AUCTORES
Globalize your Research

Journal of Thoracic Disease and Cardiothoracic Surgery
Raquel Reis Soares *

Open Access

Case Report

Left Ventricle Endomyocardial Fibrosis : A Case Report

Raquel Reis Soares [1*], Maria Clara Martins Avelar [2], Sofia Lucena Zanetti [2], Joao Victor Tavares Mendonça Garreto [3], Vinicius Dinelli Guimaraes [3], Elisa Soares Ferber [4], Mayumi de Oliveira Drumond [5], Matheus Ferber [6], Leonardo Ferber [7]

[1] Cardiovascular anesthesiologist at Biocor Rede D'Or Institute.

[2] Medical student at Faculdade Ciências Médicas de Minas Gerais.

[3] Cardiovascular surgery Resident at Biocor Rede D'Or Institute.

[4] Medical student at Universidade José do Rosário Vellano – UNIFENAS.

[5] Medical student at Universidade Federal dos Vales do Jequitinhonha e Mucuri.

[6] Cardiovascular Surgeon at Biocor Rede D'Or Institute.

[7] Cardiovascular Surgeon at Hospital das Clinicas da Universidade Federal de Minas Gerais.

Corresponding Author: Raquel Reis Soares, Cardiovascular anesthesiologist at Biocor Rede D'Or Institute.

Received date: March 05, 2023; Accepted date: March 17, 2023; Published date: March 29, 2023

Citation: Raquel R. Soares., Martins Avelar MC., Sofia L. Zanetti, Mendonça Garreto JVT., et all. (2023). Left Ventricle Endomyocardial Fibrosis :A Case Report, *J Thoracic Disease and Cardiothoracic Surgery*, 4(2); **DOI:**10.31579/2693-2156/046

Abstract

Background: Endomyocardial fibrosis (EMF) is a grim disease. It is the most common restrictive cardiomyopathy worldwide,1 but its exact etiology and pathogenesis remain unknown. It seems likely that dietary, environmental, and infectious factors may combine in a susceptible individual to give rise to an inflammatory process leading to endomyocardial damage and scar formation. 2 An extensive ventricular endocardium fibrosis distorts ventricular architecture and leads to a restrictive physiology. Impaired filling and valvular insufficiency define the disease.1 Many times it is associated to hypereosinophilic endocarditis. This tropical cardiomyopathy presents most cases in Africa, Asia, and South America.2

Case presentation: We present a 67-year-old male patient with restrictive cardiomyopathy and mitral valve insufficiency due to EMF. The patient was referred a history of schistosome infection in the childhood. He has presented no other risk factors described in the literature. After treatment with diuretics and vasodilators, he underwent a successful surgical resection of endocardial ventricular fibrosis, mitral valve replacement and two saphenous bypasses.

Conclusions: The aim of this case report is to highlight the disease and to show how challenging is the endomyocardium fibrosis's treatment and the importance of an earlier diagnosis as well as more researches on this topic.

Keywords: restrictive cardiomyopathy; endomyocardial fibrosis; heart failure cardiac surgery; hypereosinophilia; cardiac magnetic resonance

Introduction:

Previously known as "Davies disease", EMF is the most frequent restrictive cardiomyopathy worldwide with higher prevalence in sub-Saharan Africa, Asia, and South America, responsible for 10-20% of deaths by heart failure in Africa.[1,3,4] It is a chronic and insidious disease responsible for endocardial thickening, cardiac morphological modifications, reduced ventricular complacency and heart failure. Fibrous infiltrates deposit in the ventricular apex and increase atria size due to severe diastolic dysfunction and restrictive ventricular disorder.[4,5] A study conducted in Mozambique based on a sample of 1.063 individuals estimated EMF's global prevalence of 19,8%, affecting predominantly males between 10-19 years with biventricular involvement. 22% of the affected patients presented symptoms.[6]

EMF is a restrictive cardiomyopathy characterized by reduced ventricular complacency and diastolic dysfunction due to deposition of fibrous tissue in the endomyocardium, manifesting symptoms of heart failure.[1,2] EMF is a neglected disease and its insidious expression often leads to a

late diagnosis and a poor prognosis.[2] While the physiopathology remains unclear, EMF is recurrently associated to chronic eosinophilia what permits to consider endemic parasitosis (schistosomiasis, filariasis, malaria, and helminths) as a potential trigger. Other triggers would be genetics, autoimmune or nutritional due to the higher prevalence in tropical low-income countries.[2]

Case Presentation:

A 67-year-old male patient was admitted to the hospital due to decompensated heart failure.

