

Case study**ENDOMYOCARDIAL FIBROSIS: AN UNUSUAL PRESENTATION****ABSTRACT**

Endomyocardial Fibrosis is a restrictive cardiomyopathy characterized by fibrosis of the ventricular endocardium. It is usually associated with poor prognosis due to late presentation and inadequate surgical responsiveness. Some authors have described it as a neglected disease despite the growing body of knowledge on this condition. This case highlights the occurrence of EMF in an elderly woman without the typical/characteristic physical findings of EMF.

KEY WORDS; Endomyocardial fibrosis, Cardiomyopathy, Heart failure.

INTRODUCTION

Endomyocardial fibrosis(EMF), previously termed tropical endocardial disease or endocardial fibroelastosis¹ was first described by Bedford and Konstan in 1946 among soldiers of West African region who served in the British Army during the second world war² and later in Kampala, Uganda in 1948 by Jack Davies²

It is characterized by fibrosis of the right ventricular and left ventricular apical endocardium resulting in a restrictive cardiomyopathy. There is also destruction of the subvalvular apparatus, though the fibrosis does not involve the valve leaflets. When the fibrotic process involves the papillary muscles and the subvalvular mechanism, valve immobility results in atrioventricular valve incompetence.

We present a case of atypical presentation of Endomyocardial Fibrosis at the Jos University Teaching Hospital.

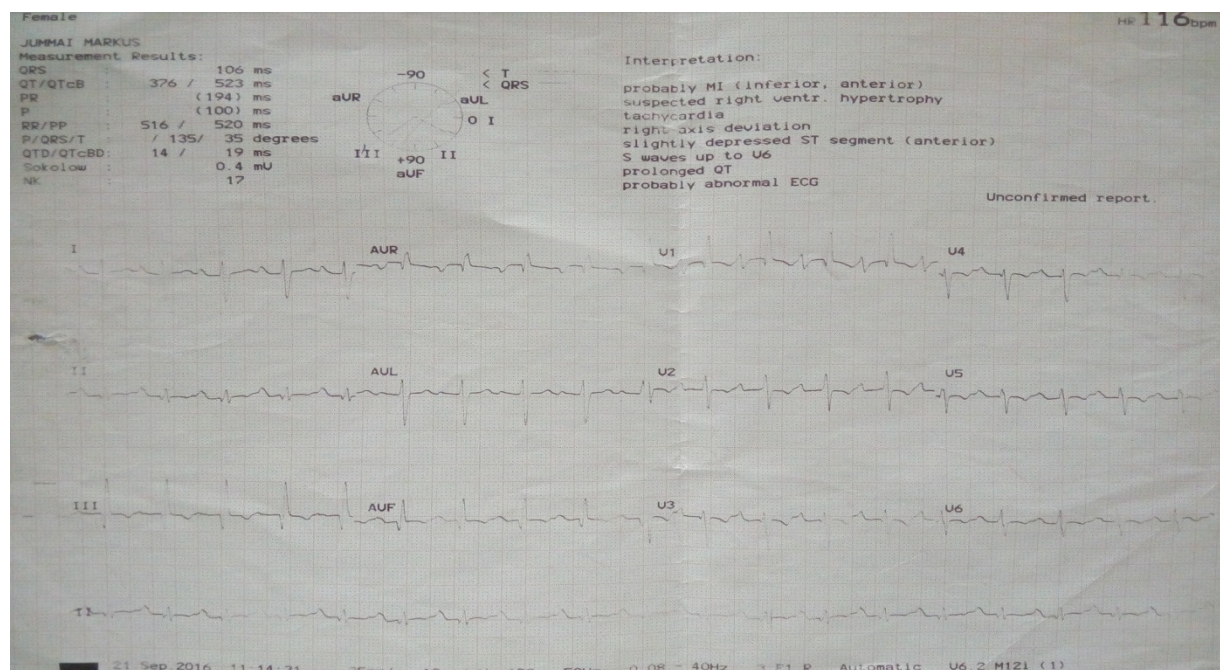
25 **CASE REPORT**

26 Mrs SJM is a 65 year old house wife and retired food vendor who now
27 occasionally farms. She presented to Jos University Teaching Hospital on account
28 of recurrent breathlessness, cough and generalized body swelling of 1 year
29 duration. She was previously known to be hypertensive but has not been
30 compliant on her medications and clinic visits. She fried and sold acara (beans
31 cake) with biomass fuel for over 30 years. There is no known family history of
32 similar illness.

