LETTER TO EDITOR (VIEWER'S CHOICE)

HYPER IGE SYNDROME PRESENTED AS EXTENSIVE CELLULITIS AND SEVERE URTICARIA

Mukesh Vir Singh, K M Shukla

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An 8-year-old boy presented with history of pain as well as redness of skin for 4 days. He developed high grade fever on the day of admission. Clinical examination revealed multiple abscesses, urticarial rashes, multiple old scar marks and hyperpyrexia. He had recurrent abscesses 2.5 years back for which patient was treated by local doctor with some oral medication and surgical drainage of abscesses. Laboratory evaluation revealed moderate anemia (hemoglobin 8.6 g/L). His total leukocyte count (8.6 × 103 cells/dL), platelet count (290 × 109/L), differential count (P58,L40,E02,), serum creatinine (0.77 mg/dL), blood urea (39 mg/dL), serum bilirubin (0.34 mg/dL), ASO titer (<150 i.u./L) and chest radiograph were within normal limits. Urine routine microscopy and culture did not show any abnormality. The ALT (55 i.u./L) and AST (50 i.u./L) were mildly raised. Serum IgE levels were raised (3000 i.u./mL) suggestive of Hyper IgE. Patient was treated with vancomycin and gentamicin combination and surgical drainage of abscess. Intravenous gamma-globulin was advised but patient's family could not afford. He was discharged on long term cotrimoxazole.

Hyper-IgE syndrome (HIES) was first described by Davis et al in 1966 in two girls with red hair, chronic dermatitis, and recurrent staphylococcal abscesses and pneumonias (1). They named the disease after the biblical character Job, whose body was covered with boils by Satan. In 1972, Buckley et al described two boys with similar symptoms as well as coarse facies, eosinophilia, and elevated serum IgE levels. These two syndromes are thought to be the same and are under the broad category of HIES (2). Abnormal neutrophil chemotaxis due to decreased production of interferon gamma is thought to cause the disease (3). Both autosomal dominant and recessive inheritance have been described. The disease was linked to mutations in the STAT3 gene after cytokine profiles indicated alterations in the STAT3 pathway (4).

Elevated IgE is the hallmark of HIES, usually > 10 times normal (5). However, patients younger than 6 months of age may have very low to non-detectable IgE levels. Eosinophilia is a common finding (5). IgE levels in HIES exceed 2000 IU/ml. However, IgE levels may decrease with age, may fall within the normal range (0.1-90 IU/ml) in about 20% of the cases (5,6).

HIES often appears early in life with recurrent staphylococcal and candidal infections, pneumonias and eczematoid skin. Finally, some patients have abnormal facial features and scoliosis and fragile bones (5). Most patients with HIES are treated with chronic antibiotics to help and protect them from staphylococcal infections. Good skin care is also important in patients with HIES. Intravenous gamma globulin has also been suggested for the treatment.

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From: Department of Pediatrics, U.P. Rural Institute of Medical Sciences and Research, Saifai Etawah, India

Address for Correspondence: Dr Mukesh Vir Singh, Lecturer Pediatrics, U.P.Rural Institute Of Medical Sciences And Research, Saifai Etawah, Uttar Pradesh, India. E-mail: mks007007@rediffmail.com

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