Cardiac involvement in hypereosinophilic syndrome

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ABSTRACT

A 9-year-old boy with hypereosinophilic syndrome (HES) was referred for cardiac magnetic resonance (CMR) imaging following an abnormal echocardiogram that showed a large mass layered on the inferolateral wall of the left ventricle, causing secondary severe mitral regurgitation. Cardiac involvement in HES usually affects the ventricular apex. In our case, CMR confirmed the presence of a large mural thrombus of 0.9 cm × 4.2 cm. This unusual cardiac involvement in HES was diagnosed in its intermediate thrombotic stage. CMR is very sensitive and specific in staging the disease. It explained the etiology of mitral regurgitation and guided therapy, especially when echocardiography was nonconclusive.

Keywords: Cardiac involvement, cardiac magnetic resonance, hypereosinophilic syndrome

A 9-year-old boy recently diagnosed with acute lymphoblastic leukemia (ALL) and hypereosinophilic syndrome (HES) on bone marrow biopsy, with a white-cell count of 49,200/μL and 80% eosinophils, was referred for cardiac magnetic resonance (CMR) imaging because of an abnormal transthoracic echocardiogram that showed a large mass on the inferolateral wall of the left ventricle [Figure 1, arrows] and severe mitral regurgitation [Video 1]. The CMR confirmed a 0.9 cm × 4.2 cm mass layered on the endocardium of the basal to distal inferolateral wall with hypokinesis of the corresponding myocardial segments [Video 2]. The mass was also attached to the posterior mitral valve leaflet and subvalvular apparatus causing significant restriction and secondary severe mitral regurgitation [Figure 2 and Video 2]. On perfusion imaging with intravenous gadolinium, there was hypoperfusion of the mass [Video 3]. There was low signal within the mass consistent with a large thrombus [Figure 3, white arrow]. There was, however, significant late gadolinium enhancement of the subendocardium (basal to the distal inferolateral wall) consistent with inflammation and/or scar [Figure 3, arrowhead].

A variety of conditions such as leukemias and lymphomas can be associated with eosinophilia and secondary manifestations of HES.[1] Patients with HES can develop ALL and patients with ALL can present clinically with HES.[1] The high resolution of CMR makes tissue characterization possible, and the increasing experience makes CMR promising for diagnosis and follow-up.[2] A hyperintense myocardial area on T2-weighted images is suggestive of increased free-water content due to myocardial edema (inflammation) and/or necrosis.[3] In HES, this is particularly seen in the ventricular apex. Cardiac involvement in HES is common and accounts for 40% of cases.[4] CMR has a high sensitivity of 88% ± 9% and specificity of 99% ± 2% for detecting thrombi.[5,6] Overlying thrombus is identifiable as a low-signal mass on the delayed enhancement images, which does not deform on tagged images. Three stages define the progression of the disease: The early stage, with myocardial eosinophilic infiltration, followed by an intermediate thrombotic stage, and finally, a late fibrotic stage, resulting in endomyocardial fibrosis.[3] In Figure 3, a characteristic three-layered image can be seen: A hypointense inner

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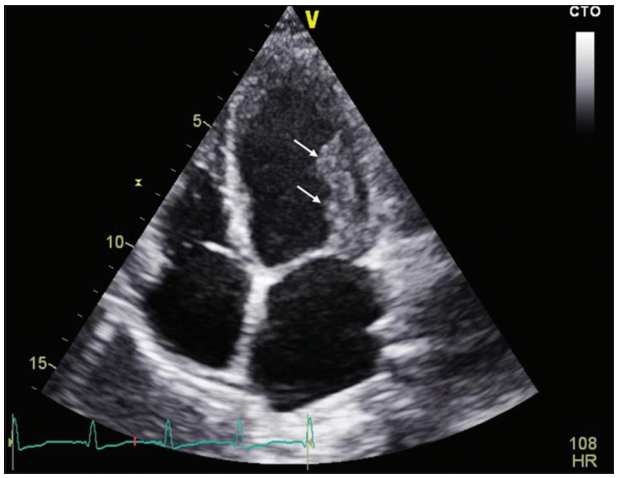

