Case Report

Right Ventricular Endomyocardial Fibrosis

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Abstract

Endomyocardial fibrosis is a variety of restrictive cardiomyopathy, in which endocardium of one or both ventricles is thickened markedly with involvement of underlying myocardium. Partial obliteration of ventricular cavities by fibrous tissue and thrombus causes diastolic dysfunction with increased resistance to ventricular filling. Systolic function is well preserved till late stages. Biventricular or isolated left ventricular involvement is common. Isolated right ventricular involvement is relatively uncommon. Case reports on endomyocardial fibrosis have declined in literature. In India, endomyocardial fibrosis is mainly reported from Kerala. A case of right ventricular endomyocardial fibrosis from West Bengal is reported here. Isolated right sided endomyocardial fibrosis, massive right atrial enlargement, complete disorganization of tricuspid valve, massive pericardial effusion, normal absolute eosinophil count and its sporadic occurrence outside 15° of the equatorial belt were interesting features in this case of endomyocardial fibrosis. X-ray features were typical of pericardial effusion masking underlying endomyocardial fibrosis. Endomyocardial fibrosis is a neglected research field. It needs more attention from biomedical researchers.

Introduction

Endomyocardial fibrosis (EMF) is a variety of oblitative restrictive cardiomyopathy of tropical and subtropical regions of the world. Endocardium of either or both ventricles is thickened markedly with involvement of the underlying myocardium. Obliteration of ventricular cavities by fibrous tissue and thrombus contributes to increased resistance to ventricular filling leading to diastolic dysfunction. In India, it is mainly reported from Kerala, while other parts of the country (e.g., Chandigarh) have reported relatively few cases. Reports of new cases of EMF have declined in medical literature.

Case report

A 35-year-old married, Hindu female of lower socioeconomic class from Kolkata, West Bengal presented with shortness of breath on exertion for two years. She also had fatigue and fullness of abdomen for same duration. She had occasional history of arthralgia without any joint swelling. She had no history of hair loss, oral ulcer, and red eyes. Her bladder and bowel habit was normal. She had no past history suggestive of filariasis, malaria, and toxoplasmosis. She had no history of conception and she never travelled outside West Bengal.

On examination, her blood pressure was 110/70 mmHg. Pulse was 110/min, irregularly irregular. Mild oedema and jaundice were present. Nutrition was subnormal. Build was average. On cardiovascular examination, apex was palpable at left 5th intercostal space, half inch medial to midclavicular line and character was normal. S1, S2 were audible but soft. No clear murmur was heard (as tricuspid annulus was grossly dilated and complete disorganization of the tricuspid valve was present, there was not enough turbulence to produce murmur). Respiratory and nervous system examination revealed no abnormality.

Blood picture revealed Hb 13.5 gm%, ESR 15 mm/hour, total leucocytes count 5600/μm mm including neutrophil 58%, lymphocyte 36%, eosinophil 6% and platelets 1.4 lakhs/μm mm. Absolute eosinophil count was 336/μm mm. Her blood group was O+ve. Blood sugar, urea, creatinine and Na+ and K+ levels were normal. Liver function test revealed bilirubin 1.4 mg% (conjugate 0.9 mg%, unconjugate 0.5 mg%), SGPT 72 unit/L, SGOT 58 U/L, total protein 8 gm% including albumin 4.3 gm% and globulin 3.7 gm%, and ALP 396 U/L (normal adult value 64-306U/L). ELISA for HIV I and II was negative. Urine routine examination was normal. Chest x-ray showed gross cardiomegaly with features of pericardial effusion (Figure 1). Ultrasonography showed enlarged heterogeneous liver and dilated hepatic veins. Moderate ascites and pericardial effusion were detected. ECG showed atrial fibrillation with non-specific ST-T changes.

Echocardiography showed giant size right atrium (without any thrombus), thickened right ventricular (RV) wall with reduced cavity size (Figures 2-5). Endocardial irregularities with bright echogenic areas suggest fibrotic changes in the right ventricular walls. Tricuspid valve showed thickened and shrivelled up leaflets, dilated annulus and passive flow across the valve to and from the small sized right ventricular cavity in diastole and systole respectively. Left ventricular (LV) cavity and wall thickness were within normal limits. Mild generalised LV wall hypokinesia and mild LV systolic dysfunction was present. LV diastolic compliance was reduced (restrictive pattern). Left atrium was mildly enlarged, with no mass in the appendage or body. Aortic valve and pulmonic valves were normal. Interventricular and interatrial septums were intact. Massive pericardial effusion was present. No evidence of impending tamponade or constrictive pericarditis was present. Pericardial fluid study showed exudative effusion; adenosine deaminase value did not support tuberculosis. Findings were suggestive of restrictive cardiomyopathy with right ventricular EMF. Patient was treated conservatively with diuretics, digoxin and anticoagulant. Her symptoms improved to some extent with reduction of abdominal distension.

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Discussion

The pathologist, Jack N. P. Davies identified EMF in Uganda in 1948. Since then, reports of EMF have come from other parts of tropical Africa, South Asia and South America that are within 15° of the equator.

