Eosinophilia to endomyocardial fibrosis: Documentation of a case

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ABSTRACT

Endomyocardial fibrosis (EMF) is an important cause of restrictive cardiomyopathy in tropical countries. The etiopathogenesis of EMF remains obscure. The role of eosinophilia in the etiopathogenesis of EMF has been debated extensively, but remains unproven. Accordingly, we present a case wherein a patient with documented eosinophilia and heart failure at the age of three-and-a-half years presented with endomyocardial fibrosis at the age of nine years. Such documentation is important to highlight the central role of eosinophils in the pathogenesis of EMF.

Keywords: Endomyocardial fibrosis, eosinophilia, eosinophilic myocarditis, Loeffler's myocarditis, restrictive cardiomyopathy

INTRODUCTION

Endomyocardial fibrosis (EMF) is an important cause of restrictive cardiomyopathy in the developing countries.[1] The etiopathogenesis of EMF remains unclear. The role of eosinophilia in the etiopathogenesis of EMF has been debated extensively but remains unproven. Accordingly, we present a case wherein a patient with documented eosinophilia and heart failure developed EMF several years later.

CASE REPORT

A 9-year-old girl presented with the complaints of progressive dyspnea and abdominal distension for 8 months. The patient had worsening orthopnea and nocturnal cough for 2 weeks before presentation and also reported episodic palpitations.

Physical examination revealed a cachectic and icteric young girl [Figure 1]. Jugular venous pressure was elevated to 8 cm above the clavicle with prominent V waves. On cardiac examination, she was in atrial fibrillation with a heart rate of 130 beats/min. The apical impulse was laterally displaced. On auscultation, S1 was soft, S2 normally split with accentuated pulmonary component, right ventricular (RV) S3 was present, and systolic murmurs of mitral and tricuspid regurgitation (TR) were present at the apex and left lower sternal border, respectively. There was tense ascites and pulsatile hepatomegaly of 4 cm below costal margin. Examination of the other systems was unremarkable.

The electrocardiogram showed atrial fibrillation, right axis deviation, and RV hypertrophy [Figure 2]. Chest radiograph showed massive cardiomegaly with prominent right atrial (RA) enlargement and no evidence of pulmonary venous hypertension or pleural effusion [Figure 3]. Echocardiography revealed obliterated RV cavity with thickened tricuspid valve, severe low-pressure TR, and a large RA thrombus. Left ventricular (LV) cavity was partially obliterated with normal LV systolic function and moderate-to-severe mitral regurgitation (MR) [Figure 4]. A limited cardiac catheterization study done after resolution of thrombus showed elevated RA pressures (mean pressure – 31 mm Hg), mildly elevated pulmonary artery pressures (systolic/diastolic/mean pressures – 38/22/29 mmHg)

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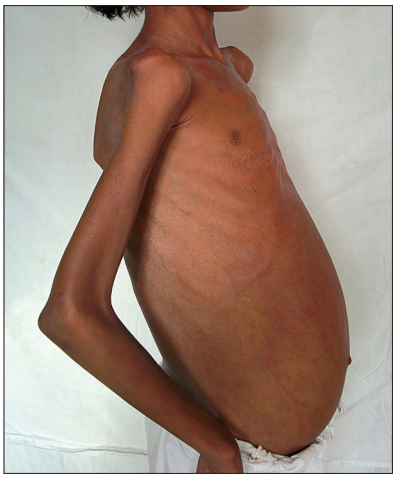

Figure 1: Tense ascites; peripheral edema was conspicuously absent

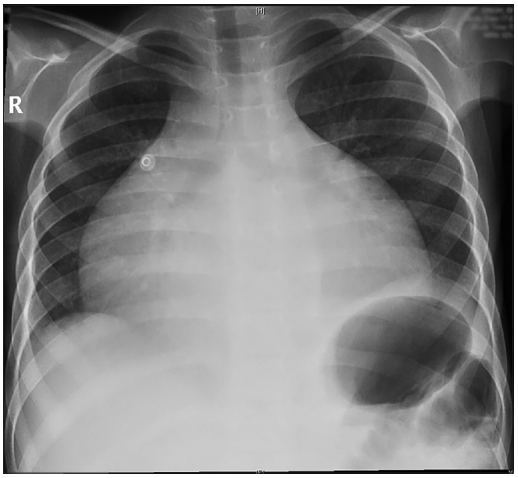

Figure 3: Chest radiograph showing massive cardiomegaly with predominant right atrial enlargement. Pulmonary venous hypertension is absent and there is no pleural effusion

with elevated LV end-diastolic pressure – 25 mm Hg. The cardiac index and pulmonary vascular resistance index were in the normal range (2.9 L/min/m^2 and 1.8 Woods units/m^2, respectively). Cardiac magnetic resonance imaging showed obliteration of RV apex with diffuse subendocardial late gadolinium enhancement in the LV and RV walls. These findings were confirmative of biventricular EMF [Figures 5 and 6].