Figure 1: Transthoracic echocardiogram apical four-chamber view showing a large mass (arrows) on the inferolateral wall attached to the mitral valve leaflet

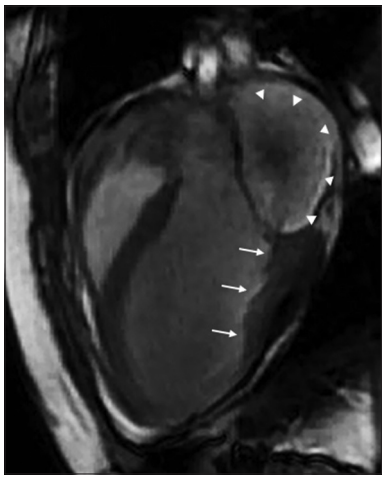

Figure 2: Cardiac magnetic resonance showing large thrombus (arrows) attached to the basal-distal inferolateral wall of the left ventricle and the posterior mitral valve leaflet, causing severe secondary mitral regurgitation (arrowheads)

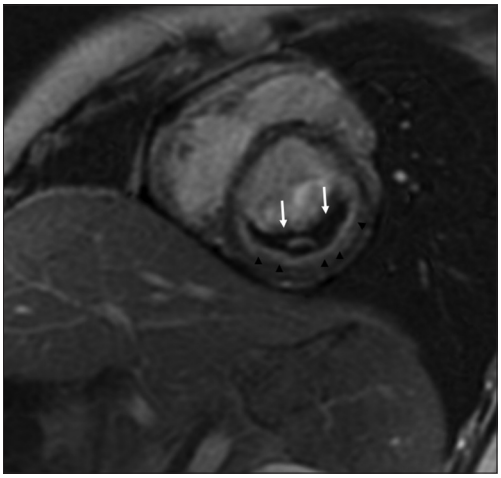

Figure 3: Cardiac magnetic resonance showing large hypointense mural thrombus (arrows) layered on the ventricular endocardium with the hyperintense signal of the subendocardium consistent with inflammation and/or scar (arrowheads)

rim of thrombus adjacent to a hyper-enhancement of the endocardium compared with the rest of the myocardium. The presence of a large mural thrombus attached to the inferolateral wall with secondary severe mitral regurgitation and centrally and posteriorly directed jets 3-4/4 (regurgitant volume 20 ml, regurgitant fraction 38%), as well as left ventricular impairment with reduced global systolic function (ejection fraction of 43%) with basal inferior/inferolateral hypokinetic walls, and distal inferior/apical akinetic walls underline the unusual extensive involvement of the pericardium and myocardium detected by CMR, which confirmed the diagnosis of HES in its intermediate thrombotic stage, explained the etiology of the mitral regurgitation, and guided therapy. Anticoagulation and steroid administration was recommended in addition to chemotherapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Brito-Babapulle F. The eosinophilias, including the idiopathic hypereosinophilic syndrome. Br J Haematol 2003;121:203-23.
2. Debl K, Djavidani B, Buchner S, Poschenrieder F, Heinicke N, Feuerbach S, *et al.* Time course of eosinophilic myocarditis visualized by CMR. J Cardiovasc Magn Reson 2008;10:21.
3. Mahrholdt H, Wagner A, Judd RM, Sechtem U, Kim RJ. Delayed enhancement cardiovascular magnetic resonance assessment of non-ischaemic cardiomyopathies. Eur Heart J 2005;26:1461-74.
4. Sen T, Gungor O, Akpinar I, Cetin M, Tufekcioglu O, Golbasi Z, *et al.* Cardiac involvement in hypereosinophilic syndrome. Tex Heart Inst J 2009;36:628-9.
5. Srichai MB, Junor C, Rodriguez LL, Stillman AE, Grimm RA, Lieber ML, *et al.* Clinical, imaging, and pathological characteristics of left ventricular thrombus: A comparison of contrast-enhanced magnetic resonance imaging, transthoracic echocardiography, and transesophageal echocardiography with surgical or pathological validation. Am Heart J 2006;152:75-84.
6. Ogbogu PU, Rosing DR, Horne MK 3rd. Cardiovascular manifestations of hypereosinophilic syndromes. Immunol Allergy Clin North Am 2007;27:457-75.