CASE REPORT

ENDOMYOCARDIAL FIBROSIS

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Abstract. We present a case report of a 26-year-old male from Bulandsahar, India. The patient presented with right heart failure. Evaluation revealed peripheral eosinophilia. An echocardiogram and MRI showed biventricular hypertrophy with obliteration of the ventricular apices, typical of endomyocardial fibrosis. This condition is rare in Bulandsahar, India.

INTRODUCTION

One of the most common causes of right heart failure is left heart failure. Rheumatic heart disease is prevalent in India, and is one of the most common causes of left heart failure leading to right heart failure, particularly in the younger population. Other causes of heart failure include cardiomyopathy, pulmonary disease and congenital heart disease. Endomyocardial fibrosis (EMF) is particularly common in tropical and subtropical countries within 15 degrees of the equator (Kutty *et al*, 1996). EMF is found in southern India, particularly in Kerala. This case is from northern India (Uttar Pradesh), 28.4 degrees north of the equator.

CASE REPORT

A 26-year-old male resident of Bulandsahar, Uttar Pradesh, India presented to the hospital with a history of dyspnea which progressed from NYHA class 1 to 3 during the previous 3 months, and a history of distension of the abdomen followed by swelling of his feet and facial puffiness for 2 months. He had a history of bronchial asthma (mild intermitant) for the past 11 years and used inhalers on an as needed basis. He had no past history of rheumatic fever, cyanosis at birth or pulmonary tuberculosis. Physical examination revealed an raised JVP, pedal edema and facial puffiness. Cardiovascular examination revealed a normal S1 and S2 with no abnormal heart sounds heard. He had hepatomegaly and free fluid in the abdomen. He had decreased air entry in the basal lungs bilaterally. The patient was investigated for restrictive cardiomyopathy and chronic constrictive pericarditis. A complete blood count showed eosinophilia with an absolute eosinophil count of 900. Stool microscopy and culture were normal. The rest of the metabolic profile was normal. The ECG showed low voltage complexes in the limb leads, left atrial enlargement and poor R wave progression. The CXR showed cardiomegaly with bilateral pleural effusions.

Echocardiogram showed biventricular hypertrophy with obliteration of the apex, diastolic dysfunction, mild mitral regurgitation and a large pericardial effusion (Fig 1). MRI showed biventricular hypertrophy with

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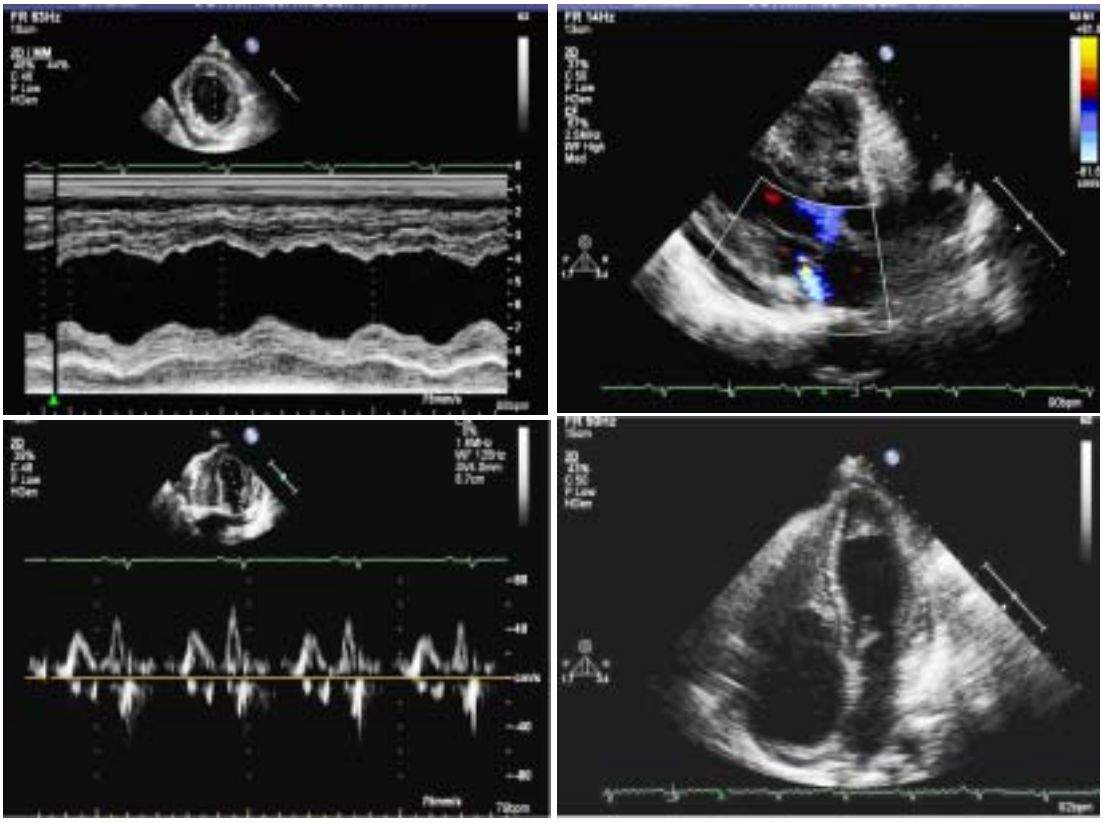

