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Case Report

Endomyocardial fibrosis with right ventricular aneurysm mimicking ARVC – A case report from India

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ABSTRACT

A 48-year-old man presented with chronic right sided heart failure. 2D echocardiography revealed the classical features of left ventricular endomyocardial fibrosis with a prominent right ventricular apical aneurysm. Right ventriculography further defined the aneurysm. Cardiac magnetic resonance images revealed a thin-walled, apical aneurysm of right ventricle with multiple septations and marked obliteration of left ventricular apex. A delayed-enhancement sequence after the infusion of contrast, demonstrated a hypersignal in the subendocardium, consistent with the right ventricular involvement of endomyocardial fibrosis. This patient had classical features of left ventricular endomyocardial fibrosis, while on the right side the typical features were missing. This aneurysm may be a passing phase of the natural history of endomyocardial fibrosis before the development of burned out stage. This aneurysm may later develop thrombus, and which may progress to fibrosis and apical obliteration. Endomyocardial fibrosis with right ventricular aneurysm has not heretofore been reported in the medical literature.

1. Introduction

Endomyocardial fibrosis (EMF) is the most common cause of restrictive cardiomyopathy in tropical climates.[1] It is characterised by fibrotic deposits on the endocardial surfaces of either or both ventricles and is frequently associated with thromboembolic disease, arrhythmias and complications of chronic heart failure.[2] This report describes a rare case of EMF with classical features of left ventricular involvement, but with right ventricular (RV) apical aneurysm which mimics arrhythmogenic right ventricular cardiomyopathy (ARVC).

2. Case report

A 48-year-old man was admitted with history of abdominal distention and bilateral pedal oedema for last 10 years. He was treated for right sided heart failure several times in the past, and one of these admissions prompted referral to a cardiac specialist. He denied having paroxysmal nocturnal dyspnoea, orthopnoea or wheezing. There was no history of chest pain, palpitation or syncope. He was a lifelong nonsmoker who abstained from alcohol and had no known significant family history. On examination he had grossly distended abdomen and bilateral pitting pedal oedema. His pulse rate was 70/min, irregular and blood pressure was 130/80 mm Hg. His oxygen saturation level was 94% on room air. Jugular venous pressure was markedly elevated up to the level of ear lobes with a prominent 'y descent'. Cardiovascular examination revealed normal heart sounds with a systolic murmur of tricuspid regurgitation. On abdominal examination there was shifting dullness and hepatomegaly. Other systemic examinations were normal as is the remainder of physical examination.

The clinical features were more suggestive of chronic right sided heart failure. A full blood count, routine biochemical screen and serum bicarbonate levels were normal. Absolute eosinophil count was with in normal range. The 12-lead electrocardiogram showed intermittent atrial fibrillation. Chest radiography revealed cardiomegaly due to significant right atrial enlargement (Fig. 1A). Echocardiogram revealed grossly dilated right atrium, reduced right ventricular volume with a prominent apical aneurysm (Fig. 1B, Supplementary Video 1 and 2). Restrictive flow pattern at both atrioventricular-valves and severe tricuspid

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regurgitation were noted. Left ventricular apex was obliterated with calcification. There was significant pericardial effusion. Endocardial calcification was detected during coronary angiography (Fig. 1C). Ventriculography confirmed right ventricular aneurysm with dilated right ventricular outflow tract (Fig. 1D, Supplementary Video 3) and obliteration of left ventricular apex (Fig. 1E, Supplementary Video 4). Pressure tracings of right atrium showed classical 'c-v waves' with prominent 'y descent' (Fig. 1F). Pulmonary artery systolic pressure was 28 mm Hg (Fig. 1G). Pullback tracings from RV aneurysm to RV cavity proper showed a pressure gradient of 15 mm Hg (Fig. 1H). The diagnosis of biventricular endomyocardial fibrosis with right ventricular apical aneurysm was made. The diagnostic criteria for EMF proposed by Mocumbi et al. was satisfied but further investigations were planned in view of unusual RV aneurysm.[1] The cardiovascular magnetic resonance imaging (CMR) study demonstrated the obliteration (Fig. 2A) and aneurysm (Fig. 2B) of left and right ventricle respectively. Dilated right atrium and systemic venous congestion were noted (Fig. 1C). Delayed enhanced phase showed high signal intensity in the right ventricular subendocardium, suggestive of fibrous tissue deposits (Fig. 2D). The patient was treated with diuretics and anticoagulation.

