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Case Report

A Heart Set in Stone: A Case of Extensive Cardiac Calcification

Roopali Khanna, Aditya Kapoor, Neetu Soni[1]

Departments of Cardiology and [1]Radiology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

ABSTRACT

Massive endocardial calcification is a rare entity. We describe a rare disease endomyocardial fibrosis associated with massive calcification of the left ventricle suspected on a chest X-ray and confirmed by echocardiography and computed tomography in an 18-year-old female presenting with breathlessness and congestive heart failure.

Key words: Echocardiography, endocardial calcification, endomyocardial fibrosis

INTRODUCTION

Cardiac calcification is not uncommon, but massive left ventricle endocardial calcifi cation is a rare entity. Endomyocardial fibrosis is a restrictive cardiomyopathy of unknown etiology that occurs almost exclusively in tropical and subtropical regions, particularly in some countries of Africa, India, and Brazil. The disease is characterized by irregular fibrous thickening of the endocardium in the apex and inflow tract of one or both ventricles. Superimposed thrombosis and endocardial calcification is seen in advanced cases. Herein, we report of a rare case of endomyocardial fibrosis associated with massive calcification of the left ventricle in a female patient presenting with progressive dyspnea and congestive heart failure.

CASE REPORT

An 18-year-old female presented with progressive shortness of breath since the last 6 months; she had an audible left ventricular (LV) S3, Grade III/VI pansystolic murmur, severe pulmonary hypertension, and elevated jugular venous pressure with giant c-v wave. Mild cardiomegaly, dilated main pulmonary artery segment, and dense ringed calcification within the cardiac silhoutte were noted on the X-ray chest [Figure 1a].

Electrocardiogram revealed right axis deviation with biatrial enlargement. Transthoracic and transesophageal echocardiography revealed biatrial dilatation, normal left ventricle systolic function, and a hyperechoic, "glittering" appearance of the left ventricle endocardium with fibrocalcific infiltration and obliteration of the left ventricle apex [Video 1]. The calcification extended up to the mitral valve apparatus [Figure 2a and b]. Transmitral and tissue Doppler flows confirmed a restrictive physiology [Figure 2c and d]. No significant involvement of the right ventricle was noted. Fluoroscopy revealed scattered massive calcification on the cardiac silhouette along the left ventricle region [Figure 1b]. A 64-slice cardiac computed tomography (CT) confirmed extensive myocardial calcification along the left ventricle anterolateral and inferior segments, mitral annulus, and the left atrium [Figure 1c and d].

Her blood investigation showed hemoglobin of 14 g/dL, total leukocyte count of 8200/mm^3 with 65% neutrophils, 28% lymphocytes, 5% monocytes, and

Address for correspondence: Dr. Roopali Khanna, Department of Cardiology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Raebareli Road, Lucknow - 226 014, Uttar Pradesh, India.
E-mail: drroopalik@gmail.com