The patient was referred a history of progressive dyspnea triggered by less than ordinary activities and lower-extremity edema throughout 1 year. He denied hypertension, dyslipidemia, diabetes, chest pain, myocardial infarction or stroke. The patient reported schistosome infection in the childhood. All his laboratory tests were normal.

A cardiac magnetic resonance imaging (MRI) revealed apical left ventricle wall thickening with left ventricular apical obliteration associated with enlargement of the respective atrium. Delayed enhancement imaging showed endomyocardium enhancement involving left ventricular apex, mitral valve regurgitation due to annulus dilation and a thrombus at left ventricular apex. (Figure 1) (Figure 2)

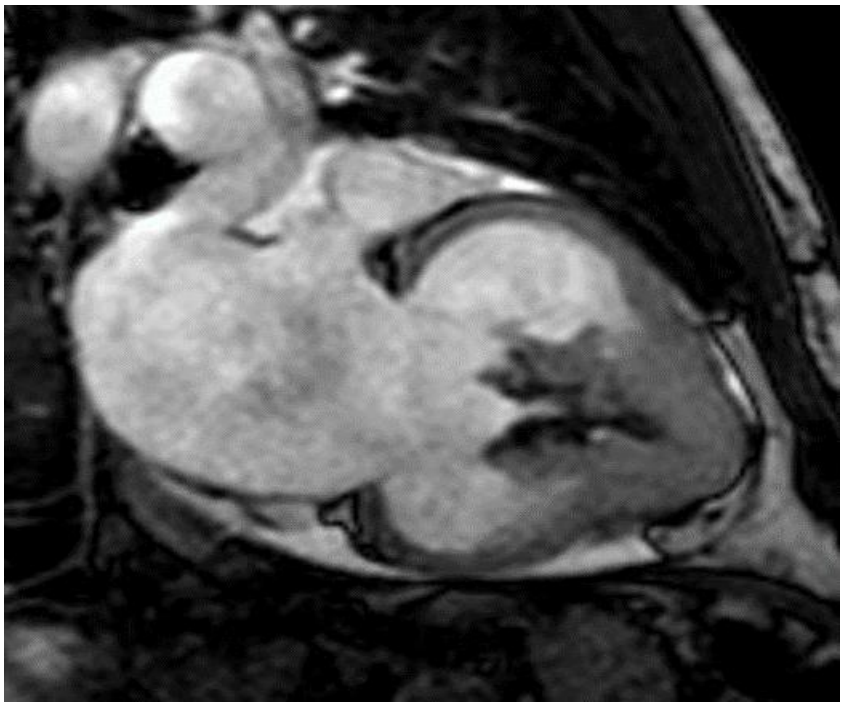

Figure 1: Cardiac MRI showing obliteration of the entire left ventricular apex.

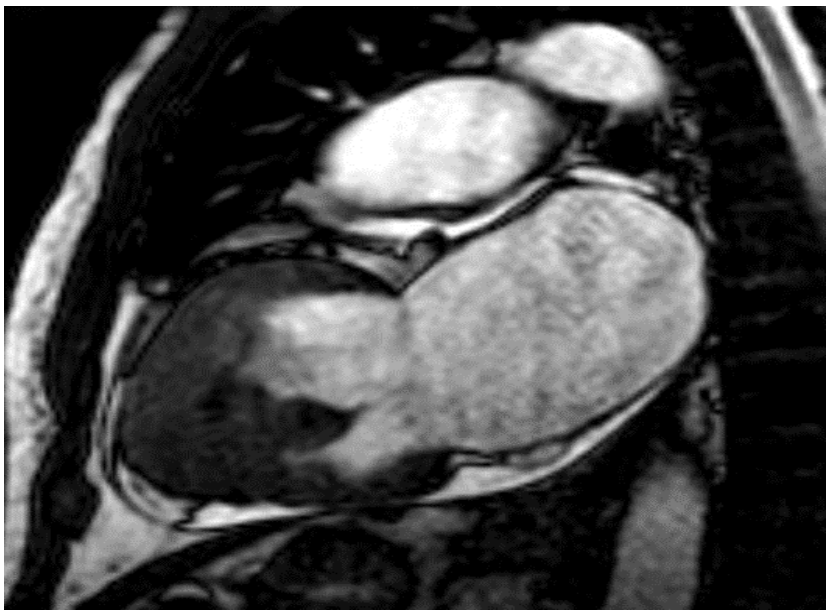

Figure 2: Delayed enhancement revealing endomyocardial fibrosis in the apical portions of all the left ventricle walls.