Laboratory investigations revealed conjugated hyperbilirubinemia (total bilirubin – 5.5 mg/dL, conjugated bilirubin – 2.25 mg/dL) and primary hypothyroidism (T3 – 39.3 ug/dL, T4 – 3.8 ug/dL, and TSH – 17.4 uIU/mL). Other liver function tests, renal function tests, and complete blood count were normal. The peripheral smear, eosinophil count, and stool routine examination were normal. The patient was diagnosed to have EMF with the involvement of both ventricles but RV was more involved than the LV.

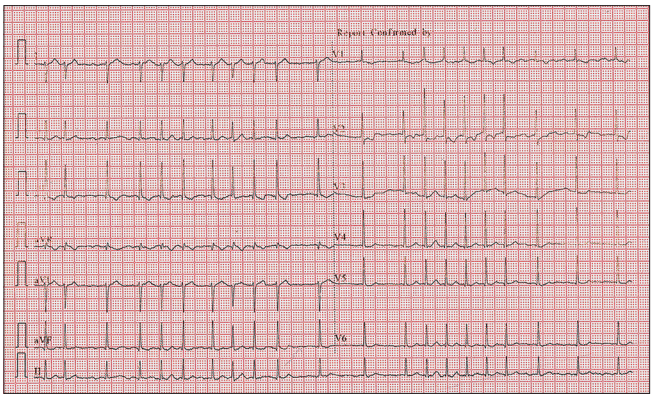

Figure 2: Electrocardiogram showing atrial fibrillation with fast ventricular rate, right axis deviation, right ventricular hypertrophy

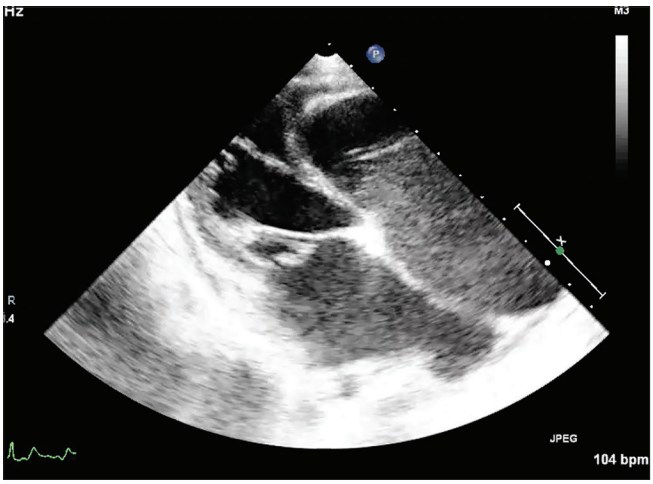

Figure 4: Four-chamber echocardiographic view showing nearly complete obliteration of right ventricular cavity with involvement of left ventricle as well

Evaluation of prior history provided insights into the patient's disease. At 3½ years of age, the patient had an acute illness with low-grade fever for 4 weeks, followed by anasarca, abdominal distension, and oliguria. The patient was admitted to another hospital for these complaints. The documented absolute eosinophil count was 9000 cells/uL, and immunoglobulin E levels were highly elevated (2662 IU/L, normal: 45–378 IU/L). The geographical location of the patient's village is in the endemic filariasis zone, and according to her parents, several residents of the village were known to have unilateral lower limb swelling typical of filariasis. The patient, however, did not receive treatment for filariasis, oral, or intravenous steroids at any point in time. Cardiac imaging during this episode of illness was not done. She improved with diuretics and was treated with oral diuretics for 1 year. The patient was then asymptomatic till her current presentation.

The final diagnosis was restrictive cardiomyopathy due to EMF, secondary to eosinophilia in the past, with biventricular involvement (RV more than LV), severe TR,

moderate MR, and normal LV function. The patient was taken up for endocardectomy and valve repairs for EMF, however, she expired in the postoperative period. The histopathology of surgically excised tissue was consistent with EMF [Figures 7 and 8].

DISCUSSION

EMF is an important cause of restrictive cardiomyopathy and heart failure in low-income tropical countries with a characteristic geographical distribution (specific geographic hot spots include Sub-Saharan Africa, coastal India, and South America).[1,2] The disease has an acute phase, an intermediate phase (when the patient is relatively asymptomatic), and a chronic phase.[3] The usual presentation is late in the chronic phase with irreversible fibrosis. The precise etiopathogenesis of EMF is still obscure although several etiological factors have been suspected including infections, eosinophilia, environmental, and dietary factors such as cassava and plantain-based diets, cerium toxicity, protein-energy malnutrition, and magnesium deficiency.[1]

Eosinophilic myocarditis in Loeffler's disease histopathologically resembles EMF; however, the lack of eosinophilia in the chronic phase of EMF and some other factors has been cited as evidence against the role of eosinophilia in the etiopathogenesis of EMF.[4] Few reports, like the present case, have documented the evolution of disease in patients with eosinophilia in the past have not been adequately emphasized in the literature.[5,6]

Eosinophilia in the population is quite common, but EMF is a rare disease; other factors that influence the occurrence of EMF in patients with eosinophilia remain to be studied. However, cases such as the present one should be documented so that research on the etiopathogenesis of EMF can be directed appropriately.