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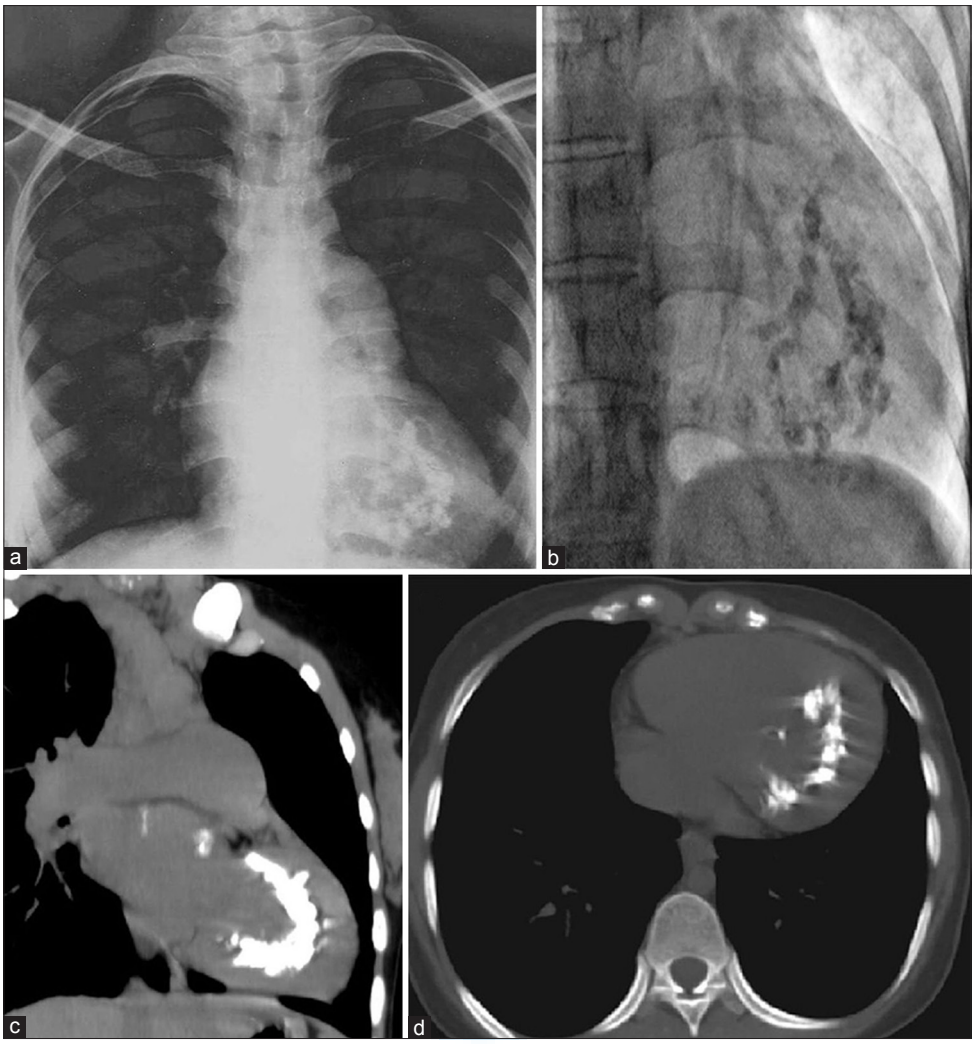


Figure 1: Chest X-ray posterioranterior view. (a) Heterogeneous opacification in the cardiac silhouette. Fluoroscopy in right anterior oblique view, (b) calcification in the left ventricle area. 64-slice computed tomography thorax sagittal, (c) and transverse view, (d) massive calcification of left ventricle endocardium anterolateral and inferior segment and calcification of left atrium superior margin

2% eosinophils. The blood chemistry including serum creatinine, bilirubin, total protein, albumin, calcium, magnesium, phosphate, and thyroid and parathyroid hormone levels was normal. Tests for antinuclear antibody and anti-HIV were negative. There was no evidence of parasitic infection. Features of progressive heart failure, restrictive cardiac physiology, and fibrocalcific infiltration/obliteration of left ventricle apex and inflow region strongly suggested a possibility of endomyocardial fibrosis. Although endomyocardial biopsy was planned, the patient declined for further intervention.

DISCUSSION

Cardiac calcification is not uncommon, but massive left ventricle endocardial calcification is a rare entity. This rare condition is associated with metastatic deposition, infarction, or other endocrine disorders. Metastatic calcification is due to deposition of calcium salts in previously normal tissue due to disturbance in calcium/phosphorus metabolism. Most commonly, it occurs due to persistently elevated calcium levels as in primary hyperparathyroidism, chronic renal disease,[1] hypervitaminosis D, widespread bone destruction from metastases, or myeloma. Dystrophic calcification is deposition of calcium salts in previously damaged tissue with normal calcium metabolism. It commonly occurs in patients with underlying ischemic heart disease, primary myocardial disease.