The coronary angiography showed severe lesion in the right coronary artery and in anterior descending artery.

The final diagnosis was left ventricle endomyocardium fibrosis (EMF), mitral valve insufficiency, thrombus at left ventricular apex and coronary artery insufficiency.

After a multidisciplinary consideration, surgical treatment was indicated. The patient was submitted to a cardiac surgery with cardiopulmonary bypass by median sternotomy, aortic, superior and inferior vena cava cannulation and anterograde cardioplegia. The procedure was done without problems and the patient was sent to the intensive care unit (ICU) using low doses of norepinephrine and dobutamine to improve cardiac function. The drugs were required in low dose for 5 days, the patient had a good outcome and was discharged from the ICU in 7 days. He was discharged from hospital 11 days after surgery.

The postoperative control transthoracic echocardiography showed a normal functioning mitral valve, without obstruction of the left ventricle outflow tract.

Tissue *fragments* recovered at *cardiac* surgery showed the endocardium thickening and fibrous extend into the underlying myocardium with calcified areas of left ventricle and septum.

Histologically the endomyocardium and mitral valve fragments presented areas of fibrosis, hyalinization and *vascular* tissue *neoformation*, multiple foci of mononuclear inflammatory infiltrate, and calcification areas. The biopsies were compatible with *the diagnostic suspicion* (endomyocarium fibrosis).

Discussion And Conclusions

Previously known as "Davies disease", EMF is the most frequent restrictive cardiomyopathy worldwide with higher prevalence in sub-Saharan Africa, Asia, and South America, responsible for 10-20% of deaths by heart failure in Africa.[1,3,4] It is a chronic and insidious disease responsible for endocardial thickening, cardiac morphological modifications, reduced ventricular complacency and heart failure. Fibrous infiltrates deposit in the ventricular apex and increase atria size due to severe diastolic dysfunction and restrictive ventricular disorder.[4,5] A study conducted in Mozambique based on a sample of 1.063 individuals estimated EMF's global prevalence of 19,8%, affecting predominantly males between 10-19 years with biventricular involvement. 22% of the affected patients presented symptoms.[6]

Many theories have been proposed to explain the etiopathogenesis of EMF, as viral infections, allergies associated with eosinophilia and autoimmune reactions, malnutrition and exposure to toxic chemicals. The relation between EMF and Loeffler syndrome (in which eosinophils gather in the pulmonary tissue in regards to a parasitic infection) and the prevalence of parasitosis in endemic countries strengthened the association between eosinophilia and myocardial lesions.[2] Approximately 40 to 50% of patients with eosinophilic syndrome develop heart conditions, such as Loeffler's endocarditis, in which the hypereosinophilic reaction is responsible for fibrosis and necrosis of the endocardium and leads to endomyocardial fibrosis.[7] However, this theory is limited by geographic inconsistency with important parasitic prevalence and insignificant difference in parasitic loads when comparing the control group to EMF patients.[2,4,8] More studies are needed to reassure a strong scientific base towards EMF's etiology.

Early stages usually manifest with fever, pancarditis, dyspnea, itching, and periorbital swelling.[2] Eosinophilia can be present mainly during the early stages of EMF.[9]

Abnormal increase of collagen deposition, specially type-1, and proliferation of fibroblasts cause myocardial rigidification and diastolic ventricular dysfunction.[3] If tendinous cords and papillary muscles are also affected, atrioventricular valve regurgitation can occur. Atrial fibrillation, conductive disorders and ventricular arrhythmias are detected in the electrocardiogram in 30% of cases. [5,10] The first necrotic stage called inflammatory phase is followed by mural thrombi generation and the risk of thromboembolic complications. Endocardial calcification and valvar incompetence are common in the final stage.[11] Symptomatic heart failure is treated with diuretics, vasodilators and beta-blockers, depending on the disease presentation.

