Idiopathic Hypereosinophilic Syndrome Manifesting As Pulmonary Oedema

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Abstract

Idiopathic hypereosinophilic syndrome is a progressive and fatal disease if not treated effectively. We report this case since hypereosinophilia is an uncommon cause of pulmonary oedema.

INTRODUCTION

The idiopathic hypereosinophilic syndrome is characterized by persistent eosinophilia with more than 1500 eosinophils for at least 6 months or until death with evidence of organ involvement usually progressive and fatal in the absence of effective medical management. Cardiac involvement occurs in more than 75% patients suffering from this disorder.

CASE REPORT

A 17 year old male student, was admitted with complaints of breathlessness, oedema feet and puffiness of face since 25 days and dry cough since 5 days. There was no history of any cardiac or renal disease in the past. There was no history of hematuria. There was no significant past history.

On examination, pulse was 126/min, blood pressure was 140/120mm Hg. The patient was pale, had oedema feet, JVP was elevated. There was no cyanosis, clubbing, icterus or lymphadenopathy. There was tender hepatomegaly. There were bilateral crepitations all over the chest. S3 gallop was present. There was no murmur. The patient was conscious and oriented. There was no focal neurological deficit.

The blood investigations revealed Hb-13gm%. WBC count was 55000/cmm with a differential count of P-11% L15%E74%. ESR was 68 at the end of one hour. The peripheral blood smear did not show any parasites. Stool examination did not show any ova or parasites. Skiagram of chest showed pulmonary oedema with cardiomegaly (Fig. 1). ECG showed sinus tachycardia. Renal function tests were normal. Liver function tests showed mild elevation of liver enzymes. USG abdomen showed hepatomegaly with bilateral pleural effusion. The kidneys were normal. 2D echo showed dilated left ventricle with generalised hypokinesia with ejection fraction of 30%. Bone marrow examination showed normocellular marrow, myeloid:erythroid ratio was 4:1 with 80% eosinophils in various stages of development. Erythropoiesis was normal. Renal Doppler and urinary VMA was done which were normal and causes of secondary hypertension were ruled out. The patient was treated with oral amlodipin 5 mg once a day (for 1 month), intravenous frusemide 40mg twice daily (for 3 days) and oral diethylcarbamazine 100mg twice daily (for 21 days). The patient had improved clinically, dyspnoea had decreased, but was present. Blood counts were repeated after completion of 21
days of diethyl carbaamazine. Total count was 15600/cmm, P-23%, L-33%, E-44%. Thus the patient was diagnosed to have idiopathic hypereosinophilic syndrome.

In view of the persistently elevated eosinophil count the patient was started on oral prednisolone at the dose of 1mg/kg/day for 4 weeks. The skiagram of chest after completion of steroids showed decrease in the heart size (Fig. 2). The steroids were gradually tapered and stopped. Amlodipin was also stopped as the blood pressure was normal. 2D echo after treatment showed normal wall motion, normal size of all chambers and EF was 55%.

**DISCUSSION**

In the absence of an identifiable cause of moderate to severe eosinophilia and in the presence of end organ involvement the diagnosis of idiopathic hypereosinophilic syndrome is to be considered.

It occurs more commonly in males (M:F=9:1) in the age group of 20 to 50 years. The WBC count may range from 10000 to 50000/cmm with eosinophil count of 30% to 70%.

The exact aetiopathogenesis of this illness is unknown. The tissue damage is caused by major basic and cationic proteins derived from cytotoxic eosinophils. The eosinophilic infiltration occurs without any evidence of an inflammatory or allergic agent and there is no vasculitis or granuloma formation. Infiltration of mature eosinophils is seen in many organs. Depending on the organs involved, there are three types:

1. Predominant pulmonary involvement in the form of tropical eosinophilia, pneumonias etc.
2. Predominant cardiac or CNS involvement. The cardiac involvement may be in the form of endomyocardial fibrosis or restrictive cardiomyopathy. The central nervous system involvement can be local deposits causing focal abnormalities or diffuse infiltration causing impairment of brain function.
3. Eosinophilic leukaemia.

Biopsies of the bone marrow and involved organs may be useful to confirm the eosinophilic infiltration. Asymptomatic patients with persistent benign eosinophilia usually do not need therapy and spontaneous resolution generally occurs within several years. However, such patients should have periodic clinical and 2D echo follow-up to detect eosinophil-mediated cardiac damage, which can occur insidiously at any time and which may not be correlated with the severity of eosinophilia. Although no therapy is indicated, it is often helpful to determine whether eosinophilia will resolve with a short course of prednisolone (1mg/kg) for 3-5 days. Steroid resistant cases may be given a trial of hydroxyurea or Interferon.

**REFERENCES**