Fig 1–Echocardiogram showing biventricular hypertrophy with obliteration of the apex, mitral regurgitation, diastolic dysfunction and a pericardial effusion.

obliteration of the apex (Fig 2). A diagnosis of extensive biventricular EMF was made. Endomyocardial biopsy was not done as it is not required for diagnosis and carries a risk for embolism. The patient was managed symptomatically and referred to a higher center for surgical treatment. The patient did not follow up after initial cosultation and died of his illness 3 months later according to a relative.

DISCUSSION

EMF was first described by Davies in 1948. It affects mainly children and young adults of low socioeconomic status and is recognized as an important cause of heart disease in Africa, southern India and Brazil (Kutty *et al*, 1996).

EMF is characterized by fibrous endocardial inflow lesions of the right, left or both ventricles, and often involves the atrioventricular valves resulting in valvular regurgitation (Freers *et al*, 2000). Oslen Ogunowo (1983) described 3 phases of EMF: the necrotic phase, the thrombotic phase and the fibrotic phase.

The pathogenesis of EMF remains unclear. There is an inconstant association between eosinophilia and EMF (Roberts *et al*, 1969; Andy *et al*, 2001). Physical findings depended on the extent and distribution of the disease. In those with right ventricular involvement, JVP elevation, ascites and edema may be present. Signs of pulmonary congestion are present in patients with left heart disease (Gupta *et al*, 1989).

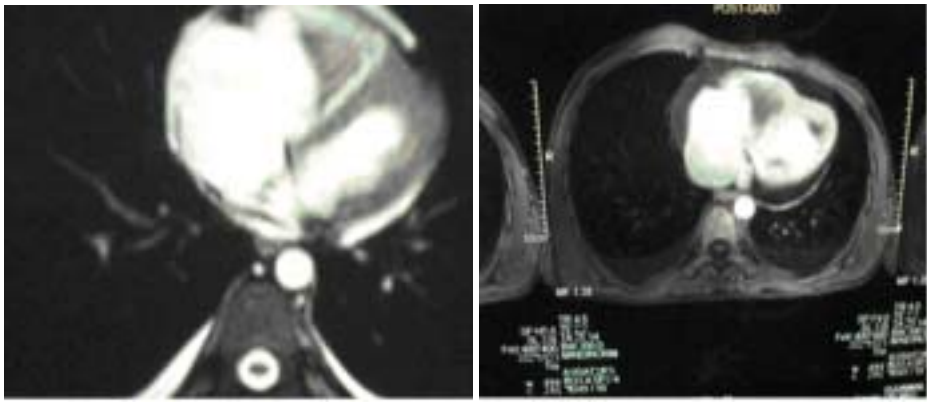

Fig 2–MRI showing biventricular hypertrophy with obliteration of the apex.

The diagnosis is made by echocardiographic examination showing obliteration of the apex of the ventricles with a variable degree of regurgitation of atrioventricular valves (Freers *et al*, 1996). Cardiac catheterizations demonstrates distortion of cardiac chamber morphology by fibrosis and obliteration of the ventricular apices also called the Mushroom Sign, along with varying degrees of mitral and tricuspid regurgitation (Cockshott, 1965). MRI shows obliterative changes in the ventricles, atrial dilatation and regurgitation of the atrioventricular valves. Delayed enhancement MRI also allows the detection of subendocardial fibrosis with good histopathological correlation (Ricardo *et al*, 2005).

The medical treatment of EMF is often difficult and not particularly effective. When EMF has reached the fibrotic stage, surgery offers symptomatic improvement and is the treatment of choice (Moreas *et al*, 1999).

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