33 Physical examination was remarkable for a well built and obese looking woman
34 with central cyanosis, respiratory distress, tachypnoea, bilateral pitting pedal with
35 sacral and anterior abdominal wall oedema. Pulse rate was 100bpm and Blood
36 Pressure 100/60mmHg Supine.

37 She had engorged neck veins and a raised jugular venous pressure up to the angle
38 of the jaw. Apex was difficult to palpate due to a thick chest wall. She had a
39 normal S1, Loud P2 and a parasternal S3. With bibasal fine crepitations. SPO2 was
40 53% on room air. Her liver was tipped and tender. An initial assessment of
41 Corpulmonale 2⁰ COPD was made.

- 42 ECG done (Figure 1) showed sinus tachycardia, right axis deviation, right atrial
43 enlargement, right ventricular hypertrophy and SI ,QIII and TIII pattern.



- 44
- 45 Figure 1.

- 46 Chest X-ray (Figure 2) showed cardiomegaly[CTR 0.7], upper lobe diversion,
47 widespread interstitial shadows and blunted right recess.



48

49 Figure 2.

50 Pack Cell Volume was 56%. Erythrocyte Sedimentation Rate (ESR) was 20mm/hr.

51 Total White Blood Cell Count was 4,200/ml with Eosinophils constituting 4% of it.

52 Echocardiography done [Figure 3] revealed a markedly dilated right atrium[Major

53 axis 71cm and minor axis 65cm] and right ventricle with fibrotic plaques on the

54 right ventricular apex. She had a restrictive pattern of relaxation [EA ratio 2.9,

55 Deceleration time 107ms], a dilated pulmonary trunk measuring 26mm,

56 pulmonary regurgitation and evidence of severe pulmonary hypertension.

57 Tricuspid annular plane systolic excursion [TAPSE] was 11. And there was mild

58 pericardial effusion [4mm]. Tricuspid valve area by planimetry was 7.03cm².



60 Figure 3.

A substantive diagnosis of Endomyocardial Fibrosis with pulmonary embolism in Failure was made. She was placed on diuretics, anticoagulants, sildenafil citrate and intermittent intranasal oxygen. And was discharged home after a month on admission to continue follow-up at the cardiology clinic.

DICUSSION

EMF, though worldwide in occurrence, it is mainly seen in the tropical and subtropical regions; relatively uncommon in Northern and southern Africa³. It has been reported in regions within the tropical belt 15⁰ of the equator.

Most of studies on EMF describe clinical series of patients in advanced stages the disease with large-scale epidemiologic studies lacking⁴ . A community survey in rural Mozambique, involving 1249 patients revealed a prevalence of 19.8% (211 cases) out of which 33 had surgery⁵. In Uganda, EMF accounted for 20% of patients referred for echocardiography⁶

The aetiopathogenesis of EMF remains much of an enigma, with only probable causes postulated; ranging from filariasis-induced occlusion of cardiac lymphatics, schistosomiasis, loa-loa infection, Toxoplasma gondii infection, hypersensitivity reaction to streptococcal infection^{7,8,9,10,11,12}. Excessive consumption of plantain which is rich in serotonin, environmental exposure to Cerium(a rare element present in affected areas) and Magnesium and Protein deficiency have also be thought to be likely causes¹.

The debate on it being a part of Loeffler's hypereosinophylic syndrome still rages decades later.

EMF has a bimodal peak age of presentation with onset in the first decade (10 years) and a second peak occurring in the second to fourth decades of life¹⁴.

Earlier reports gave female sex preponderance with ratio of 2:1². Mocumbi et al⁷, in Mozambique reported more males affected than females (23% versus 17%). The Jos series by Isiguzo et al¹⁵ also showed a male preponderance with average age of presentation being 28 years.

