Unusual cardiac manifestation of hypereosinophilic syndrome

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Hypereosinophilic syndrome was diagnosed in a 14 year old girl who presented with recurrent syncpe. An ambulatory electrocardiogram revealed intermittent type 2 second degree left atrioventricular (AV) block. Focal thinning of the interventricular septum was noted on echocardiography, contrary to the commoner finding of regional ventricular wall thickening among patients with hypereosinophilic syndrome. High grade AV block as a manifestation of hypereosinophilic syndrome is rare and focal thinning of the left ventricle as a manifestation of the syndrome has not been described earlier.

A 14 year old girl presented with history of recurrent syncopal attacks of one years’ duration. She was on antiepileptic drugs at presentation based on an electroencephalogram showing epileptiform activity in the parieto-temporal region noted during evaluation for syncpe. Further evaluation for persistent syncopal episodes, despite antiepileptic medications, had shown intermittent second degree atrioventricular (AV) block on the electrocardiogram (ECG), focal thinning of the interventricular septum, and wall motion abnormality on echocardiography. She was referred to our institute for permanent pacemaker implantation.

She had no history suggestive of collagen vascular disease, bronchial asthma, atopy, vasculitis, or worm infestation. Physical examination revealed a regular cardiac rhythm at a rate of 72 beats/min, blood pressure of 90/60 mm Hg, and an unremarkable cardiac examination. Blood tests showed a raised leucocyte count (18 × 10^9/l) with an absolute eosinophil count of 8.3 × 10^9/l, and platelet count of 141 × 10^9/l. Serum IgE was normal (85 IU/ml). Stool examination for ova and cysts was repeatedly negative. Peripheral blood and bone marrow smear showed marked eosinophilia and there were no abnormal cells. Peripheral smear for filarial parasite was negative. Antinuclear antibody, antineutrophil cytoplasmic antibody, and hepatitis B surface antigen were negative. An ECG revealed sinus rhythm, right bundle branch block, and inverted T waves in the anterior leads. Ambulatory ECG monitoring revealed intermittent type 2 second degree AV blocks (fig 1). The basal portion of the interventricular septum was grossly thinned out (fig 2) with mildly dilated left ventricle (left ventricular internal dimension diastole 51 mm, left ventricular internal dimension systole 39 mm) and impaired global left ventricular contractility (left ventricular ejection fraction 48%) on echocardiography. There was no AV valve regurgitation. There was no cardiomegaly and no pulmonary infiltrates on the frontal chest radiograph. After excluding all possible causes of eosinophilia including drugs, a diagnosis of idiopathic hypereosinophilic syndrome with predominant cardiac involvement was established. She was started on oral corticosteroids and advised permanent pacemaker implantation.

DISCUSSION
The diagnosis of hypereosinophilic syndrome in this patient was based on a total eosinophil count greater than 1.5 × 10^9/l, cardiac involvement, and lack of evidence for any other known cause of eosinophilia.¹ ² Syncope is an uncommon manifestation of cardiac involvement in hypereosinophilic syndrome and has not been described in detail.¹ ² Higher degree of AV block in hypereosinophilic syndrome has not been reported in the literature to the best of our knowledge.

There are several unique features in our patient. This patient had focal interventricular septal thinning on echocardiography, contrary to the commoner finding of regional ventricular wall thickening in patients with hypereosinophilic syndrome.² However, there are few reports of advanced dilated cardiomyopathy in the literature.⁵ ⁶ ⁷ Hypereosinophilic syndrome presenting as symptomatic AV block is a new occurrence. It is reasonable to presume that the involvement of the basal portion of the interventricular septum is responsible for the AV block in this patient.

The effect of treatment with corticosteroids and cytotoxic agents on the cardiac manifestations of hypereosinophilic syndrome is variable, acute involvement responding better to such treatment.² ³ ⁷ However the response of conduction abnormalities to medical treatment is not clear, more so in this patient. The echocardiographic finding of basal septal thinning would probably indicate the irreversibility of the AV
block. Further follow up of this patient is needed to understand the behaviour of conduction system disturbances in hypereosinophilic syndrome.

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Figure 2  Two dimensional echocardiography, parasternal long axis view showing thinned out septum.

Learning points

- Cardiac involvement is common among patients with hypereosinophilic syndrome.
- Focal thinning of the interventricular septum can occur as a manifestation in hypereosinophilic syndrome.
- Cardiac conduction abnormalities as a manifestation of hypereosinophilic syndrome, although uncommon, should be considered in the evaluation of unexplained syncope.

REFERENCES
