

CASE REPORT

HYPEREOISNOPHILIC SYNDROME: A CASE REPORT

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ABSTRACT: We report a 28years old male presenting with syncope. A thorough clinical evaluation directed us towards cardiac syncope. Doppler echocardiographic study was used as a main modality of diagnosis and cardiac catheterization confirmed the diagnosis of restrictive cardiomyopathy. We express the contribution of clinical findings and appropriate diagnostic measures in approaching a rare case of systemic eosinophilic syndrome, presenting as restrictive cardiomyopathy.

KEYWORDS: Hypereoisophilia, cardiomyopathy, eosinophilic gastritis.

INTRODUCTION: The hypereosinophilic syndrome involves several organ systems beyond the heart, including the lungs, brain and bone marrow. Both chambers of the heart are involved and show endocardial thickening of the inflow regions and ventricular apices.^[1] On histological examination, there are variable degrees of eosinophilic myocarditis of the myocardium and subendocardium, thrombosis and inflammation of small intraluminal coronary vessels, mural thrombosis containing eosinophils and endocardial thickening of several millimeters.

CASE HISTORY: A 28years old male presented to the out-patient department with breathlessness on exertion, fatigability, pedal edema and epigastric pain since 2years, aggravated since last 2 months. O/E- Pulse rate 88/min, irregularly irregular low volume. Blood pressure 100/70 mm Hg, respiratory rate 18/min, bilateral pedal edema present, jugular venous pressure (JVP) raised (10cm H₂O) with prominent "y" descent. CVS examination revealed apical impulse well defined in left fifth intercostal space 1cm lateral to mid clavicular line, variable S₁, loud P₂, systolic murmurs in mitral and tricuspid areas.

Respiratory system revealed clinically no abnormalities. Per abdomen showed congestive hepatomegaly with ascites. On investigations, ECG showed normal voltage complexes with atrial fibrillation, right axis deviation with ST-T changes (reverse tick sign) (Fig. 1). Chest x ray showed enlarged cardiac silhouette with pulmonary congestion.

Echocardiography showed marked biatrial enlargement, (fig. 2) mildly hypertrophied but normal sized ventricles, moderate MR, severe TR, mild PAH (SPAP 40 mmHg), fair biventricular function, no intracavitary clots, with minimal pericardial effusion. Echocardiography measurements LA 64mms, IVS 10 mms, LVID(d) 45mms, LVID(s) 35mms, PWD 8 mms, EF 50% (fig. 3).

Cardiac catheterization revealed grade III MR, severe TR severe PAH, mild RV systolic dysfunction, no apical obliteration, no endocardial calcification, fair LV systolic dysfunction with normal coronaries. On routine investigation absolute eosinophilic count is 3000per/cumm. Upper gastro intestinal endoscopy revealed multiple ulcers and biopsy has been done, which is showing eosinophilic gastritis (fig. 4).

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DISCUSSION: This patient of heart failure presented with symptoms of systemic and pulmonary congestion was evaluated and significance was given to the raised JVP with prominent “y” descent suggestive of restrictive heart disease. Clinical signs of mitral regurgitation (MR), tricuspid regurgitation (TR) and pulmonary arterial hypertension (PAH) with atrial fibrillation (AF) were significant.

ECG showed normal QRS voltage with AF, right axis deviation and ST-T changes (suggestive of digoxin toxicity). Presence of AF with negative “y” descent made us to evaluate regarding restrictive right heart diseases.^[2]

Echocardiography showed biatrial dilatation, restrictive Doppler flows, dilated inferior vena cava, preserved systolic function and atrio ventricular regurgitations which were characteristic of RCM. Echocardiography also helped us in excluding endomyocardial involvement as seen in endomyocardial fibrosis by the absence of obliteration of ventricular cavities and right ventricular outflow tract dilatation.^[3]

This was confirmed by cardiac catheterization which revealed markedly elevated ventricular filling pressure with characteristic restrictive haemodynamic pattern and no obliteration of cavities.

