regurgitation were also described.



**Figure 1:** Transthoracic echocardiogram on the 4-chamber view demonstrating a tumor-like lesion at the right ventricle measuring 46,5mm and occupying 2/3 of the cavity. It also spreads to the left ventricle.

The investigation continued with a cardiac magnetic resonance (MRI) [4-6, 9, 10]. An irregular tumor-like lesion that obliterated the apical portion of the right ventricle was described. There was also close relation to the interventricular septum and left ventricle. Late gadolinium enhancement (LGE) demonstrated the presence of thrombus at the basal portion of the lesion (Figure 2). A clear pattern of LGE that represented endocardial fibrosis, strongly suggested the diagnosis of Endomyocardial fibrosis.



**Figure 2:** Cardiac MRI findings were highly suggestive of Endomyocardial fibrosis.

The gastrointestinal symptoms were controlled after institution of directed medical therapy, including salicylates and disease-modifying agents. Regarding the management of the cardiovascular findings, surgical approach was indicated. Treatment options for the endomyocardial disease included conservative management and surgical approach. In this case, we opted for the later considering the disease extension and impairment of RV filling. The patient was also at a low-surgical risk group. The selected procedure was a biventricular resection of the lesion.

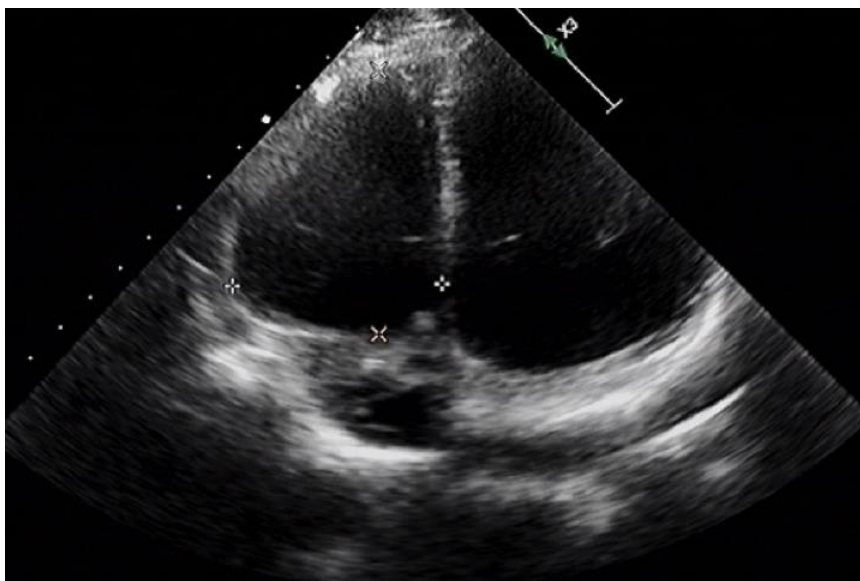
There were no major complications, post-operative period went uneventful, and the patient was discharged for outpatient follow-up. Histopathology report demonstrated degenerative changes, fibrous reaction, hyalinization, spumes and pigmented histiocytes. These findings were conclusive for the initial hypothesis of Endomyocardial fibrosis. On the first post-operative appointment there were no specific signs and symptoms. A control transthoracic echocardiogram was performed (Figure 3) with a significant improvement if compared to the previous study.

A 24-hour Holter monitoring demonstrated non-sustained ventricular tachycardia (Figure 4). The patient received beta-blocker and remained in asymptomatic for over 2 years with no relapse of disease. There was an overall improvement of the perceived quality of life, as reported by the patient. In terms of long-term follow-up, it is believed that remission of the gastrointestinal condition is a critical component in management. We also hypothesize that acute flares of the disease may lead to increased inflammation and eosinophilia that can predispose recurrence of cardiac involvement. The follow-up implemented after the first two years in this case consists in annual appointments in Cardiology and Gastroenterology with imaging exams requested at the providers discretion and according to patients' complaints.