Several hypotheses exist regarding aetiopathogenesis of EMF indicating the role of filarial infections, helminths, eosinophilia, and malnutrition. But, geographical distribution of EMF is not related to the prevalence of filariasis and eosinophilia. Toxoplasma-related myocarditis, malaria and HIV-infection have also been described in association with EMF. The endemic variety of EMF can be related to high cerium and low magnesium levels. Coexistence of high incidence of EMF and deposits of monazite element cerium in soil in two coastal talukas in Kollam and Alapuzha districts in south Kerala in India support the geochemical hypothesis. EMF is postulated to be related to prolonged ingestion of tapioca (tuber) which accumulates cerium from the surrounding sand. Food contaminated by sand could also lead to accumulation of cerium in the body. EMF is frequent in lower socioeconomic groups, in children and women of reproductive age. Tremendous change in socioeconomic and

Fig. 1: Straight x-ray chest showing pericardial effusion (CT ratio is increased, cardiophrenic angles are acute, pulmonary vessels are not engorged).

Fig. 2

Fig. 3

Fig. 4

Fig. 5

Figs. 2-5: Apical four chamber view of two-dimensional echocardiograms showing right ventricular EMF with partial obliteration of the cavity (mainly at the apex with echogenic masses). Echogenic areas are visible throughout the right ventricular endocardium. Right atrium is grossly dilated. Pericardial effusion is present. Tricuspid valve shows thickened and shrunken up leaflets, and dilated annulus.
health status of Kerala over past four decades is associated with decline in new cases of EMF in young age groups. This decline parallels the decline of rheumatic fever in developed countries. Endomyocardial diseases causing restrictive obliterator cardiomyopathies include EMF and hypereosinophilic (Loeffler) syndrome. EMF is principally found in tropical countries (geographic area within 15° of the equator), while Loeffler endocarditis is seen mainly in temperate climate. Loeffler endocarditis is rapidly progressive, aggressive, affecting principally males, and associated with intense eosinophilia and thromboembolic phenomenon. It is usually related to parasitic infections, leukaemia, and immunologic reactions. In contrast, EMF has insidious onset, indolent course, no gender predilection and has an inconstant association with eosinophilia. It most often affects children and young adults.

EMF mainly involves the apex and subvalvular regions of one or both ventricles. It involves the inflow tracts and may affect the atroventricular valves. Tethering of papillary muscles leads to tricuspid and mitral regurgitation. Outflow tract remains unaffected. EMF may be biventricular, isolated left ventricular or occasionally isolated right ventricular. Right or left atrial enlargement occurs according to involvement of respective ventricles. Mural thrombi may develop in either ventricle, serving as a source of pulmonary and systemic emboli. Thrombi overlying the endocardial lesions may lead to widely distributed endocardial calcific deposits. Epicardial coronary arteries remain free of obstructive lesions.

Right ventricular EMF is associated with JVP elevation, ascites, and edema. Kussmaul’s sign may be present. Ascites may be disproportionate to the amount of peripheral edema (due to concomitant presence of protein-losing enteropathy and subsequent hypoalbuminemia). Supraventricular arrhythmias may occur. Echocardiographic findings include right ventricular thickening, obliterated apex, dilated right atrium, strong echoes emanating from endocardial surface, and abnormal septal motion in patients with tricuspid regurgitation (TR). Pericardial effusion is seen occasionally in both right and left ventricular EMF. A study from Nigeria described six patients in whom right ventricular EMF was accompanied by a large pericardial effusion. Aetiology of pericardial effusion is possibly inflammatory. EMF is to be considered as a pancarditis as all the layers are involved.

Right ventricular EMF should be distinguished from constrictive pericarditis. In contrast to constrictive pericarditis, the apex beat is usually easily palpable, and TR is more common in EMF. Ascites responds to diuretic therapy in EMF more than constrictive pericarditis. Other factors to exclude constrictive pericarditis are the presence of atrial fibrillation and cardiac enlargement due to aneurysmal dilatation of right atrium in EMF. Ebstein’s anomaly is to be excluded as it is associated with giant right atrium and TR. Endomyocardial biopsy may be helpful in establishing the diagnosis of EMF. As the disease is often focal, the biopsy may miss the pathological process, particularly when right ventricular biopsy is performed in a patient with left sided disease. Biopsy may dislodge a mural thrombus with resultant embolisation. So biopsy is rarely performed and left sided biopsy is not recommended. Management usually includes diuretics, after load reducing agents, and anticoagulation. Surgical treatment with resection of fibrotic tissue and valve repair or replacement may be helpful, but recurrence is common. Overall prognosis of EMF is very poor.

The presently reported case had isolated right ventricular EMF. X-ray features were typical of pericardial effusion, masking underlying EMF. Though she did not have left ventricular EMF, she had mild diastolic and systolic dysfunction of left ventricle with generalized LV wall hypokinesia. Massive right atrial enlargement, complete disorganization of tricuspid valve, massive pericardial effusion, normal absolute eosinophil count and its sporadic occurrence outside 15° of the equatorial belt were the interesting features in this case of EMF. EMF needs more attention from biomedical researchers.

References