
Case Reports

Hyperimmunoglobulin E Syndrome

**Manju Salaria
Surjit Singh
Lata Kumar**

Hyperimmunoglobulin E syndrome (HIE) is a rare immunodeficiency disorder characterized by atopic like dermatitis, recurrent systemic and cutaneous pyogenic infections, markedly increased serum levels of IgE and peripheral eosinophilia. This syndrome has only rarely been reported in the Indian literature (1).

Case Reports

Case 1: A ten-year-old male child presented to us with history of recurrent skin infections since the age of two months and recurrent respiratory infections since the age of seven years. He was fully vaccinated. On examination his weight was 18 Kg and height was 127 cm. He had coarse facial features, evidence of infected eczema and multiple healed scars over legs and face. The child was given a course of penicillin on which he showed gradual improvement. Four months later, he presented with an erythematous swelling in the left

inguinal region but with little rise of local temperature. Investigations revealed normal hemoglobin, total leucocyte count of 9400/cu mm with absolute eosinophil count 470/cu mm which later increased to 846/cu mm. He had normal serum electrophoresis, normal nitroblue tetrazolium dye reduction and negative HIV serology. His IgE levels were 4790 IU/ml (N=0.1-90 IU/ml). The abscess was drained and pus grew *Staphylococcus aureus* on culture. He was treated with oral cloxacillin for four weeks. Oral cimetidine (10 mg/kg thrice a day) and low dose cotrimoxazole prophylaxis (3 mg/kg/day of trimethoprim + 15 mg/kg/day of sulfamethoxazole) were initiated. He again developed skin boils and inguinal lymphadenopathy when therapy was discontinued due to financial constraints. This infection responded to a course of oral cloxacillin.

Case 2: A nine year old male child presented with history of fever and swelling over the parotid region for 15 days. His growth and development was normal and there was no history of recurrent bacterial infections in early childhood. He was fully vaccinated. On examination he had coarse facial features, rough skin and a swelling over parotid region. He had tender enlarged right cervical and left axillary lymph nodes with little rise of local temperature. Investigations revealed a normal hemoglobin level, total leucocyte count of 14,000/cu mm with absolute eosinophil count being 840/cu mm. He had normal serum electrophoresis and normal nitroblue tetrazolium dye reduction. His serum IgE levels were 2072 IU/ml. The pus from enlarged left axillary lymph node grew *Staphylococcus aureus*. He was treated with oral cloxacillin

From the Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Reprint requests: Dr. Lata Kumar, Professor and Head, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

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for 2 weeks and the recovery was uneventful. Repeat IgE estimation done after 3 weeks showed a titer of 3564 IU/ml). He was started on oral cimetidine and was asymptomatic at six month follow up.

Discussion

In 1966, Davis *et al* described a clinical syndrome of recurrent staphylococcal abscesses, chronic sinusitis and infected eczematous lesions in two girls (2). Buckley *et al.*(3) later reported the syndrome in a male child with raised IgE levels and recurrent pyogenic infections suggesting that this entity is not restricted to girls. On literature review, it was concluded that the syndrome entails a disorder of recurrent bacterial infections of skin and sinopulmonary tract starting in childhood in the presence of serum IgE levels at least 10 times normal (>2000 IU)(4). This disease usually appears before the age of 2 years; however, the age at onset of first infection has at times been reported to be as high as 17 years(4). One of our cases had no major skin infection till the age of 9 years.

These patients are most commonly infected with *Staphylococcus aureus*, *Haemophilus influenzae*, *Streptococcus pneumoniae* and Group A beta hemolytic *streptococcus*^). Gram negative and fungal infections are said to be less common. Predominant sites of infections have been found to be skin and lungs, though ears, eyes, oral mucosa, sinuses, joints, blood and viscera can be involved (5). Similar to our cases, staphylococcal abscesses associated with hardly any features of local inflammation have been reported (2). The precise reason for this phenomenon is not clearly understood, though defective neutrophil chemotaxis (which is often a feature of HIE syndrome) may partly be responsible. Although most of these infections respond to antibiotics, recovery may be slow (1).

Dermatitis is present in more than 80% of patients and usually begins at 2 months to 2 years of age (5). Peripheral eosinophilia, along with elevated serum IgE levels, is the most common laboratory finding in HIE syndrome (6).

The exact defect leading to repeated injections remains to be defined. Elevated levels of IgE *per se* have not been proven to be responsible for the immunodeficiency. No T cell abnormality has been described. Defective polymorphonuclear chemotaxis has been hypothesized as the basis for patient's susceptibility to infections (7), but these abnormalities are highly variable and inconsistent. Polymorphonuclear phagocytosis, metabolism, and killing have been found to be normal (5).

As the etiology of this syndrome is uncertain, no definitive therapy is known. Antimicrobial agents form the mainstay of therapy. Improvement in neutrophil functions has been noticed with levamisole and ascorbic acid (6,8-10). Treatment with cimetidine has been shown to improve clinical parameters as well as neutrophil chemotaxis (11). Isotretinoin has also been reported to be effective in HIE syndrome. It probably causes decrease in sites for colonisation by bacteria by reduction of both sebum excretion and sebaceous gland mass(12). Two other forms of therapy have recently been reported-intravenous immunoglobulin (IVIG) and plasmapheresis (6). IVIG probably acts by providing circulating antibodies to bacterial antigens, which are deficient in these individuals (6).

The role of prophylactic antibiotics in HIE syndrome is difficult to evaluate as patients may have several months free of infections and then have several infections within a few months(4). Many patients can reach adulthood indicating that this defect is compatible with a prolonged survival (5).

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Carbamazepine Induced Pseudo-Lymphoma Syndrome

H. Paramesh
D.A. Satish
Salim A. Khatib
Mukta Jain M.

Pseudolymphoma syndrome, as a hypersensitive reaction to anticonvulsant drugs especially phenytoin, carbamazepine, tridone and phenobarbitone is well known(1-4). Its pathogenesis is uncertain. It consists of a triad of fever, generalized rash and lymphadenopathy. Varying degrees of

malaise, hepatosplenomegaly, abnormal liver functions, arthralgia, eosinophilia and blood dyscrasias have also been reported⁵. We present a case of carbamazepine induced pseudolymphoma syndrome.

From the Departments of Pediatrics and Dermatology, Lakeside Medical Center and Hospital, Bangalore.

Reprint requests: Dr. H. Paramesh, Director, Pediatrician-in-Chief, Lakeside Medical Center and Hospital, 33/4 Meanee Avenue, Bangalore 560 042.

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