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CASE STUDY

HYPEREOSINOPHILIC HYPER IgE SYNDROME – A RARE CASE REPORT

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ABSTRACT

Hyper immunoglobulin (Ig) E syndrome is a rare complex immunoregulatory disorder characterized by hyperglobulinemia, recurrent bacterial infections and chronic eczematous dermatitis. The onset of this syndrome may occur at any time from early childhood onwards, and may include eosinophilia and osteoarticular and dental abnormalities. The mortality in untreated patients three years after diagnosis can be high as 75%. Cardiac involvement is the most common cause of increased morbidity and mortality. Early diagnosis of this condition and starting appropriate therapy is of paramount importance in establishing a diagnosis and decreasing the morbidity and mortality associated with this condition.

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INTRODUCTION

In this report, we describe the case of a 9 year old boy who had a combination of hypereosinophilia, marked elevated IgE levels in order to illustrate the difficulty of establishing the diagnosis and initiating the treatment promptly.

Hyper immunoglobulin (Ig) E syndrome is a rare complex immunoregulatory disorder characterized by hyperglobulinemia, recurrent bacterial infections and chronic eczematous dermatitis. The onset of this syndrome may occur at any time from early childhood onwards, and may include eosinophilia and osteoarticular and dental abnormalities. The mortality in untreated patients three years after diagnosis can be high as 75%. Cardiac involvement is the most common cause of increased morbidity and mortality. Early diagnosis of this condition and starting appropriate therapy is of paramount importance in establishing a diagnosis and decreasing the morbidity and mortality associated with this condition.

In this report, we describe the case of a 9 year old boy who had a combination of hypereosinophilia, marked elevated IgE levels in order to illustrate the difficulty of establishing the diagnosis and initiating the treatment promptly.

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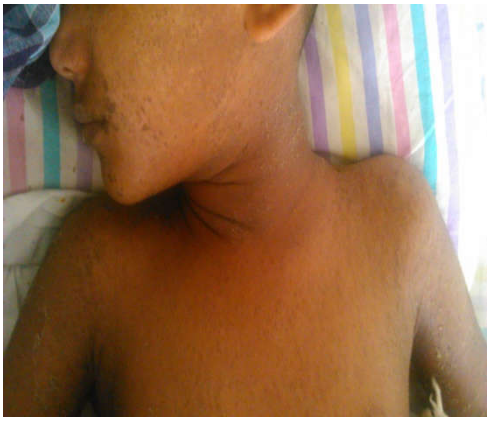
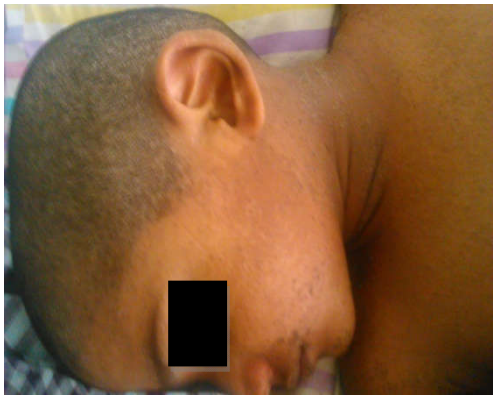
Case report

A 9 year old previously healthy boy was brought with history of fever and malaise for the past three months duration and maculopapular rash with scaling for the past two weeks with history of itching. The rashes started initially in the thigh and spread all over the body. He had history of jaundice and decreased urine output for past 1 week duration. The patient complained of throat pain. There was erythema and desquamation over the forehead, perioral area, trunk, upper limb, thigh upto the leg. On examination the boy was well built with no pallor. There was icterus and the patient had bilateral cervical and inguinal lymphadenopathy of size 1-1.5 cm which were firm, non matted, non tender. He had facial and periorbital edema. The maculopapularrashes were seen predominantly over the forehead. The oral, buccal mucosa and ventral surface of the tongue showed multiple candy white patches. There was congestion of pharynx. His cardio vascular system was normal with blood pressure of 100/60 mm Hg. The respiratory rate was 18/ minutes. His investigations are as follows hemoglobin 14.1gm%, total count 17,300 cells /cummwith eosinophils 35%, Absolute eosinophil count (AEC) 3000/$mm^{3.}$ His ESR was 9mm/1 hour. IgE level by chemiluminiscence was 606.7 IU/mL (Normal - < 80 IU/mL). Peripheral blood smear showed Normocytic normochromic blood picture with eosinophilic leukocytosis with toxic features.





His liver function test showed serum bilirubin 3.2 mg/dl with direct fraction 2.8 mg/dl, SGOT 80 IU/L (5-45 IU/L), SGPT 70 IU/L (5-45 IU/L), serum Alkaline phosphatase 1645 IU/L (130-525 IU/L). Ultrasonogram of abdomen showed hepatosplenomegaly and bilateral splenomegaly with no free fluid. Echo cardiogram showed no abnormalities with normalvalvular function. His total serum proteins were 7.6 gm/dl, albumin 2.8 gm/dl, increased globulin 4.8 gm/sl, blood urea 75 mg/dl, serum creatinine 2 mg/dl with normal serum electrolytes. Urine showed pus cells 0-1/hpf with no cellular/ RBC casts, 2+proteinuria. Stool examination did not reveal any ova/cysts, before and after deworming.

Throat swab showed growth of normal upper respiratory flora. Since the patient had elevated AEC and elevated IgE levels, it was diagnosed as a case of hypereosinophilic, hyper IgE syndrome and the patient was started on IVmethyl prednisolone (30mg/Kg/day) for 5 days followed by oral prednisolone 1mg/Kg/day. Within 10 days of initiation of corticosteroid therapy his liver function improved with disappearance of skin rashes with normal hepatic and renal functions. The patient was discharged with oral prednisone, which was tapered and stopped over a period of 6 months. He was reviewed after 6 weeks. On review after 6 weeks his AEC was 2200 mm^3, 3rd month 1450 mm^3, 6th month 880 mm^3. His repeat renal and liver function test continues to be normal. He is symptom free till date.

DISCUSSION

Hypereosinophilic, hyper IgE syndrome is a rare disease in children and its exact incidence is uncertain. It is a group of heterogenous disorders many of which remain ill defined and are characterized by persistent eosiniophilia (defined as > 1500/mL for 6 months) and organ involvement/dysfunction (in which other clinical entities have been excluded). It can be asymptomatic or may present as life threatening diseases. The pathophysiology of this disease is eosinophil mediated with tissue damage causing organ dysfunction. Multiple organ systems are usually affected namely cardiac, central nervous system, hepatic, renal, hematologic and skin. Some have underlying malignancy while others progress to eosinophilic leukemia. In our case we have involvement of skin in the form of maculopapular rashes, hepatic in the form of hepatospleenomegaly and jaundice and renal involvement in the form of proteinuria and elevated serum creatinine and hematological picture showing eosinophilic leukocytosis, elevated IgE and raised absolute eosinophilic count. There was no cardiac, pulmonary or neurological involvement. Since the patient had elevated IgE, and raised AEC persisting for more than 6 months with favorable response to steroids, a case of hypereosinophilic, hyper IgE syndrome was diagnosed.

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