Eosinophilic heart disease in a paediatric patient

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A 12-year-old child with no previous medical history was referred with a 4-day history of cough, shortness of breath, and peripheral blood eosinophilia. Transthoracic echocardiography showed a soft tissue infiltrating the left ventricular free wall, the lateral mitral annulus, and the mitral valve leaflets. A soft tissue strand connecting the lateral left atrial wall and mitral leaflets across the mitral valve orifice was also identified, causing reduced opening and functional mitral stenosis. The diagnosis of Löffler endocarditis was made, and after 10 weeks of treatment with oral prednisolone, there was complete resolution of symptoms and of the infiltrative tissue with normalization of mitral valve function. The present case highlights some atypical features of eosinophilic heart disease-like occurrence in paediatric age, the complete preservation of the right ventricle and left ventricular apex, and the presentation with mitral stenosis compared with mitral regurgitation typically observed in the late phase of the disease.

Keywords
Inflammation • Mitral valve • Echocardiography • Endocardium

A 12-year-old child with no previous medical history was referred with a 4-day history of cough, shortness of breath, and peripheral blood eosinophilia. Examination was unremarkable except for a mild tachycardia. A 12-lead ECG showed sinus tachycardia, normal atrio-ventricular and intraventricular conduction. Laboratory values were notable for a haemoglobin 8.1 g/dL, white blood cell count of 47 170 cells/mL with absolute eosinophilia of 36 790. Troponin T was elevated to 0.14 μg/mL with a normal creatine kinase. Lung function tests showed a reduced vital capacity and reduced expired volume in the first second. An urgent transthoracic echocardiogram showed the presence of a dense soft tissue density infiltrate affecting primarily the left ventricular free wall (Figure 1 and Supplementary data online, Video S1), mitral annulus, and the mitral valve leaflets. In particular, it involved the lateral aspect of the posterior and anterior wall of the left ventricle, as well as the mitral annulus, and P1 and A1 scallops of mitral valve leaflets (Figure 2). Furthermore, the lateral aspect of the left atrium was infiltrated with a strand of tissue connecting the left atrial wall and mitral leaflets across the mitral valve orifice (Figures 1 and 2 and Supplementary data online, Video S2), causing reduced opening and functional mitral stenosis. Left ventricular diameters and systolic function appeared normal.

The patient was started on high-dose oral prednisolone, which led to rapid decrease of cough and dyspnoea. Intravenous heparin was started to avoid thrombus deposition on the infiltrate. Laboratory investigations from peripheral blood samples and bone marrow biopsy excluded all causes of secondary eosinophilia, whereas genetic testing excluded the presence of platelet-derived growth factor receptor alpha gene Fip1-like1 gene re-arrangement or cysteine-rich hydrophobic domain 2 deletion and the presence of any myeloproliferative disorder. Haematologic derangement improved before the patient was discharged from the hospital, and warfarin was initiated to avoid thrombus formation during the healing process. At 10 weeks after discharge, there was evidence of complete resolution of the LV infiltrate and disappearance of the infiltrate affecting both mitral leaflets, with improved excursion. The resolution of the infiltrate on the posterior mitral leaflet resulted in fibrosis with leaflet thickening and tethering and consequent mild mitral regurgitation.

Discussion
Eosinophilic infiltration of the heart was originally described in 1936 by Löffler in the post-mortem examination of two patients dying after two decades of afebrile leucocytosis and eosinophilia, progressive right-sided heart failure with hepatosplenomegaly, and ascites. Cardiac autopsy showed a layering of fibrosis that obliterated the ventricles but spared the valves. Eosinophilic heart disease is now a recognized manifestation of the hypereosinophilic syndrome, in which up to 50% of patients have evidence of cardiac involvement. Typical cardiac findings include endocardial fibrosis with extensive mural thrombus occupying the apices of both ventricles. Advanced forms include progressive myocardial damage, conduction system disease, and refractory heart failure. The present case highlights some atypical features of eosinophilic heart disease. First, eosinophilic heart disease is extremely rare...
in children. Second, the child described by us presented with infiltration limited to the lateral left atrial and ventricular walls, with preservation of the right ventricle and the apex of the left ventricle. Also, the patient had direct infiltration of the mitral valve, which caused functional valve stenosis rather than regurgitation as it is more commonly seen, probably due to inflammatory tissue infiltration.

Figure 1  Four-chamber view focusing on the left ventricle showing the eosinophilic infiltrate of the endocardium of the left ventricular lateral wall (A). The arrow shows the inflammatory tissue strand connecting the lateral atrial wall and the anterior mitral leaflet. AML indicates anterior mitral leaflet. Magnified image showing in detail the same findings of A (B). Asterisks indicate normal left ventricular myocardium. The arrows show the inflammatory tissue strand connecting the lateral atrial wall and the anterior mitral leaflet.

Figure 2  Short-axis view of the left ventricle at the level of the mitral valve (A) showing the eosinophilic infiltrate of the left lateral ventricular wall and the anterior and posterior mitral valve leaflets (scallops A1 and P1). Long-axis view of the left ventricle (B) showing the infiltration of the posterior mitral leaflet and the inflammatory tissue strand restricting anterior mitral valve leaflets. AML indicates anterior mitral leaflet; PML, posterior mitral leaflet.
strand detected in the acute phase, as opposed to the mitral regurgitation seen after fibrotic scarring in the late phase. Two-dimensional echocardiography is the primary method for the diagnosis of eosinophilic heart disease and should be performed in all patients with peripheral blood eosinophilia.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

Conflict of interest: none declared.

References