The above case report did not typify the presentation of EMF. First, it occurred in the northern part of Nigeria, where until a couple of years ago, was not thought to occur. Danbauchi et al reported cases of EMF in Zaria, an ancient city in the guinea savannah of North West Nigeria¹⁶. The low prevalence in the region by earlier reports could have been hinged on the non-availability of echocardiographic machines and low index of clinical suspicion. Interestingly, in 2002, the prevalence of EMF showed a decrease in a hospital audit in Enugu, southern Nigeria from a country prevalence of 10% to 0.8%. And the authors questioned if this was a true reflection or from a missed diagnosis¹⁷. Akinwusi et al reported a decline in prevalence of EMF in South West Nigeria [from 10% in the 1960s and 1970s to 0.02% to 0.04%]¹⁸

Secondly, it occurred in a 65year old woman with history of daily biomass exposure for 30 years ; a scenario that depicts risk for COPD and probably corpulmonale. EMF is usually seen before the fifth decade of life¹⁴.

Clinical features of endomyocardial fibrosis depend on the stage of the disease and the anatomical involvement of the heart. About 50% of children and

107 adolescents report an initial illness with fever, chills and night sweat with facial
108 swelling and urticaria¹⁸. This was not seen in the patient presented.

109 The eosinophil hypothesis of EMF gained prominence in the 1960s with reports
110 of eosinophilic endomyocardial disease among European visitors to tropical
111 regions¹⁹. In a study of patients with EMF, Andy and colleagues found eosinophilia
112 in 80% of their cohorts. The authors believed that eosinophilia is seen in the early
113 stages of the illness. This patient did not have eosinophilia probably because she
114 presented late. Yet, the “egg on stick appearance”, which is a characteristic
115 clinical feature of EMF was also not present in her.

116 Patients with severe RVEMF can have cyanosis due to right to left atrial shunting
117 through a patent and stretched foramen ovalis defect¹⁵. In this case, the cyanosis
118 was attributed to the concurrent pulmonary embolism the patient had. Atrial
119 thrombus, commonly seen in the right atrium is due to stagnating blood in the
120 large right atrium, especially in the presence of AF.

121 The clinical presentation varies based on the extent and stage of the disease.
122 Patients with isolated RVEMF present with purely right sided heart failure with
123 oedema, ascites, cachexia and malnutrition, elevated JVP and hepatomegally.
124 These features, with the exception of cachexia and malnutrition were evident in
125 the patient. The patient had severe TR and PR with markedly elevated pulmonary
126 pressure. EMF does not usually involve the outflow tracts but in this patient, the
127 pulmonary incompetence can be attributed to the pulmonary hypertension and
128 dilated pulmonary trunk. Features of pulmonary arterial hypertension usually
129 depend on the presence of pulmonary venous hypertension from mitral
130 incompetence as well as diastolic dysfunction. This patient did not have mitral

incompetence. It's been said that few patients may present with pulmonary hypertension entirely due to LV diastolic compliance failure in the absence of mitral incompetence, but this is distinctly rare²⁰. The patient had severe pulmonary hypertension with impaired relaxation.

EMF is often times detected as an incidental finding in patients with rheumatic or other heart diseases²⁰. The above case was initially managed as corpulmonale.

Medical treatment options are decongestive therapy with diuretics, anticoagulation for pulmonary embolism, antiarrhythmic treatment for AF and abdominal paracentesis to relieve tense ascites.

Surgical options for treatment include LV endocardectomy, AV valve repair or replacement and exclusion of fibrotic RV in pure RVEMF by a bidirectional Glenn (BDG) connection²¹. A plane of cleavage can easily be developed and all the thickened myocardium removed. Successful surgical resection of the endocardial fibrosis with valve repair or replacement can have dramatic effect on symptoms and survival, although the operation itself is associated with a significant risk for morbidity and mortality¹. In the Mozambique community survey, the most prominent complication in patients who underwent surgery was pericardial effusion requiring surgery²².

CONCLUSION

Although Endomyocardial fibrosis occurs primarily in the subtropical regions of Africa in its typical manifestation, in this report, it was in the Savannah belt. And the case did not occur in the typical age group (adolescents and young adults). It is therefore important to realize that EMF has indeed remained an enigma. And

154 despite advances in medical knowledge, majority of health facilities in Africa lack
155 adequate capacity to effectively treat this neglected disease.

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