A score was developed by Mocumbi and col. in 2018 to guide EMF's patient's diagnosis and prognosis.[5,6] The presence of two major criteria or one major and two minor criteria is considered as a diagnostic proof of the disease. The prognosis is based on the scores, being poor when ≥ 15 points and better if < 8 point.[6] Among the major criteria are: "obliteration of the right ventricular or left ventricular apex", "thrombi or spontaneous contrast without severe ventricular dysfunction", "retraction of the right ventricular apex" and "atrioventricular valve dysfunction due to adhesion of the valvular apparatus to the ventricular wall". The minor criteria are "restrictive flow pattern across mitral or tricuspid valves", "pulmonary valve diastolic opening" and "enlarged atrium with normal-size ventricle".[6]

EMF's diagnosis can be guided by echocardiographic evidences of thrombotic and fibrotic obliteration of the ventricular apex, increase of the endocardial diameter and valvar insufficiency.[5,12] Differential diagnosis with other restrictive cardiomyopathies is substantial for the treatment's guidance and can be better achieved by cardiac MRI using gadolinium-based contrast. MRI provides accurate morphological evaluation by assessing diastolic and systolic functions, tissue characterization and stratifying prognosis of most cardiomyopathies. Delayed enhancement imaging helps to identify the presence of fibrosis.[7]

In this case we presented a patient that was classified as NYHA III - New York Heart Association Classification of Heart Failure (comfortable at rest; less than ordinary physical activity causes fatigue, dyspnea, palpitations) and who had poor response to pharmacological treatment. He was in the final stage of the disease and the left ventricular endomyocardium was replaced by fibrosis, as well as the mitral valve tendinous cords. The final result was a restrictive dilated cardiomyopathy and mitral valve incompetence. Superimposed thrombosis and endocardial calcification were present.

To improve his clinical condition the endomyocardial resection and valve replacement were performed after a multidisciplinary consideration, aiming to increase the end systolic and diastolic diameter and to avoid left ventricle diastolic dysfunction.

There is no specific drug therapy for EMF. Surgical intervention is reserved for more advanced cases (patients graded NYHA functional classification III and IV) as it has a high postoperative mortality.[4,5] If the left ventricle is affected, the severity of the mitral insufficiency and pulmonary hypertension establish surgical indication.[12] Despite the fact that Right ventricular EMF is the most common site of presentation either in isolation or as part of the biventricular disease, our patient had a Left ventricular form of the disease. Involvement of both ventricle, right ventricle fibrosis, and tricuspid or mitral regurgitation is associated with a higher mortality rate. Ascites is observed in half of cases of EMF and it is associated with greater involvement of the right ventricle or a longer duration of the disease. It defines a worse prognosis. The ascites without peripheral edema appears to be caused by peritoneal inflammation with increased sequestration of fluid as well as fibrosis with decreased reabsorption.[13]

Surgical intervention is expected to increase the survival when is undertaken before the development of irreversible cardiac and hepatic damage. It requires experienced operators and well-equipped cardiac facilities.[14]

The imaging techniques progress, detecting myocardial damage, contribute to earlier diagnosis and survival in EMF patients.[14] Surgical intervention, in a less advanced stage of the disease, may improve quality of life and prognosis of the patient, but it is not curative.[14,15]

Drugs that block different pathways of eosinophilic inflammation altering immunologic mechanism of hypereosinophilic syndromes make the

monoclonal antibody Imatinibe a promising potential treatment for EMF, but they are ineffective when the heart failure is present.[5]
The EMF remains a challenging disease, it is mandatory a better comprehension of its physiopathology to find answers for its treatment.

List of Abbreviations

Endomyocardial fibrosis - EMF
Magnetic resonance imaging - MRI
Intensive care unit - ICU
New York Heart Association - NYHA

Declaration

We declare for whom it may concern that written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

We also confirm that the disclosed information is correct and that no

other situation of real, potential or apparent conflict of interest is known to us.

We confirm that we do not have a financial or other interest in the subject of the work

we wrote, which may be considered as constituting a real, potential or apparent

Conflict of Interest.

We also afirm that we have not received any funds to write the: " Left ventricle endomyocardium fibrosis - a case report".

Every author contribute to make this article and all of them are responsible for writing the paper.

