

# Diagnostic challenges in a long-term follow-up of hypereosinophilic restrictive cardiomyopathy

Długoterminowa obserwacja hipereozynofilowej kardiomiopatii restrykcyjnej — wyzwania diagnostyczne

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A 21-year-old woman was admitted for progressive dyspnoea, non-productive cough, and fever over the previous two weeks. Physical examination revealed fine pulmonary crackles, and bilateral pulmonary infiltrates were noted at chest X-ray (Fig. 1). Sinus tachycardia and ST-T abnormalities on electrocardiogram (ECG) were present (Fig. 2). Echocardiography revealed normal-sized, hyperkinetic left ventricle with a thrombus attached to the ventricular apex (Fig. 3). The mitral inflow had a restrictive pattern with increased filling pressures ( $E/e' = 15$ ). Laboratory data showed moderate anaemia and increased white cell count with  $3500/\text{mm}^3$  eosinophils. All the data supported the diagnosis of hypereosinophilic syndrome (HeS) with pulmonary and cardiac involvement. An extensive work-up excluded haematological disease (leukaemia, lymphoma), tumours, and vasculitis, and *Toxocara canis* infection was diagnosed based on ELISA-positive specific antibodies. Anticoagulant, corticosteroid, beta-blocker, and albendazole therapy was initiated, and clinical improvement was noticed with disappearance of pulmonary infiltrates and cured helminthic infection. However, the ventricular thrombosis and the restrictive pathophysiology persisted and hypereosinophilia was only partially remitted after one month of treatment ( $2000/\text{mm}^3$ ). Initially, HeS aetiology strongly suggested a *Toxocara canis* infection, despite the fact that it is a mild, self-limited disease, rarely affecting the heart. Based on the persistence of hypereosinophilia and the presence of severe restrictive cardiomyopathy, a diagnosis of idiopathic HeS with Loeffler endocarditis and incidental parasitic infection was established. A few months later, the patient developed acute femoral artery occlusion under anticoagulation therapy and was successfully treated by surgical thromboendarterectomy. The intensity of anticoagulation was increased thereafter. After eight years of follow-up, the patient had moderate heart failure symptoms with abnormal ECG and mild hypereosinophilia ( $700/\text{mm}^3$ ). She was on long-term corticosteroid, beta-blocker, and anticoagulant therapy without recurrence of cardioembolism. Echocardiography revealed severe endocardial fibrous thickening dividing the left ventricle in two pseudo-cavities with compensatory dilatation of the ventricular base (Fig. 4). There was persistent restrictive mitral inflow, left atrial dilatation, and moderate mitral regurgitation without right ventricular involvement. Idiopathic HeS is a rare disorder defined by persistent hypereosinophilia ( $> 1500/\text{mm}^3$ ) associated with organ damage. Restrictive cardiomyopathy with endomyocardial thrombosis and fibrosis occurs in about 20% of cases, having an unpredictable evolution. Echocardiography yields pathognomonic features showing apical ventricular thrombus adjacent to the normal contracting myocardium, restrictive mitral inflow pattern, and extensive areas of fibrosis involving the mitral valve in the late stages. This case is remarkable for many reasons. It has brought together two possible causes of hypereosinophilia. The simultaneous acute onset of idiopathic HeS with heart and lung involvement had different outcomes, with pulmonary remission and incessant cardiac disease progression. There was single ventricle damage. The major role of anticoagulation in the prevention of thromboembolism and the partial efficacy of medication were highlighted.



Figure 1. Chest X-ray — bilateral pulmonary infiltrates

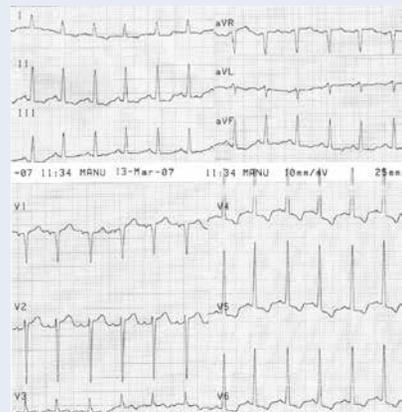


Figure 2. Electrocardiogram — sinus tachycardia and ST-T abnormalities



Figure 3. Echocardiography — normal-sized, hyperkinetic left ventricle with a thrombus attached to the ventricular apex



Figure 4. Echocardiography — severe endocardial fibrous thickening dividing the left ventricle in two pseudo-cavities with compensatory dilatation of the ventricular base

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**Conflict of interest:** none declared

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