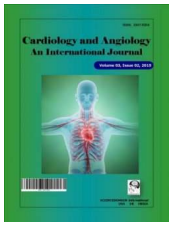

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Endomyocardial Fibrosis: An Unusual Presentation

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Authors' contributions

This work was carried out in collaboration between all authors. Author CMC designed the study and wrote the first draft of the manuscript. Authors SCR, VSG and YCK managed the literature searches of the study. Author SSD managed the analyses of the study. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Endomyocardial fibrosis is a tropical disease, characterised by fibrosis in the endocardium and subjacent myocardium affecting particularly the inflow tract and the apex of one or both ventricles. This case highlights the occurrence of EMF in an elderly woman without the typical/characteristic physical findings of EMF.

Keywords: Endomyocardial fibrosis; heart failure.

1. INTRODUCTION

Endomyocardial fibrosis (EMF), previously termed tropical endocardial disease or endocardial fibroelastosis [1] 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 war [2] and later in Kampala, Uganda in 1948 by Jack Davies [2].

It is usually associated with poor prognosis due to late presentation and inadequate surgical responsiveness. Early diagnosis is almost impossible because of absence of fibrosis [3].

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Some authors have described it as a neglected disease despite the growing body of knowledge on this condition. We present a case of an atypical presentation of Endomyocardial Fibrosis at the Jos University Teaching Hospital. Jos Nigeria.

2. CASE REPORT

Mrs SJM is a 65 year old retired food vendor now subsistent farmer born and bred in a village near Jos, Central Nigeria. She presented to the Jos University Teaching Hospital on account of recurrent breathlessness, cough and generalized body swelling of about a year's duration. She was previously known to be hypertensive, but neither regular on her medications nor any regular clinic visits. She had been exposed to biomass fuel for over 30 years as a food vendor. There was no known family history of cardiac disorder.

On physical examination, she was obese, with marked central cyanosis, in respiratory distress with tachypnoea. She had bilateral pitting pedal, sacral and anterior abdominal wall oedema.

On cardiovascular examination, pulse rate was 100 bear per minute and Blood Pressure was 100/60 mmHg in the Supine position. She had engorged neck veins and a raised jugular venous pressure up to the angle of the jaw. Apex was difficult to palpate due to a thick chest wall. She had a normal first heart sound, Loud P2 component of second heart sound and a left parasternal third heart sound. Chest auscultation revealed fine crepitations at the bases. Oxygen saturation (SPO2) was 53% on room air. Abdominal examination revealed a tender tipped liver. An initial assessment of Cor-pulmonale 2° COPD in failure was made.

ECG done (Fig. 1) showed sinus tachycardia, right axis deviation, right atrial enlargement, right ventricular hypertrophy and SIQIIITIII pattern.

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

Echocardiography done (Fig. 3) revealed a markedly dilated right atrium [Major axis 71 cm and minor axis 65 cm] and right ventricle with fibrotic plaques in the right ventricular apex. The left ventricle and atrium were smaller the right ventricle and atrium. She had a restrictive pattern of relaxation [EA ratio 2.9, Deceleration time 107 ms] in Doppler study. A dilated pulmonary trunk measuring 26 mm, pulmonary regurgitation and evidence of severe pulmonary hypertension was seen. Tricuspid annular plane systolic excursion [TAPSE] was 11. And there was mild pericardial effusion [4 mm] posteriorly. Tricuspid valve area by planimetry was 7.03 cm2.

