



Right ventricular endomyocardial fibrosis – A case report

Deepak Madi, Basavaprabhu Achappa, Narasimha Pai, Padmanabha Kamath

Kasturba Medical College Hospital, Mangalore, Affiliated to Manipal University

CASE REPORT

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Corresponding Author:

Basavaprabhu Achappa
Department of General Medicine,
Kasturba Medical College, Mangalore, India
Affiliated to Manipal University
[Email:bachu1504@yahoo.co.in](mailto:bachu1504@yahoo.co.in)

Abstract

Endomyocardial fibrosis (EMF) is a progressive type of restrictive cardiomyopathy. It affects inflow portion of right and/or left ventricle and apex. It is a neglected tropical disease. Here we report a rare case of right ventricular endomyocardial fibrosis. A 70-year-old female presented to us with history suggestive of right-sided heart failure of two months duration. There was no eosinophilia. Chest X-ray showed cardiomegaly. Echocardiogram showed dilated right atrium and obliteration of the apex of the right ventricle. A diagnosis of Right ventricular Endomyocardial fibrosis was made. She was treated with diuretics and anticoagulants and she improved.

Key Words

Endomyocardial Fibrosis; Restrictive cardiomyopathy; Right heart failure.

Implications for Practice

1. **What is known about such cases?** Endomyocardial fibrosis is the commonest cause of restrictive cardiomyopathy worldwide. It usually affects children and young adults. Overall prognosis of Endomyocardial fibrosis is poor.

2. **What is the key finding reported in this case report** We report a rare case of isolated right ventricular endomyocardial fibrosis in an elderly female without eosinophilia.

3. What are the implications for future practice?

Endomyocardial fibrosis must figure in the list of differential diagnosis of heart failure in tropical countries. Endomyocardial fibrosis can mimic other common conditions like Constrictive pericarditis, Rheumatic heart disease and Ebstein's anomaly.

Background

Endomyocardial fibrosis (EMF) is a progressive type of restrictive cardiomyopathy. It affects the inflow portion of right and/or left ventricle and apex. Myocardial fibrosis (due to collagen deposition and fibroblast proliferation) is responsible for the restrictive physiology. It can present as right heart failure, left heart failure or bi-ventricular failure. The highest prevalence of this condition is seen in regions of sub-Saharan Africa.¹ Cases of EMF have been reported from south India.² We report a rare case of right ventricular endomyocardial fibrosis from south India. We used Echocardiography as a non-invasive investigative tool to diagnose EMF in our case.

Case details

A 70-year-old female came to our hospital with complaints of pedal oedema of two month duration. She also had exertional breathlessness. There was no history of similar complaints in the family. She was diagnosed to have a heart problem, was treated with diuretics, digoxin and she was referred to our institution for further management. Past history of diabetes, hypertension and stroke was present.

On examination she had elevated jugular venous pressure, pulse rate of 48 per minute and bilateral pitting pedal oedema. Cardiovascular examination revealed pansystolic murmur of tricuspid regurgitation. She also had hepatomegaly.

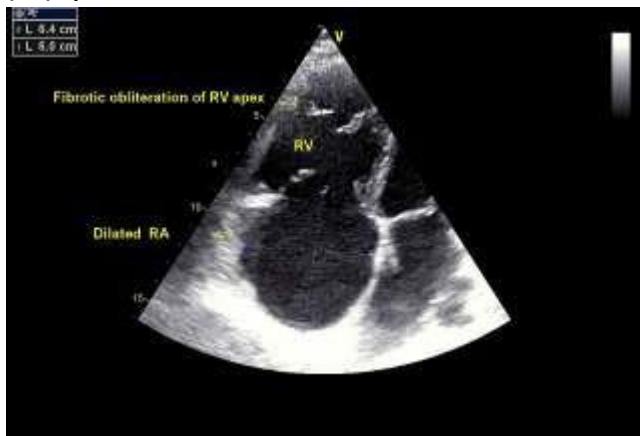
Lab investigations showed: Hb-10.8gm/dl, TC: 4,500/ cm². Differential count – N59, L29, E2, M10 and Platelets: 2,66,000/cm². Electrocardiogram showed junctional rhythm with evidence of digoxin effect. Chest radiography revealed cardiomegaly (Figure 1). Echocardiogram showed grossly dilated right atrium, mild dilated right ventricle, tricuspid regurgitation and obliteration of the apex of the right

ventricle (Figure 2). Restrictive flow pattern across mitral and tricuspid valve was present. Our patient had one major criteria (obliteration of the apex of the right ventricle) and two minor criteria (dilated right atrium and restrictive flow pattern across mitral and tricuspid valve). So a diagnosis of Right ventricular Endomyocardial fibrosis was made (Table 1).

