CASE REPORT

Complex right atrial mass in endomyocardial fibrosis: a diagnostic dilemma

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SUMMARY

Endomyocardial fibrosis, though a vanishing disease from India, remains an important cause of heart failure in children, adolescents and young adults. It may be complicated with arrhythmias and thromboembolism and is an important cause of mortality and morbidity. Moreover, usual presentation of this condition is in advanced stage with poor prognosis. Ventricular endocardial fibrosis with organised thrombus is the hall mark of this entity. Presence of associated cardiac mass poses a diagnostic challenge. We present one such case of endomyocardial fibrosis, in which a large thrombus was seen adherent to the anterolateral wall of right atrium, posing further risk of thromboembolism with complex management issues.

right ventricular apical obliteration, small ventricular cavity and normal tricuspid valve leaflets attachment (figure 1B). RA was grossly dilated with large mass in it (figure 1B,C). The mass was attached to the anterolateral wall and showed no perfusion uptake or contrast enhancement (figure 1D,E) leading to the diagnosis of a thrombus. Characteristic three-layered appearance was seen on late postcontrast scans (figure 1E) in the region of right ventricle apex with the inner layer showing no perfusion due to presence of thrombus, a middle layer of late gadolinium enhancement reflecting the fibrotic tissue and an outer layer of almost normal myocardium. In addition to this, it showed presence of ascites, bilateral pleural effusions and pericardial effusion (figure 1B,C,F).

BACKGROUND

Endomyocardial fibrosis (EMF) is a form of restrictive cardiomyopathy, earlier reported commonly from coastal areas of Kerala in India. In endemic areas, it may account for upto 20% of all causes of heart failure in children and young adults.¹ Generally, the diagnosis is made in the advanced stage of the disease, and these patients usually die within 2 years of diagnosis. Incidence of the disease is decreasing with the improvement of the socioeconomic status and presently, it represents a vanishing disease from India. Death is usually from associated complications like arrhythmias and thromboembolism is an important cause of morbidity. Atrial thrombus is rarely seen in this condition. Moreover, a very large adherent thrombus has not been described so far in literature.

CASE PRESENTATION

A 27-year-old woman presented with gradually progressive shortness of breath and palpitations since last 6 years and generalised body swelling for last 4 years following febrile illness of about 2 weeks duration. This time, the patient presented with right heart failure with atrial fibrillation. Jugular venous pressure (JVP) was raised with presence of pedal oedema. There was mild eosinophilia with raised IgE levels. Chest radiograph showed enlarged globular heart with right atrial enlargement (figure 1A). ECG showed low voltage QRS complexes. Echocardiography revealed hugely dilated right atrium (RA) with large mass (10×9 cm) and normal sized left heart chambers. The patient was referred to us for MRI examination for cardiac evaluation and characterisation of the RA mass. MRI demonstrated

OUTCOME AND FOLLOW-UP

The patient was started on oral anticoagulation in view of increased risk of pulmonary thromboembolism owing to presence of right atrial thrombus. A follow-up transthoracic echocardiogram after 2 weeks demonstrated mild reduction in the right atrial thrombus size. She was also on continued medical management for heart failure and is now planned for bidirectional Glenn.

DISCUSSION

Pathophysiology of EMF involves focal or diffuse endocardial thickening involving mainly the inflow, apices, and the subvalvular region leading to valvular regurgitation, diastolic dysfunction and obliteration of the ventricular apices. Biventricular involvement occurs in about 50% cases with pure left ventricular involvement in 40% and pure right ventricular involvement in the remaining 10% cases.² Ventricular endocardial fibrosis with organised thrombus formation produces characteristic trilayered appearance. Advanced right-sided disease may show propensity of thrombus formation especially in the right atria due to slow flow of blood through cardiac chambers, however, atrial thrombus is less frequently observed as compared with ventricular thrombus. Fibrosis involving the right ventricular myocardium increases the stiffness of the heart, resulting in a restrictive physiology. Right ventricular stiffness along with atrioventricular valvular regurgitation results in atrial enlargement. Stasis of blood in the grossly dilated RA along with postulated prothrombotic tendencies in EMF result in atrial thrombus formation.

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Figure 1 Frontal chest radiograph (A) reveals cardiomegaly with right atrial enlargement. Bright blood steady-state free precession images in the four chamber (B) and short axis (C) views show presence of a large adherent thrombus (*) within the right atrium (RA) along its right anterolateral wall with obliteration of apex of the right ventricle (RV) (region indicated by dotted circle). Bilateral pleural effusion (black arrows) and pericardial effusion (dotted arrow) were also noted. Perfusion image (D) shows no vascularity within the RA mass. Late gadolinium enhancement image (E) reveals non-enhancing RA mass with subendocardial enhancement (black arrows) in the region of RV apex with a small linear overlying thrombus along the RV apex (black arrowhead), giving the typical trilayered appearance with the inner layer showing no perfusion due to presence of thrombus, a middle layer of late gadolinium enhancement reflecting the fibrotic tissue and an outer layer of almost normal myocardium. The linear thrombus is better appreciated on the bright blood image (F). LV, left ventricle.

Presence of associated cardiac mass poses a diagnostic challenge. Echocardiography remains the initial investigation of choice. Cardiac MRI is one of the non-invasive confirmatory investigations in diagnosis of EMF and elucidation of associated cardiac masses (current case highlights the importance of delayed gadolinium enhancement in the diagnosis of EMF and cardiac thrombus). Moreover, MRI has prognostic implications as it allows characterisation and quantification of fibrosis

with good histopathological correlation.^{3 4} Presence of large atrial thrombus poses additional risk of thromboembolism and development of pulmonary hypertension, thus complicating management issues. Medical management is largely supportive with surgical endocardectomy (with or without atrioventricular valve repair) as definitive treatment method in EMF; however, it carries high postoperative mortality and chances of recurrence after surgery.⁵

Learning points

- ▶ Ventricular endocardial fibrosis with organised ventricular thrombus is the hall mark of endomyocardial fibrosis (EMF).
- ▶ Cardiac MRI plays an important role in identification and characterisation of associated cardiac mass in addition to the characteristic findings of EMF.
- ▶ Adherent atrial thrombus is rare, posing further risk of thromboembolism with complex management issues.

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