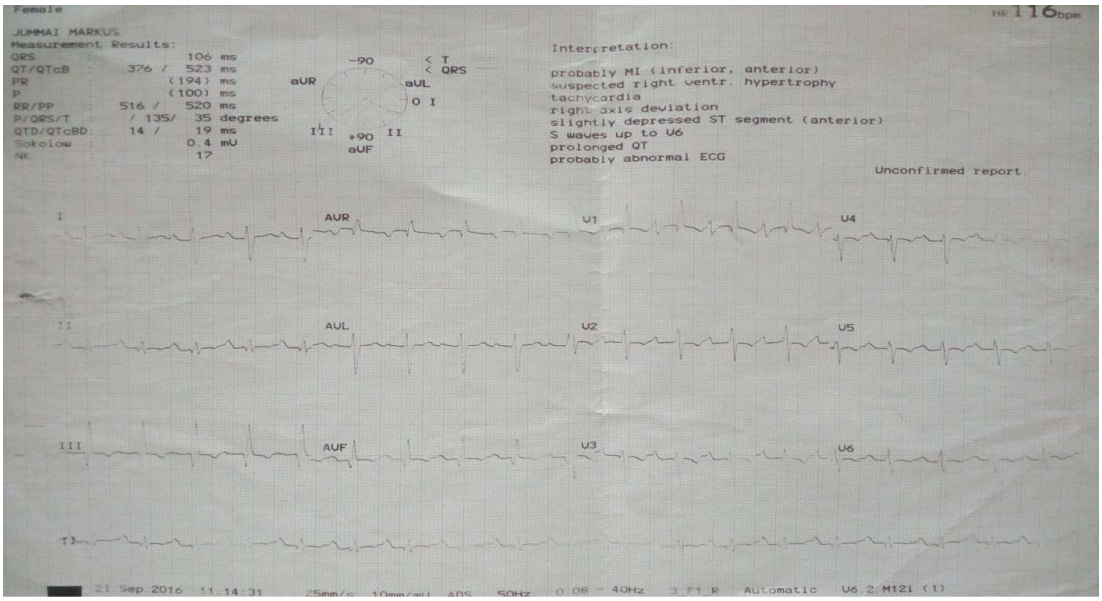

Fig. 1. Electrocardiogram of patient

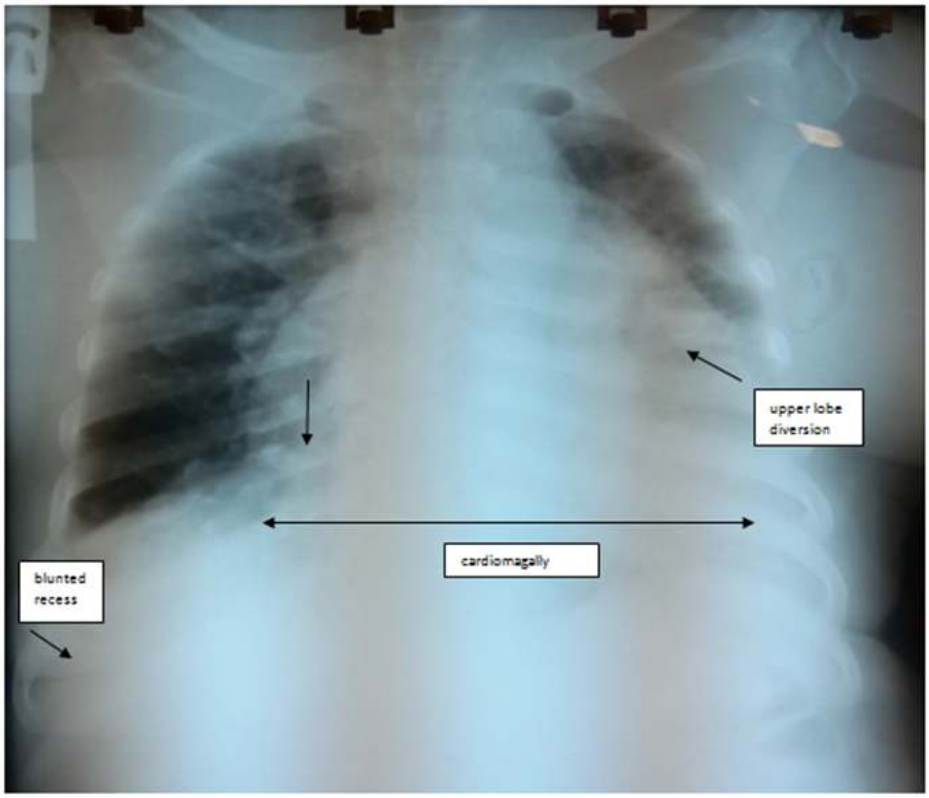

Fig. 2. Chest radiogram of patient

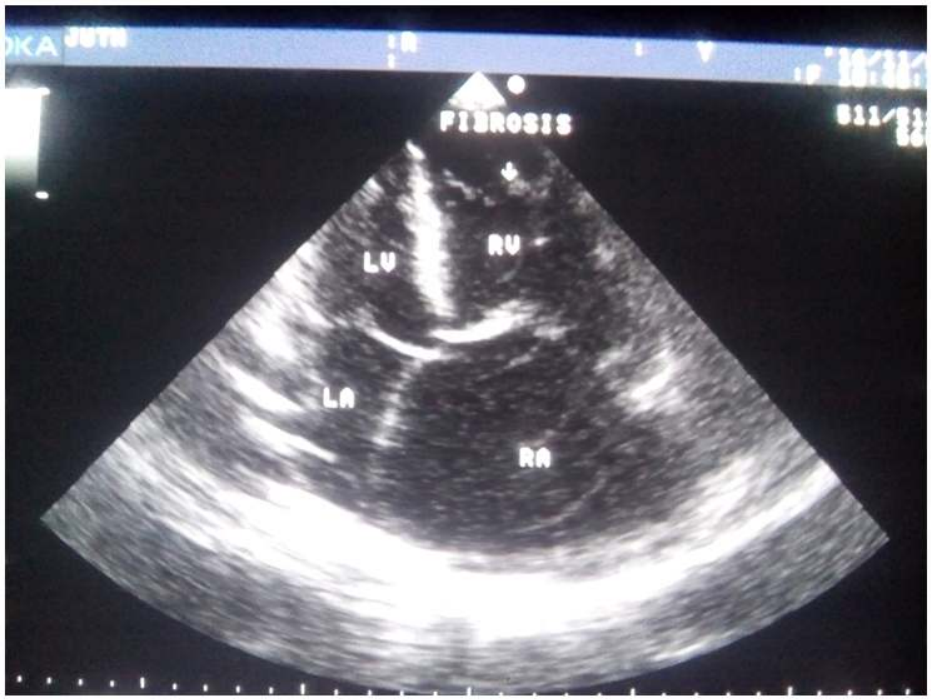

Fig. 3. Echocardiogram of patient

Other investigations done included, Pack Cell Volume was 56%, with mild anisocytosis. Erythrocyte Sedimentation Rate (ESR) was 20mm/hr. Total White Blood Cell Count was 4,200/ml with Eosinophils constituting 4% of it. Pulmonary function test revealed an obstructive pattern with a peak expiratory flow rate of less than 20% for her age. Sputum cultures were negative for common organisms. Liver function test showed raised total and conjugated bilirubin, alkaline phosphatase and transaminases.(total bilirubin 56micromol/l, conjugated bilirubin 46micromol/l, alkaline phosphatase 105IU/l). Cardiac catheterization, magnetic resonance imaging (MRI) were not available in this centre.

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. There was no significant improvement in her condition to enable her fit for pericardial stripping and was discharged home after a month on admission to continue follow-up at the cardiology clinic.

3. DISCUSSION

EMF, though worldwide in occurrence, it is mainly seen in the tropical and subtropical regions; relatively uncommon in Northern and southern Africa [3]. Diagnosis of EMF was challenging, made only when an echocardiography was done. Markedly dilated right atrium and right ventricular apical fibrosis were the hall-mark of EMF diagnosis. Most of studies on EMF describe clinical series of patients in advanced stages the disease with large-scale epidemiologic studies lacking [4]. A community survey in rural Mozambique, involving 1249 patients revealed a prevalence of 19.8% (211 cases) out of which 33 had surgery [5]. In Uganda, EMF accounted for 20% of patients referred for echocardiography [6]. These reports were based on echocardiography findings. The patient features were more in keeping with cor-pulmonalea, but the finding of apical fibrosis is in favor of EMF. 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 life [7].

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, it was considered non-existent in the savannah region. Danbauchi et al. reported cases of EMF in Zaria, an ancient city in the guinea savannah of North West Nigeria [8]. 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 in early stages. 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 diagnosis [9]. 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%] [10].

Secondly, it occurred in a 65 year old woman with history of daily biomass exposure for about 30 years, a scenario that depicts risk for COPD and probably cor-pulmonale.

Clinical features of endomyocardial fibrosis depend on the stage of the disease and the anatomical involvement of the heart. About 50% of children and adolescents report an initial illness with fever, chills and night sweat with facial swelling and urticaria [10]. This was not seen in the patient presented.

