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A case of typical right ventricular endomyocardial fibrosis

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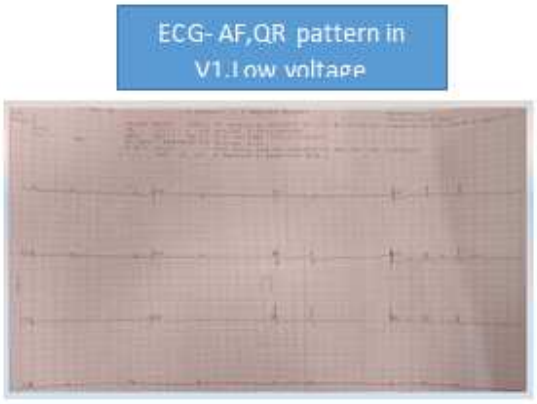
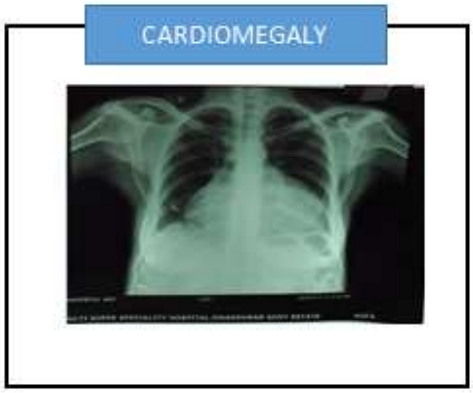
ABSTRACT

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 (1) Here we report a rare case of right ventricular endomyocardial fibrosis .A 48 -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





CASE HISTORY

48 yrs old female admitted with complaints of abdominal distension and difficulty in breathing for the past 6 month duration. History of left lower limb varicose vein on regular scelerotheraphy. No recent history of syncope, chest pain, palpitation. on examination vitals seems stable with elevated JVP and features of right heart failure present. Not a known diabetic or hypertensive. ECG showed atrial fibrillation with controlled ventricular rate and low voltage complex, with T wave inversion in V1 to v3 . Echocardiogram showed huge dilated right atrium with small right ventricle, obliteration of right ventricular apex, RV dysfunction, dilated RVOT of more than 36mm with RVseptum, free wall and RVOT showed dyskinesia. low pressure TR present. Cardiac MRI showed right atrial aneurysm with diverticulum like out pouching, dilated IVC with no delayed myocardial hyper enhancement. with this scenario RV cardiomyopathy was suspected, By applying the echo criteria (NEJM), EMF was diagnosed ,features include RV apex obliteration (HEART OF AFRICA), MERLON SIGN positive (hypercontracting basal segments and obliteration of RV apex),RV dysfunction, RVOT dilatation with anterior mital leatlet thickening and

restrictive flow pattern present. This case contains all the typical findings for the right ventricular endomyocardial fibrosis ,hence it was reported.

