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Hypereosinophilic Syndrome with Loeffler Endocarditis, A Rare Case of Heart Failure in A Child

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Abstract: *An eight-yr-old girl presented with history of progressive dyspnoea and cough for 12 days with fatigue and rapid worsening of symptoms. There was features of cardiogenic shock ,congestive cardiac failure and mild cardiomegaly. Investigations revealed marked leucocytosis with 63% eosinophils. Cardiac MRI was suggestive of endomyocardial fibrosis with mural thrombi in the left ventricle. Extensive work up was consistent with the diagnosis of hypereosinophilic syndrome (HES) with Loeffler endocarditis , a rare differential of heart failure in children.*

Keywords: *Cardiogenic shock, Heart failure, Hypereosinophilic syndrome, Loeffler endocarditis .*

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I. Introduction

Hypereosinophilic syndrome (HES) is characterized by any form of hyper eosinophilia (HE) associated with organ damage [1]. We describe here a girl with HES with severe cardiac involvement, so-called Loeffler endocarditis ,who presented to us with congestive heart failure and shock .

II. Case Report

An 8-year-old-girl presented with complaints of progressive dyspnea and cough for 12 days before admission. There was no history of fever, sore throat , joint pains, skin manifestations, hematuria, diarrhea, jaundice, bleeding manifestations, allergic manifestations, worm infestations, contact with tuberculosis or any drug intake. At admission, the girl appeared sick looking, pale and tachypneic having a pulse rate of 144/min, respiratory rate of 48/min and BP-78/40 mm Hg , pedal edema and raised JVP, but no lymphadenopathy, bony tenderness , skin lesions, jaundice, cyanosis or clubbing. Examination of the cardiovascular system revealed mild cardiomegaly with gallop rhythm and soft holosystolic murmur at apical area without any conduction. There was bilateral basal crepitations and soft, tender hepatomegaly . Rest of the systemic examination was within normal limit.Investigations revealed Hb- 10.9 g/dl, total leucocyte count (TLC)-64,380/mm^3, polymorphs- 27%, lymphocytes- 8%, monocytes- 2%, eosinophils- 63% with absolute eosinophil count 40,716/mm^3 , platelet count - 2.86 lakh/mm^3, ESR - 30mm , CRP < 0.3 mg/dl, ASO - 204 IU/ml , troponin T-370 ng/L (N-<3 ng/L), creatine kinase- 138 U/L and creatine kinase-M B mass 2.5 ng/ml. Arterial blood gas analysis was within normal limit. Serum C3 was 149 mg/dl. Immunoglobulin E was raised (532 IU/ml). No OPC found on stool examination. Other tests including, liver function tests , kidney function tests, blood cultures, urine examinations, connective tissue and vasculitis screening tests were within normal limit. X-ray chest revealed mild cardiomegaly. Electrocardiographic monitoring showed sinus tachycardia. Echocardiography revealed hyper-echoic shadow attached to the posterior and lateral walls of left ventricle suggestive of cellular infiltration, prolapsed posterior mitral leaflet, severe mitral incontinence and chunk of pericardial effusion. Ejection fraction was 39.7% Cardiac magnetic resonance imaging (CMRI) was suggestive of endomyocardial fibrosis with mural thrombus in left ventricle.

It showed a sessile lesion (about 1.3 cm x 2.3 cm) projected with irregular margins from the posterior wall of left ventricle and also was extended upto the posterior mitral leaflet. There was poor enhancement in the lesion in first pass images , and delayed contrast enhancement was noted along the endocardial surface. Left atrial chamber was dilated and left ventricle was also mildly enlarged . Bone marrow biopsy revealed hypercellular marrow with eosinophilic precursors with 2% blasts.

Pharmacologic and supportive therapy were initiated promptly for heart failure and shock . Vasoactive drugs needed were milrinone (as an inodilator) and low dose epinephrine . In view of clinically evident heart failure , echocardiographic and cardiac MRI image in the background of severe peripheral hypereosinophilia , the diagnosis of Loeffler endocarditis was made. The patient was put on IV methylprednisolone (30mg/kg/day) for three days followed by oral prednisolone (2 mg/kg/day), and also anticoagulants , beta-blockers , diuretics and antiplatelet agents. There was a marked decline in her eosinophil count (TLC 26,900/cmm with 41% eosinophil) after 11 days of initiation of therapy . But she had persistent tachycardia and was dependent on oxygen and vasoactive drugs . Repeat ASO titre was 198 Iu/ml. Due to poor general condition percutaneous endomyocardial biopsy and surgery was not attempted. Further investigations could not be done as the patient succumbed after 26 days of admission due to refractory shock and hypoxemia at the end .