The eosionphil hypothesis of EMF gained prominence in the 1960s with reports of eosinophilic endomyocardial disease among European visitors to tropical regions [11]. In a study of patients with EMF, Andy and colleagues found eosinophilia in 80% of their cohorts [11]. The authors believed that eosinophilia is seen in the early stages of the illness. This patient did not have eosinophilia probably because she presented late. Yet, the ‘’egg on stick appearance’’, which is a characteristic clinical feature of EMF was also not present in her.

Patients with severe RVEMF can have cyanosis due to right to left atrial shunting through a patent and stretched foramen ovalis defect [12]. In this case, the cyanosis was attributed to the concurrent pulmonary embolism the patient had. Atrial thrombus, commonly seen in the right atrium is due to stagnating blood in the large right atrium, especially in the presence of AF.

The clinical presentation varies based on the extent and stage of the disease. Patients with isolated RVEMF present with purely right sided heart failure with oedema, ascites, cachexia and malnutrition, elevated JVP and hepatomegally. These features, with the exception of cachexia and malnutrition were evident in the patient. The

patient had severe tricuspid regurgitation and pulmonary regurgitation with markedly elevated pulmonary pressure. Features of pulmonary arterial hypertension usually depend on the presence of pulmonary venous hypertension from mitral 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 rare [13]. The patient had severe pulmonary hypertension with impaired relaxation pattern.

EMF is often times indolent, detected as an incidental finding in patients with rheumatic or other heart diseases [14]. The above case can be considered as an example, where initial management was as case of cor-pulmonale only to diagnose EMF following echocardiography.

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 endocardiectomy, AV valve repair or replacement and exclusion of fibrotic RV in pure RVEMF by a bi-directional Glenn (BDG) connection [15]. 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 mortality [1]. In the Mozambique community survey, the most prominent complication in patients who underwent surgery was pericardial effusion requiring surgery [16,17].

4. CONCLUSION

Endomyocardial fibrosis occurs primarily in the subtropical regions of Africa in adolescents and young adults classically. More cases are now reported in the Savannah belt in our case in the elderly. It is therefore important to realize that EMF has indeed remained an enigma. And despite advances in medical knowledge, majority of health facilities in Africa lack adequate capacity to make early diagnosis and effectively treat this neglected disease.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

All authors hereby declare that all documentations have been examined and approved by the appropriate ethics committee of the Jos University Teaching Hospital and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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