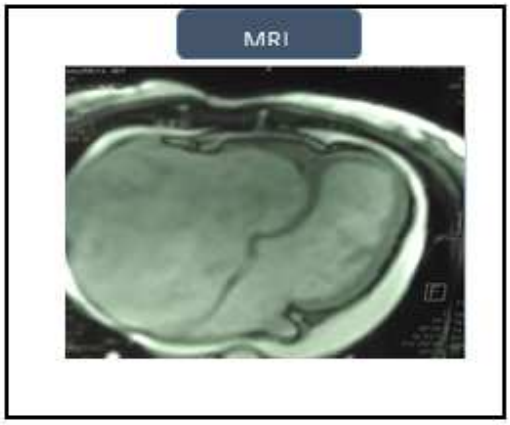
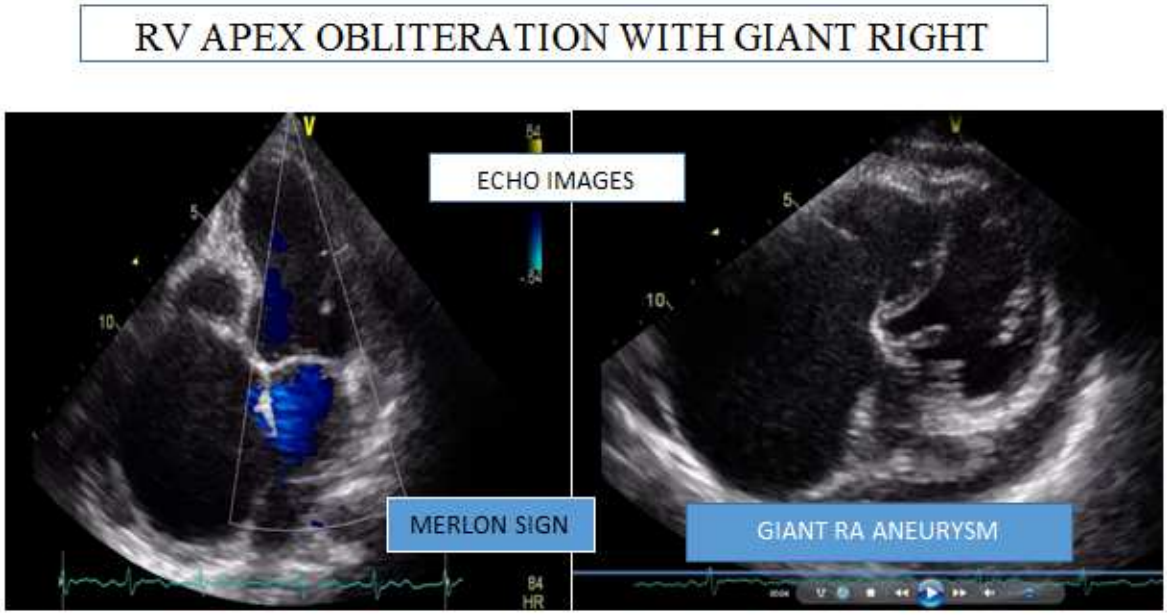


Table 1. Criteria for Diagnosis and Assessment of the Severity of Endomyocardial Fibrosis.*

Criterion	Score
Major criteria	
Endomyocardial plaques >2 mm in thickness	2
Thin (≤1 mm) endomyocardial patches affecting more than one ventricular wall	3
Obliteration of the right ventricular or left ventricular apex	4
Thrombi or spontaneous contrast without severe ventricular dysfunction	4
Retraction of the right ventricular apex (right ventricular apical notch)	4
Atrioventricular-valve dysfunction due to adhesion of the valvular apparatus to the ventricular wall	1–4†
Minor criteria	
Thin endomyocardial patches localized to one ventricular wall	1
Restrictive flow pattern across mitral or tricuspid valves	2
Pulmonary-valve diastolic opening	2
Diffuse thickening of the anterior mitral leaflet	1
Enlarged atrium with normal-size ventricle	2
M-movement of the interventricular septum and flat posterior wall‡	1
Enhanced density of the moderator or other intraventricular bands	1

* A definite diagnosis of endomyocardial fibrosis was made in the presence of two major criteria or one major criterion associated with two minor criteria. A total score of less than 8 indicates mild endomyocardial fibrosis, 8 to 15 moderate disease, and more than 15 severe disease.



MOZABIQUE ECHO CRITERIA FOR EMF nejm med 359;1 www.nejm.org july 3, 2008,43-9.

DISCUSSION

Jack N. P. Davies described EMF in Uganda so it is also known as Davies disease. It is mainly a tropical disease. EMF is usually seen in areas that are located within 15 degrees of the equatorial belt (3) .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 (2) leading to diastolic dysfunction.

In a study done in Mozambique the most common form was biventricular endomyocardial fibrosis (55.5%), followed by right-sided endomyocardial fibrosis (28%). In the same study left-sided disease was seen in 16.6% of cases(6). 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(2).Santra et al. have reported a case of right ventricular EMF from India (5). Echocardiography is the most valuable tool for diagnosis of EMF. Mocumbi et al. 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 (6)CMR was done in our case. Endomyocardial biopsy is not essential to diagnose EMF.Medical management usually includes treatment with diuretics and anticoagulants. Surgical treatment consists of resection of fibrotic tissue and valve repair or replacement. Surgical treatment of EMF should be considered a palliative procedure – **Bidirectional Glenn shunt** can also done as a palliative procedure , because surgery does not alter the progressive nature of the disease (3)

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. EMF is a neglected tropical disease (4)More research needs to be done to find the exact pathogenesis of EMF.

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