Ethical Approval

This study was approved by the Ethical Committee of Cardiovascular Department of Biocor Institute, Nova Lima , Minas Gerais, Brasil.

Availability of data and materials

Every data and materials' statement can be found in our institution archives and in the bibliographic references cited in the text.

Competing interests

The authors declare that they have no competing interests.

Funding

The authors declare that they have no sources of funding for the research

Author's contributions

MF, LF, ESF analyzed the surgery indication and the clinical evolution. JVG, VD, MD analyzed the patient's images and laboratorial tests. RRS, SZ, MCA were major contributor in writing the manuscript. All authors read and approved the final manuscript.

Acknowledgements

Not applicable

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

1. Lachaud M, Lachaud C, Sidi D, Menete A, Jouven X, Marijon E, Ferreira B. (2018) Fibrose endomyocardique tropicale: perspectives [Tropical endomyocardial fibrosis: Perspectives]. Ann Cardiol Angeiol (Paris). 67(2):74-81.
2. Grimaldi A, Mocumbi A, Freers J, Lachaud M, Mirabel M, Ferreira B,et all (2016), Tropical Endomyocardial Fibrosis. American Heart Association. Circulation. 133(24): 2503-2515.
3. Duraes AR, de Souza Lima Bitar Y, Roe ver L, Neto MG. (2020) Endomyocardial fibrosis: past, present, and future. Heart Fail Rev. 25(5):725-730.
4. Lino D, Filho F, Néri A, Silva F. (2011) Endomyocardial Fibrosis as a Rare Cause of Mitral Insufficiency: case study and literature review. Rev Bras Cardiol. 24(4):254-257.
5. Scatularo CE, Posada Martínez EL, Saldarriaga C, Ballesteros OA, Baranchuk A, Sosa Liprandi A, Wyss F, Sosa Liprandi MI. (2021) Endomyocardiofibrosis: A Systematic Review. Curr Probl Cardiol. 46(4):100784.
6. Mocumbi A, Ferreira M, Sidi D, Yacoub M. (2008) A population study of endomyocardial fibrosis in a rural area of Mozambique. N Engl J Med. 3;359(1):43-49.
7. Kharabish A, Haroun D. (2015) Cardiac MRI findings of endomyocardial fibrosis (Loeffler's endocarditis) in a patient with rheumatoid arthritis. J Saudi Heart Assoc. 27(2):127-131.
8. Bukhman G, Ziegler J, Parry E. (2008) Endomyocardial fibrosis: still a mystery after 60 years. PLoS Negl Trop Dis. 27;2(2):e97.
9. Andy JJ, Ogunowo PO, Akpan NA, et al. (1998) Helminth associated hypereosinophilia and tropical endomyocardial fibrosis (EMF) in Nigeria. Acta Trop 69(2):127–140.
10. Disertori M, Rigoni M, Pace N, et al. (2016) Myocardial Fibrosis Assessment by LGE Is a Powerful Predictor of Ventricular Tachyarrhythmias in Ischemic and Nonischemic LV Dysfunction. J Am Coll Cardiol Img. 9 (9) 1046–1055
11. Jallal H, Belabbes S, Khatouri A. (2022) An unusual tropical endomyocardial fibrosis: a case report. Pan Afr Med J. 4;41:48
12. Almeira D, Silveira J, Brito F. (2011) Restrictive cardiomyopathies. Rev Soc Cardiol Estado de São Paulo. 21(1):14-20 RSCESP (72594).
13. Barretto AC, da Luz PL, de Oliveira SA, Stolf NA, Mady C,Bellotti G, Jatene AD, Pileggi F (1989) Determinants of survival in endomyocardial fibrosis. Circulation 80(3 Pt 1):I177–I182
14. Mocumbi, A.O., Stothard, J.R., Correia-de-Sá, P. *et al.* (2019) Endomyocardial Fibrosis: an Update After 70 Years. *Curr Cardiol Rep* 21, 148.
15. Salemi VM, Rochitte CE, Shiozaki AA, Andrade JM, Parga JR, et al. (2011) Late gadolinium enhancement magnetic resonance imaging in the diagnosis and prognosis of endomyocardial fibrosis patients. Circ Cardiovasc Imaging. 4(3):304-311.

DOI: 10.31579/2693-2156/046