Endomyocardial biopsy was not performed as studies have shown its limitation reserved in excluding infiltrative disorders and being a blind procedure^[4] Löffler endocarditis is more aggressive and rapidly progresses, affects mainly males and is associated with hypereosinophilia, thromboemboli, systemic arteritis; endomyocardial fibrosis occurs in a younger distribution, affects young children and is only variably associated with eosinophilia.^[5]

In temperate climates, endomyocardial disease is closely associated with significant hypereosinophilia, which can have several different causes. Hypereosinophilia associated with Löffler endocarditis usually is characterized by eosinophil counts exceeding 1500/cumm for at least 6 months. Most patients with this degree of hypereosinophilia will have cardiac involvement.

The eosinophilia may be secondary to leukemia, reactive disorders such as parasite infection, allergies, granulomatous syndromes, hypersensitivity, or neoplastic disorders. The mechanism by which eosinophils participate in the development of cardiac disease remains incompletely understood. These cells have the capacity to directly infiltrate tissues or to release factors that may exert toxicity.

The observation that patients with Löffler endocarditis have degranulated eosinophils in their peripheral blood supports the idea that these granules contain cardiotoxic substances, capable of causing the necrotic phase of endomyocardial disease, which leads to the thrombotic and fibrotic phases once the eosinophilia resolves.^[6]

Here we are reporting a case of hypereosinophilic syndrome presenting as restrictive cardiomyopathy and eosinophilic gastritis, a high suspicion should be kept in mind for the above diagnosis when patient presents with hypereosinophilia. Eosinophilic gastritis is a rare and heterogeneous condition characterized by patchy or diffuse eosinophilic infiltration of gastrointestinal tissue; presentation may vary depending on location as well as depth and extent of bowel wall involvement. It usually runs a chronic relapsing course.^[7]

Antiplatelets, digoxin, warfarin, Corticosteroids mainstay of treatment.

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Fig 1: ECG atrial fibrillation

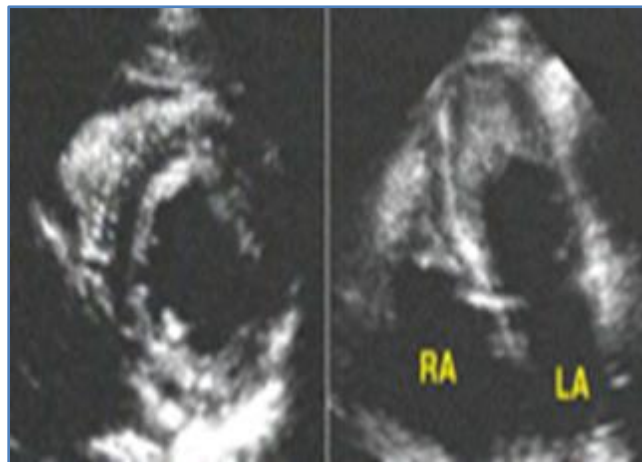


Fig 2 : 2d echo showing bi atrial enlargement with TR

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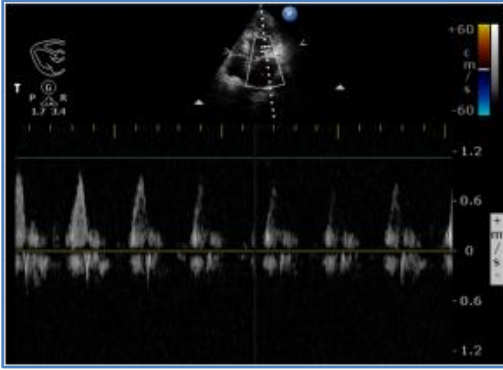


Fig 3 : 2d echo showing bi atrial enlargement with TR



Fig 4: Upper g.i endoscopy showing nonhealing ulcers

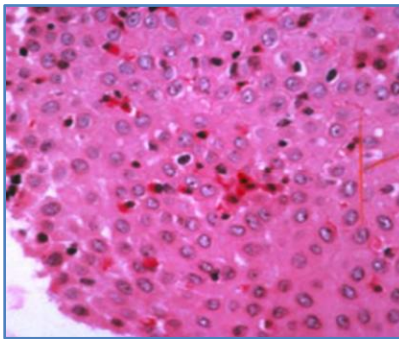


Fig 5: biopsy showing eosinophilic gastritis

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