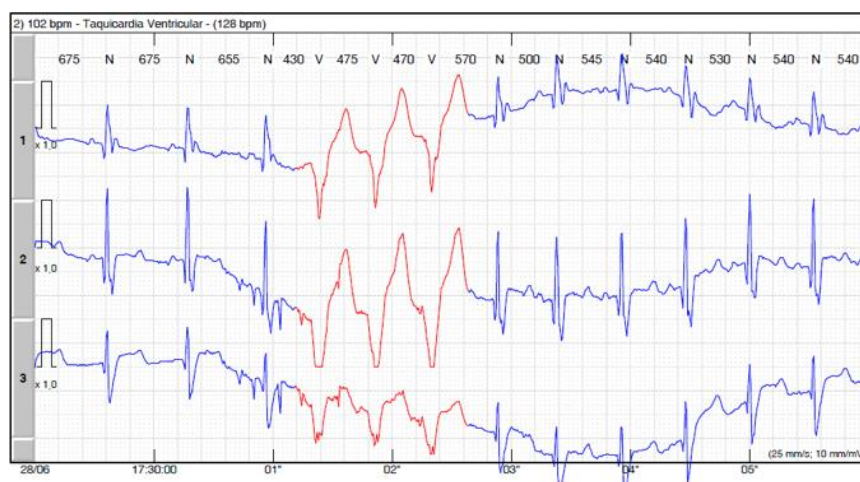
### 3. Discussion and conclusion

Most of the patients with the condition are completely asymptomatic, which contributes to a delayed diagnosis. The disease development usually occurs in three phases: active inflammatory state, transitional period, and chronic fibrotic state. The typical clinical presentation of the disease occurs particularly in the advanced stages (chronic fibrotic state) with cachexia, malnourishment, hypoalbuminemia, and low cardiac output. Cardiac involvement is a pancarditis with endocardial fibrosis and diastolic dysfunction. In 55% of patients there is biventricular disease [2]. The restrictive condition results in cavity

enlargement, which facilitates arrhythmias and electrical disturbances [5]. The cardiac injury mediated by eosinophils progresses in three stages. The early necrotic form is usually silent and can be diagnosed by MRI [9,10].



**Figure 3:** Control transthoracic echocardiogram with complete resolution after lesion resection.



**Figure 4:** Non-sustained ventricular tachycardia on 24-hour Holter monitoring.

In the intermediate phase, which is characterized by endocardial damage, there is overt thrombosis and increased risk of embolization. Finally, a fibrotic stage, with fibro-inflammatory remodeling of valve structures and chordae tendineae, ultimately leads to ventricular heart failure<sup>6</sup>. The diagnosis requires a throughout investigation that includes full blood tests, electrocardiogram, chest radiography, transthoracic echocardiogram and cardiac magnetic resonance, if available. The initial choice is usually the echocardiogram, a cheap non-invasive method with an elevated degree of specificity (82,8%) [5, 11]. Not only it can indicate the degree and extent of fibrotic involvement, but it can also measure the hemodynamic compromise of the disease.

Routine imaging with other advanced modalities such as cardiac MRI is not mandatory, but if available it can contribute with important additional information such as the presence of thrombi and a specific LGE pattern [9,10]. Medical management includes basically control of heart failure symptoms and other associated conditions like arrhythmias.

Surgical procedures are usually indicated for symptomatic patients and/or those with high-risk situations, such as biventricular involvement, hemodynamic compromise, and large lesions. Despite the recent advancements in therapy and the increased availability of imaging methods, prognosis is still reserved with elevated mortality and morbidity. This is partially explained by delayed diagnosis [7].

In this case report, we hypothesized that there is an association between Endomyocardial Fibrosis and Ulcerative Colitis (chronic inflammatory intestinal disease). The presence of hypereosinophilia in patients with Ulcerative colitis is considered a marker of disease severity at diagnosis [3]. Common aspects for both conditions include immune hyperactivity and eosinophilia [6]. Therefore, the gastrointestinal condition may be considered a predisposing factor that have aggravated the cardiac condition. The association between a pro-inflammatory state generated by the inflammatory bowel disease and the development of immune hyperactivation and eosinophilia that may possibly lead to a cardiac pathology remains unproven but is a plausible pathophysiologic explanation for the case presented in this paper [3].

As it is an essentially tropical pathology that mainly affects regions of low socioeconomic development, diagnosis is generally late and treatment options are limited, contributing to high mortality and morbidity rates [8]. The recognition in its initial stages represents a challenge to clinical practice. It is necessary to increase interest and knowledge about this disease to reduce the serious social, psychological and economic impacts that it confers [1]. In view of the above, Endomyocardial fibrosis shares several pathophysiologic and epidemiological aspects with Ulcerative colitis. The association between these conditions remains relatively underrepresented and requires further research and investigation of the specific mechanisms that may contribute to symptom development and overlapping between the cardiac and gastrointestinal diseases.

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**Research Ethics Committee Approval:** We declare that the patient approved the study by signing an informed consent form and the study followed the ethical guidelines established by the Declaration of Helsinki.

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**Conflicts of Interest:** None.

**Supplementary Materials:** None.

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