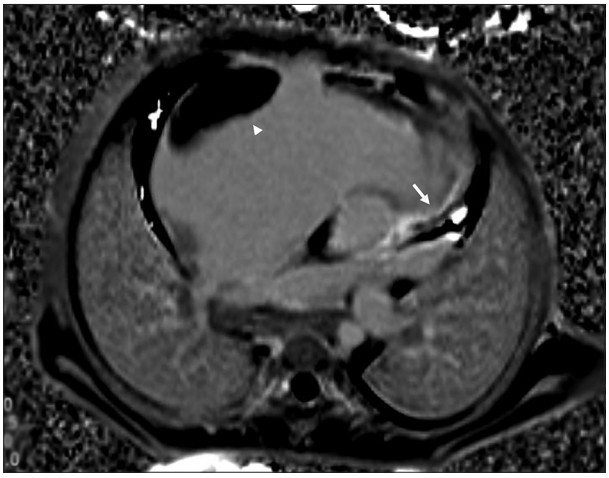

Figure 5: Cardiac magnetic resonance image showing obliteration of right ventricular apex with subendocardial late gadolinium enhancement in the right ventricular wall (arrow). Grossly dilated right atrium with right atrial thrombus (arrowhead) are also noted

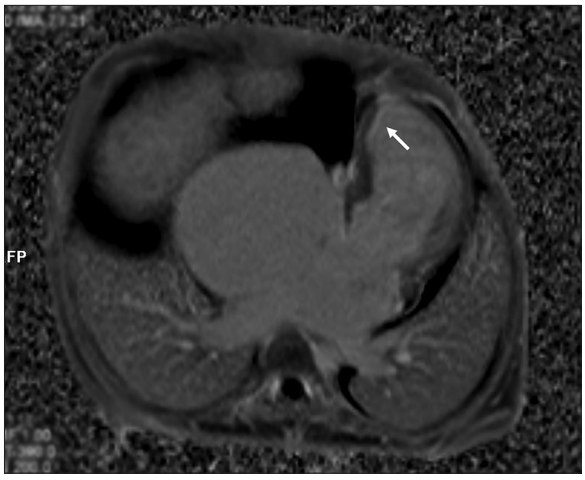

Figure 6: Cardiac magnetic resonance image showing subendocardial late gadolinium enhancement in the left ventricular wall

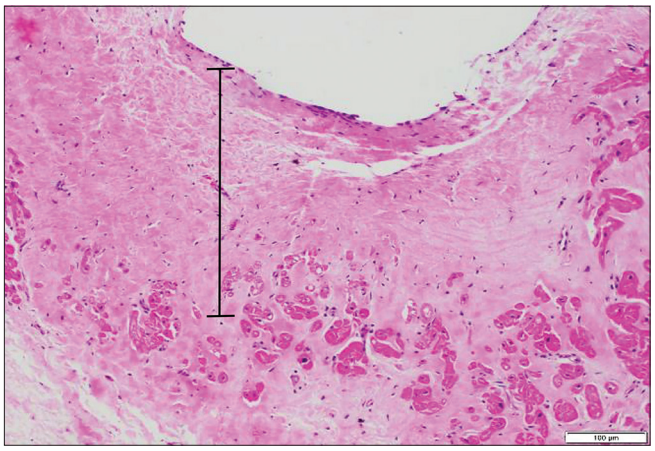

Figure 7: Markedly thickened endocardium with extension of fibrosis into the underlying myocardium (straight line, ×10)

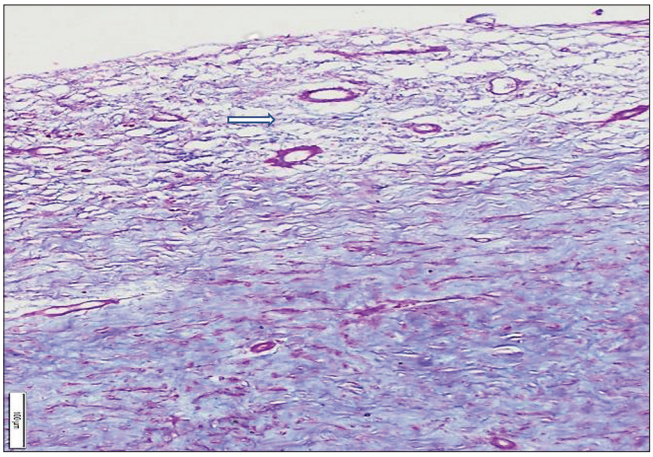

Figure 8: Masson trichrome stain demonstrating the deposition of collagen in the subendocardium with presence of blood vessels (blue color, arrow ×20)

CONCLUSION

We document a case of biventricular EMF in a patient who had eosinophilic heart disease in the past attesting to the role of eosinophilia in the pathogenesis of EMF.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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