3. Discussion

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.[2] Echocardiography is the most useful and standard technique for diagnosing this condition.[1] RV apical aneurysm with multiple septations was an unusual echocardiographic finding in this patient. Ventricular aneurysm is not a classical echocardiographic feature of EMF and hence conditions like ARVC and diverticula (congenital or acquired) were kept as differential diagnosis. Cardiovascular magnetic resonance imaging (CMR) is complimentary to transthoracic echocardiography for diagnosing EMF and is done only for research purposes.[2] In this instance CMR was performed to rule out ARVC and confirm the diagnosis. CMR with Gd-DTPA infusion allowed the characterisation of subendocardial fibrosis or scarring by delayed enhancement. Scarring or necrosis of the endomyocardium increases gadolinium concentration, which explains the delayed hyperenhancement and confirms the diagnosis of EMF.[3] Furthermore, the characteristic CMR feature of ARVC like transmural fatty replacement of myocardium or diffuse RV myocardial thinning were not observed in this patient.[4] Cardiac catheterisation demonstrated the characteristic right atrial 'c-v waves' with 'y descent' of restrictive physiology and ventriculography disclosed unique RV apical aneurysm. Reduced RV volume and absence of hypertrophied trabeculae are against the diagnosis of ARVC.[5] Biventricular EMF with prominent aneurysm of RV was a unique finding in this case. EMF with RV aneurysm has not heretofore been reported in world literature. The aetiology of EMF is speculative and the pathogenesis remains an enigma. Patients with EMF generally presents with features of haemodynamic compromise due to the late fibrotic stage of illness.[5] In this patient, RV aneurysm may be a passing phase of the

