GENETICS AND GENODERMATOSES

A CASE OF HYPERIMMUNOGLOBULIN E SYNDROME AND KIMURA DISEASE

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Background: Hyperimmunoglobulin E syndrome is characterized by recurrent skin abscess, pneumonia, eczematous dermatitis, and elevated serum IgE (Immunoglobulin E) levels. Eczematous dermatitis and lichenification, accompanied with pruritus, often lead to excoriation, which is similar to atopic dermatitis. In addition to skin lesions, Hyperimmunoglobulin E syndrome shows characteristic facial features, and skeletal anomalies.
Kimura disease commonly presents as painless lymphadenopathy or subcutaneous masses in the head and neck region. Often patients with Kimura disease demonstrate peripheral eosinophilia and elevated level of serum IgE.

Observation: A 15-year-old male patient presented with disseminated erythematous papules on whole body with itching. In addition to skin lesion, scoliosis and a mass on left neck, diagnosed as Kimura disease, were also present. Blood test showed elevated level of IgE (> 5,000KU/L) and hypereosinophilia. The use of oral and topical steroids and oral immunosuppresant improved lesions of the trunk. However, the lesions of the extremities, including hands and feet, remained unchanged as they continued to improve and deteriorate. IVIG treatment was attempted, but there was no significant improvement so far.

Key message: Kimura disease ususally shows eosinophila or elevated serum IgE levels. However, it is rarely accompanied by hyperimmunoglobulin E syndrome. We report an unusual case of Kimura disease and hyperimmunoglobulin E syndrome.