

MEETING ABSTRACT

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Hyperimmunoglobuline e syndrome: case report

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Background

Hyperimmunoglobulin E syndrome (HIES) is a rare disease caused by a primary complex immunodeficiency, with IgE serum levels over 2,000U/mL. The syndrome is characterized by the following triad: 1) recurrent staphylococcal abscesses, 2) recurrent airway infections; 3) elevated serum IgE concentration. Our patient presented signs that are compatible with HIES in distinct moments of his life – microabscesses associated with eczema at adolescence, respiratory infections from childhood until present moment and serum IgE level of over 2,000 in recent dosage.

Methods

This is a case report.

Results

55 year old male complains of intense pruritus over entire body, with 100°F fever and shiver episodes when pruritus was more intense, associated with 11kg weight-loss during one year. During first year of life, he presented furunculosis; at adolescence, featured diffuse eczematous lesions, highly pruritic and associated with frequent impetiginization. Patient has asthma since childhood, with two previous hospitalizations due to pneumonia.

At dermatologic exam, patient presented hyperkeratotic erythematous-desquamative plaques, with edema, erosions and desiccated serous crusts, as well as fissures and ulcers. Linear excoriation signs and xeroderma were present in all integument. IgE dosage evidenced levels over 2,000 U/mL, without specified exact value. The result of pathological exam presented spongiosis and psoriasiform superficial dermatitis with prominent eosinophils.

Several therapeutic approaches have been tried in the past, all unsuccessful. It was then prescribed

Metotrexate with folic acid and prednisone – patient responded with significant clinical improvement, especially decrease of pruritus. The dose of prednisone was progressively decreased until 40mg/day associated with Metotrexate 7.5mg 3 times/week and folic acid 5mg 4 times/week. Patient persists with important decrease of lesions and consequent improvement of life quality.

Conclusions

Cutaneous infections often happen, and furunculosis and cellulitis may be observed. "Cold abscesses", which are neither erythematous nor painful, occur mainly in patients that are not submitted to prophylactic antibiotic therapy; they are pathognomonic of HIES, though not indispensable for diagnosis. Recurrent respiratory infections cause pulmonary sequelae that lead to chronic respiratory failure, the main cause of death in patients with this disease.

HIES is a condition whose diagnosis is hard, mostly due to its rarity. Frequently, patients pass by several medical institutions and go through partially or completely unsuccessful therapeutic approaches, what delays even more the correct diagnostic conclusion and the beginning of effective treatment. Nevertheless, after the introduction of appropriate immunosuppressive therapy, patients tend to stabilization of clinical condition and important improvement of life quality.

Consent

Written informed consent was obtained from the patient for publication of this abstract and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

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