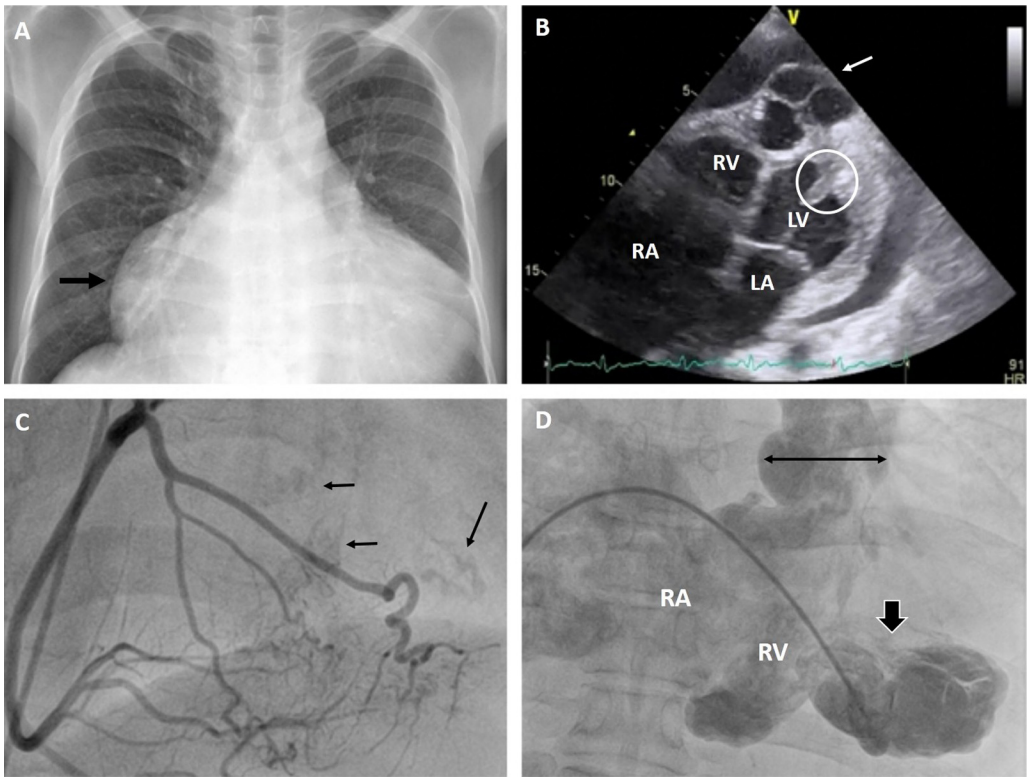

Fig. 1. (A) Chest radiograph. Note the enlargement of cardiac silhouette due to significant right atrial dilatation (arrow). Left atrial enlargement is also noted. (B) 2D echocardiogram modified apical 4-chamber view. Left atrium is enlarged (size 46 mm). Left ventricular apex is obliterated with calcification (encircled). Note enlarged right atrium (size 54 mm) and large aneurysm at right ventricular apex (arrow) with multiple septations. Significant pericardial effusion is noted. (C) Arrows delineate endocardial calcific deposits on the left ventricle detected during coronary angiography. Epicardial coronaries are free of lesion. (D) Cine angiographic frames of the right ventricle in the postero-anterior view at systole showing large apical aneurysm with septations (single headed arrow). Right atrium is dilated. Note the dilated right ventricular outflow tract (double headed arrow). (E) Cine angiographic frames of the left ventricle in the right anterior oblique view at end-diastole showing obliteration of ventricular apex. (F) Pressure tracings of right atrium showing classical 'CV waves' with prominent 'y' descent (Arrow). (G) Pulmonary artery pressure tracings showing pulmonary artery systolic pressure of 28 mm Hg. (H) Pull-back pressure tracing from right ventricular aneurysm to the ventricular cavity proper showing a pressure gradient of 15 mm Hg (arrow). RA – right atrium, RV – right ventricle, LV – left ventricle.

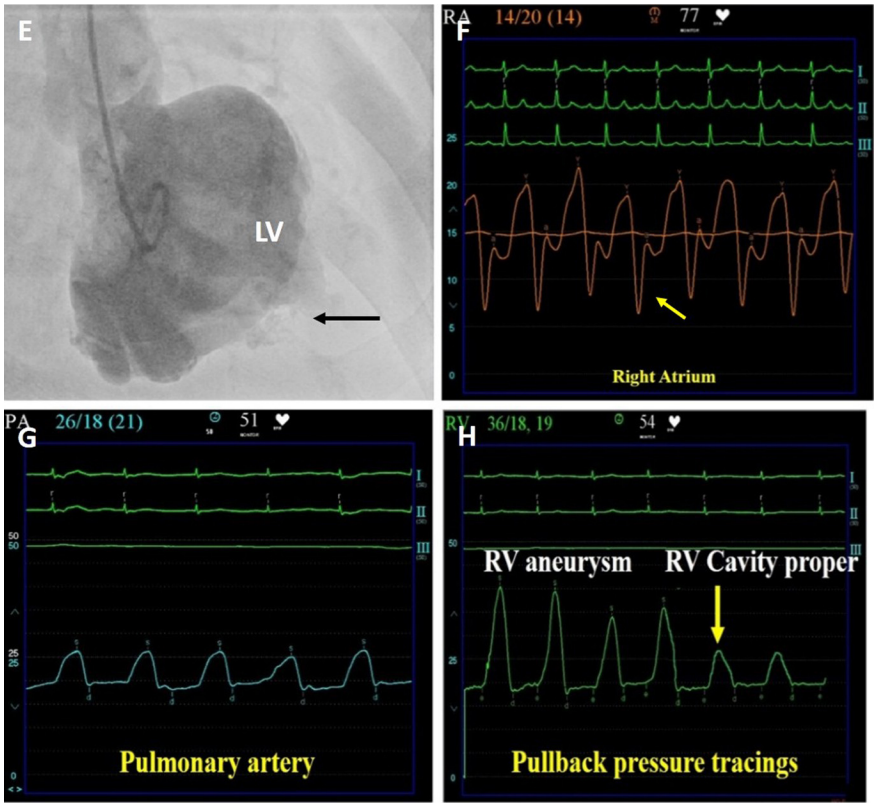

Fig. 1. (*Continued*).