Figure 1: Chest X-ray showing cardiomegaly



Figure 2: Echocardiogram shows grossly dilated right atrium (RA) and fibrotic obliteration of right ventricular (RV) apex.



She was treated conservatively with diuretics. Pedal oedema decreased and she felt better. Digoxin was stopped. She was also started on tablet warfarin. Insulin was administered for diabetes. She is on regular follow up for the past four months.

Discussion

Jack N. P. Davies described EMF in Uganda so it is also known as Davies disease.³ It is mainly a tropical disease. EMF is usually seen in areas that are located within 15 degrees of the equatorial belt. It is the commonest cause of

restrictive cardiomyopathy worldwide. Endomyocardial fibrosis is a disease of uncertain origin. Several factors have been implicated in its origin. Poverty, malnutrition, infections (viral and parasitic), autoimmunity, allergy (eosinophilia), toxic agents (cerium, cassava, serotonin) and heredity have all been implicated¹. It can mimic other common conditions like apical hypertrophic cardiomyopathy, Constrictive pericarditis, rheumatic valve disease or Ebstein’s anomaly.⁴

Table 1: Criteria for diagnosis of EMF⁶	
Major criteria	
☐	Endomyocardial plaques >2 mm in thickness
•	Thin (≤1 mm) endomyocardial patches affecting more than one ventricular wall
☐	Obliteration of the right ventricular or left ventricular apex
☐	Thrombi or spontaneous contrast without severe ventricular dysfunction
☐	Retraction of the right ventricular apex
☐	Atrioventricular-valve dysfunction due to adhesion of the valvular apparatus to the ventricular wall
Minor criteria	
•	Thin endomyocardial patches localized to one ventricular wall
•	Restrictive flow pattern across mitral or tricuspid valves
•	Pulmonary valve diastolic opening
•	Diffuse thickening of the anterior mitral leaflet
•	Enlarged atrium with normal size ventricle
•	M movement of the interventricular septum and flat posterior wall
•	Enhanced density of the moderator or other intraventricular bands
*A definite diagnosis of EMF is made in the presence of two major criteria or one major criteria associated with two minor criteria.	

The important feature of this disease is the formation of fibrous tissue in the endocardium and to a lesser extent in the myocardium of one or both ventricles. Obliteration of ventricular cavities by fibrous tissue and thrombus contributes to increased resistance to ventricular filling leading to diastolic dysfunction.⁵

In a study done in Mozambique the most common form was biventricular endomyocardial fibrosis (55.5%), followed by



right-sided endomyocardial fibrosis (28%).⁶ In the same study left-sided disease was seen in 16.6% of cases. Our patient had right-sided EMF and she had clinical features of right heart failure. In a case series of 30 cases of EMF from India only two patients had isolated right ventricular EMF.⁷ Santra et al.⁵ have reported a case of right ventricular EMF from India.

Echocardiography is the most valuable tool for diagnosis of EMF.⁸ Mocumbi et al.⁶ have described echocardiographic criteria to diagnose EMF. Late gadolinium enhancement (LGE) cardiovascular magnetic resonance (CMR) can also help in the diagnosis and prognosis of EMF.⁹ CMR was not done in our case due to financial constraints. Endomyocardial biopsy is not essential to diagnose EMF.⁴

Medical management usually includes treatment with diuretics and anticoagulants. Surgical treatment consists of resection of fibrotic tissue and valve repair or replacement.¹⁰

Surgical treatment of EMF should be considered a palliative procedure because surgery does not alter the progressive nature of the disease.¹¹ Overall prognosis of EMF is very poor. Change in socioeconomic and health status over past four decades is associated with decline in new cases of EMF in our country.¹²

EMF is a neglected tropical disease. More research needs to be done to find the exact pathogenesis of EMF.

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests

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PATIENT CONSENT

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1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient.
3. This submission is compliant with the requirements of local research ethics committees.