

Low carbohydrate low sodium diet, spironolactone and diclofenamid have also been tried in the management of HPP(7). None of these measures prevent progressive myopathic changes.

To conclude, in addition to rarity, the present case also emphasizes the usefulness of acetazolamide in the treatment and the case with which recurrent attacks can be prevented by potassium supplementation.

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Hyperimmunoglobulin E Syndrome

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Hyperimmunoglobulin E syndrome (HIE) is a rare immunodeficiency disorder which

presents with repeated pulmonary infections, pyoderma, otitis media and subcutaneous abscesses, in the child who has coarse features, short stature, eczema and ungal candidiasis. Serum IgE levels are usually greatly raised. Although 130 cases have been reported world-over, to the best of our

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knowledge no case has been reported in the Indian literature.

Case Report

A male, born of non-consanguineous marriage, was first seen in May 1986, at the age of 2 years. He had pneumothorax and an abscess 8 x 8 cm on the left thigh. His weight and height (8 kg and 87 cm, respectively) were both below the 5th percentile. He had a course facies, a square jaw, and brown hair. He had, received OPV, DPT and BCG at appropriate ages.

In the past history, he was well only for 2 months after birth. He had suffered from diarrhea, paronychia, and had 3 attacks of pneumonia. His investigations revealed a Hb of 9.6 g/dl, total leukocyte count 15.2×10^9 /dl, with 3,800 band cells, neutrophils 65%, lymphocytes 20% and eosinophils 15%. Culture of pus from the pleural cavity and abscess grew *Staphylococcus epidermidis* and *Pseudomonas aeruginosa*, respectively.

He was treated with intercostal drainage and appropriate antibiotics. Over a 7 year follow up, he had 3 attacks of pneumonia along with bronchopleural fistula in which staphylococci were grown from the pleural aspirate, persistent large pneumatoceles on X-ray, repeated attacks of eczema and pyoderma, had 4 attacks of deep multiple "cold" abscesses of the scalp, cervical, axillary and gluteal regions (from which staphylococcus was cultured), and had repeated attacks of otitis media, and fungal onychia. He has now developed Grade II clubbing.

Serum proteins, albumin globulin ratio, fractional globulins, serum calcium, phosphorus and alkaline phosphatase levels were normal. Immunologic tests done on the child

were as follows: there were 54% T-cells and 45% B-cells in the lymphocyte series. CD4+ cells were 15% and CD8 + cells were 51%. Absolute eosinophil counts showed high levels between 700-1500/mm³ during periods of illness. PAS and myeloperoxidase staining showed normal morphology of polymorphonuclear (PMN) cells. The *in vivo* leukocyte migration test (Rebeuk window test) showed abnormal PMN mobility but the nitrobluetetrazoline (NBT) dye test (which is a test for intracellular killing and oxidative response) was normal. The complement system was normal as seen by a normal total hemolytic complement activity (CH⁵⁰ = 100 mg/dl) and a normal level of C3 (14 mg/dl). Skin testing with recall antigens (*Candida albicans*, *Aspergillus fumigatus*, streptokinase, trichoderma and PPD) was non-reactive, indicating an abnormal CMI. The serum immunoglobulins showed a normal IgG and IgM (200 and 249 IU/ml, respectively), a low IgA (48 TU/ml) and a markedly raised IgE (40,000 IU/ml).

Lymphnode biopsy revealed non-specific lymphadenitis and infiltration with eosinophils. Skin biopsy showed dense infiltration with mononuclear cells.

In view of the clinical features of repeated infections with staphylococci, fungal onychia and laboratory findings of normal WBC morphology, normal granularity, killing of neutrophils but abnormal chemotaxis (Rebeuk window test), abnormal T4 : T8 ratio, blood and tissue eosinophilia, abnormal response to recall antigens, markedly increased serum IgE with low IgA levels, we labelled this case as HIE syndrome.

Discussion

Davis *et al.* in 1966 reported the association of infected eczematous dermatitis

with sinusitis, acute pulmonary infections and recurrent "cold" staphylococcal abscesses in two Caucasian red haired girls. They named it as Job's syndrome, after the Biblical character(1). Rebecca Buckley in 1972 reported two further children with eczema, multifocal infections, dysmorphic syndrome and high serum TgE concentrations(2).

HIE is a rare disease, which appears usually before the age of 2 years. The clinical features include recurrent multifocal infections(3) affecting lungs, skin, ears, and sinuses caused by *Staphylococcus aureus*, *Streptococcus pneumoniae*, or Gram negative bacilli, and infected eczematous dermatitis. Half the patients have mucocutaneous candidiasis involving nail beds, esophagus and vagina. Growth is dysmorphic with course facies and prognathism. Other reported disorders include asthma (10%), osteogenesis imperfecta and axial osteoporosis (about 50%). TgE levels are usually more than 4000 IU/ml.

The host defence defect results probably from a T-suppressor cell abnormality thereby allowing unregulated T-helper activity to augment IgE production. Though, there is no structural defect in the IgE molecule, the IgE is nevertheless ineffective. B-cell abnormality is shown by defective immunoglobulin synthesis *in vitro*. Mononuclear cells from HIE patients have been shown to produce an inhibitor of leukocyte chemotaxis, but this defect of neutrophil chemotaxis is fluctuating(4). There is usually a normal E-rosetting count and the CD4+ cell count is also usually normal, but CD3+ and CD8+ lymphocytes are usually depressed(5). Blood and tissue

eosinophilia is usually present. The anamnestic response to vaccines and recall antigens is usually poor, but immediate reaction to inhaled or ingested allergens are frequent even though skin tests for the same may show no response. Several diseases like systemic mastocytosis, systemic lupus erythematosus, Hodgkin's disease and non-Hodgkin's lymphoma, may be seen in association with HIE. The complications mainly involve the respiratory system, and include chronic bronchitis, bronchiectasis, empyema, pneumothorax and bronchopleural fistulas(6). Pneumatoceles are common and may need surgical excision. Treatment lies in long acting Penicillinase resistant penicillins to prevent infections of vital organs.

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