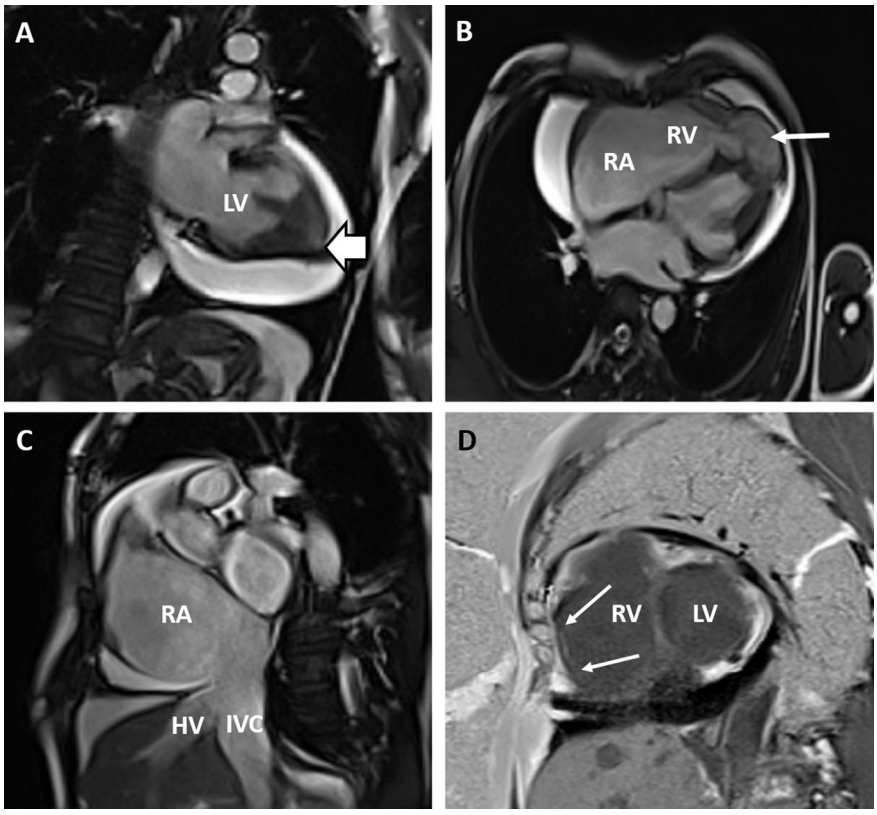

Fig. 2. (A) CMR imaging, gradient echo sequence (white-blood pool phase) 2-chamber view, 2 min after 0.2-mmol/kg Gd-DTPA infusion demonstrates left ventricular apical obliteration (arrow). Note the apical filling of the left ventricle exhibiting a lower signal than adjacent myocardium. Significant pericardial effusion is noted. (B) Same sequence, four chamber view demonstrates small right ventricle with prominent apical aneurysm (asterisk). (C) Same sequence, short axis view showing dilated right atrium, hepatic vein and inferior venacava. (D) Delayed enhanced sequence in short axis 10 min after Gd-DTPA infusion. Right ventricular subendocardium showing high signal suggesting fibrosis (arrows). HV – hepatic vein, IVC – inferior venacava, CMR imaging – cardiac magnetic resonance imaging. Gd-DTPA – gadolinium diethylene triamine penta acetic acid.

natural history of EMF before the development of burned out stage or the chronically elevated RV pressure might have contributed to the development of aneurysm in an actively inflamed ventricle. Whether an active inflammatory or thrombotic stage of the disease exists or fibrotic stage is a continuation, and whether this aneurysm itself is a squeal of inflammation is open to speculation.[6] In this case, the LV apical obliteration is diagnostic of LVEMF while on the right side the classical features of EMF are missing (especially obliteration of ventricular apex). But, humongously dilated right atrium, characteristic right atrial 'c-v waves' with 'y descent', restrictive flow pattern across tricuspid valve, significant tricuspid regurgitation and imaging evidence of significant systemic venous congestion are impressive findings in favour of right ventricular involvement of EMF. We conjecture that, this aneurysm may later develop thrombus, and which may progress to fibrosis and apical obliteration. Visualisation of RV apical aneurysm by conventional ventriculography and confirmation of subendocardial fibrosis by delayed enhancement CMR provided a comprehensive tool for diagnosing an uncommon manifestation of a rare, neglected cardiovascular disease. Endomyocardial biopsy was deferred due to poor general condition of the patient.

4. Conclusion

Biventricular EMF with RV aneurysm is a very unique finding not so far reported in medical literature. Although there is a significant decline in the number of new cases of EMF in hospital admissions nowadays, it should be kept in mind as a differential diagnosis of RV aneurysm; especially in tropical countries.

Conflicts of interest

The authors have none to declare.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.ihj.2016.07.019.

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