III. Discussion

The differential diagnosis of cardiac disease with peripheral eosinophilia include HES, Churg-Strauss syndrome, giant cell myocarditis, parasitic infections, and malignancy [2]. Hypereosinophilic syndrome (HES) and hypereosinophilia (HE) , first described by Chusid et al [3] has recenty been revised [1] . HE is now defined as any form of persistent and marked eosinophilia , documented on at least two occasions with a minimum time interval of four weeks (except when immediate therapy is required because of hypereosinophilia mediated organ dysfunction) , and HES, as any form of HE associated organ damage, in the absence of an alternative explanation for the damage [1]. Organ damage is secondary to eosinophils' degranulation toxicity and common target organs are cardiac system, nervous system, skin, lung and gastrointestinal tract [4] . The index case was probably suffering from acute necrosis of myocardium due to eosinophilic infiltration followed by formation of mural thrombus and endomyocardial fibrosis as evident from CMRI. Troponin T level was elevated in our case because eosinophilic infiltration may cause necrosis of cardiac myocytes leading to myocarditis and congestive cardiac failure. Corticosteroids are currently recommended as first-line therapy and our patient responded well. Surgical removal of the big mural thrombus could have saved the child from intractable cardiac failure.

HES is considered very rare in children. One study estimated the crude incidence of HES in adult population is to be about 0.035 / 100,000 person/year and median age at diagnosis is 52.5 years and male to female ratio is 1.47[5]. Ventricular thrombosis in HES is reported in adults [6]. Heart failure in older children beyond 2 years are usually due to the causes like rheumatic heart diseases, myocarditis, cardiomyopathies and congential heart diseases with complications .What makes this case unique as Loefller endocarditis is a rare cause of heart failure and persistent shock in pediatric age group .

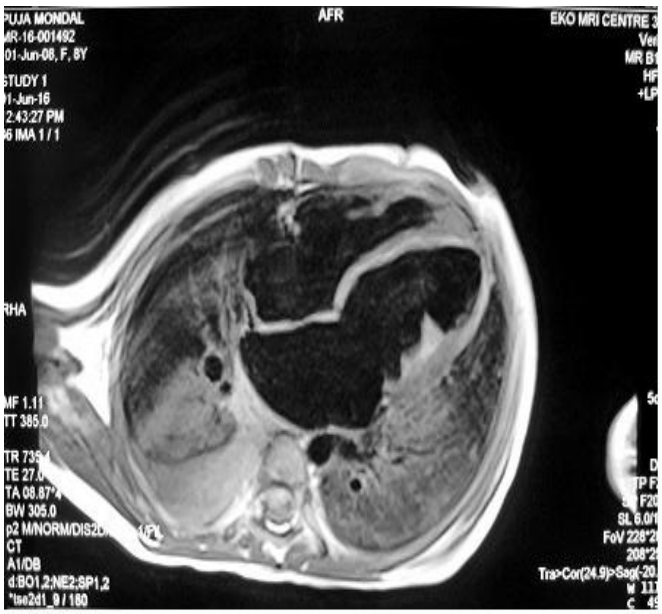

Figure 1

Cardiac MRI suggestive of a sessile lesion projecting with irregular margins from the posterior wall of left ventricle.

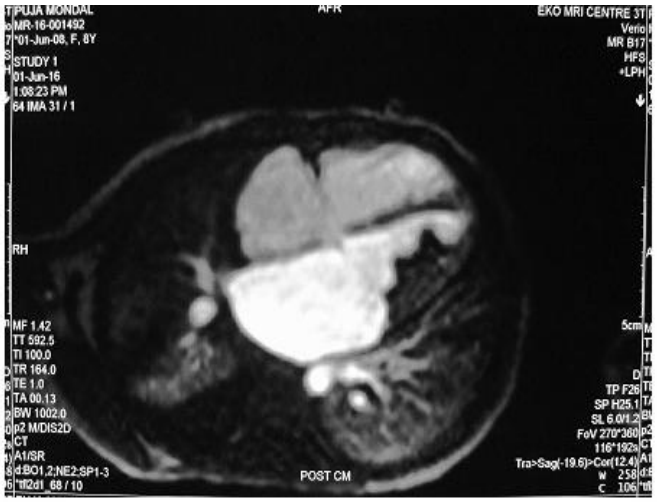

Figure 2

MRI of the same lesion shows poor enhancement.

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