Patients with myeloproliferative disorder may have associated Loeffler endocarditis secondary to chronic prolonged eosinophilia which may be complicated by calcification of the endocardial sclerotic lesions.[2] In hyperparathyroid condition, there is an increased prevalence of cardiac structural abnormalities such as left ventricle hypertrophy, calcification, and deranged functional properties of the heart.[3]

Endomyocardial fibrosis is a disease of unknown etiology. In 1984, Silver *et al*.[4] described the first case of massive endocardial calcification of the left ventricle,

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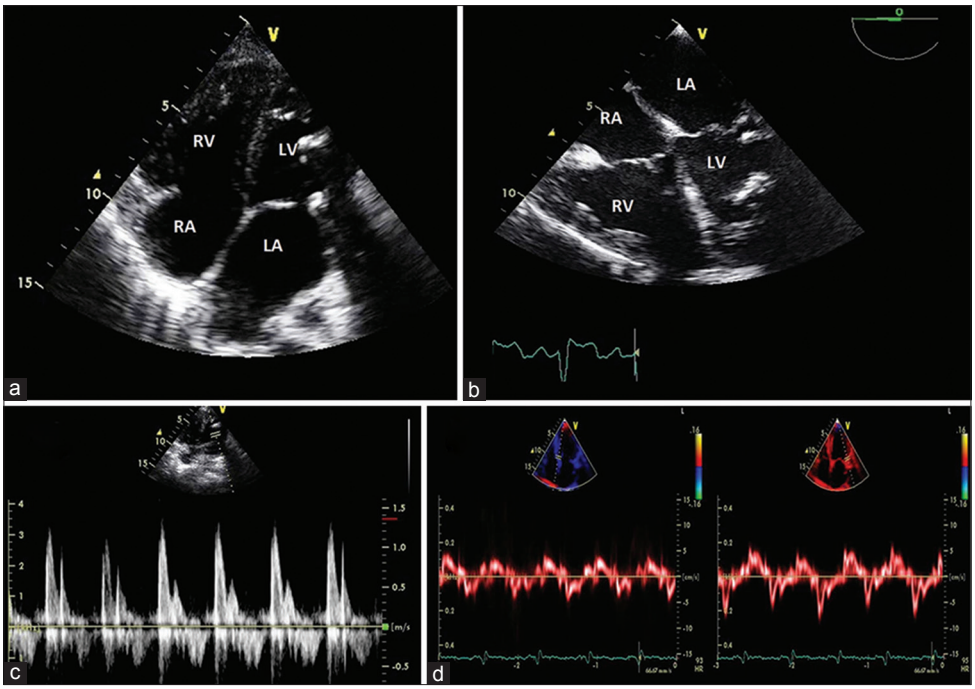


Figure 2: Transthoracic (a) and transesophageal echocardiography, (b) revealed biatrial dilatation, hyperechoic left ventricular endocardium with fibrocalcific infiltration of the left ventricular apex; the calcification extended up to the mitral valve apparatus, transmitral (c) and tissue Doppler flows, (d) confirmed a restrictive physiology

suggesting it was a different entity causing restrictive cardiomyopathy. This suggestion was refuted by Lengyel *et al*.,[5] who suggested that the endocardial calcification was a clue for the diagnosis of endomyocardial fibrosis. The etiology of endomyocardial fibrosis remains unclear. Most of the patients of endomyocardial fibrosis present with symptoms of heart failure, generalized weakness, and fever. There is no definitive treatment of endomyocardial fibrosis, and symptomatic therapy is usually given for heart failure and prevention of thrombus. However, the prognosis of endomyocardial fibrosis is poor after manifestation of heart failure symptoms.[6]

In our patient, the blood biochemistry profile revealed normal blood counts, renal profile, and parathyroid hormone. The ventricular systolic function and wall motion were normal on echocardiography. Hence, calcification secondary to chronic renal failure, myeloproliferative disease, and ischemic heart disease were unlikely. Her echocardiography showed massive LV and atrial calcification with restrictive morphology. Endomyocardial biopsy would have been definitive. Features of progressive heart failure, restrictive cardiac physiology, and fibrocalcific infiltration/obliteration of LV apex and inflow region strongly suggested a possibility of endomyocardial fibrosis. Endomyocardial fibrosis is characterized by progressive heart failure, and a restrictive physiology secondary to subendocardial fibrosis involving ventricular apices and inflow tracts and multimodality imaging is often useful in such situations. This shows that a rare disease such as endomyocardial fibrosis associated with massive calcification of the left ventricle may be suspected on a simple chest X-ray and confirmed by CT.

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Conflicts of interest

There are no conflicts of interest.

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