

Genodermatoses

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Epidermolytic hyperkeratosis associated with generalized pustular psoriasis

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A 27 year-old Phillipino female with bullous congenital ichthyosiform erythroderma and lifelong epidermolytic hyperkeratosis (EHK) was hospitalized for a febrile and generalized erythematous pustular eruption. The eruption had begun one-week prior to admission with increasing erythema and scaling as well as tenderness over her neck, trunk and arms which later generalized.

A few days prior to presentation, she was treated with prednisone at a local clinic. She denied signs or symptoms of antecedent infection but had been under more stress. She had no personal or family history of psoriasis. Cutaneous exam revealed generalized polycyclic erythematous scaling plaques, some studded with pustules on a background of ichthyotic skin. A biopsy showed psoriasiform epidermal hyperplasia with subcorneal and interepidermal neutrophilic abscesses. DIF was non-diagnostic. On admission, the patient was febrile and had leukocytosis ($24.3 \times 10^9/L$) with a predominance of neutrophils. KOH and swab culture of skin was negative. Infectious serologic studies were also negative. The patient was started on methotrexate and infliximab, which was associated with dramatic improvement of the erythroderma. Her fever and leukocytosis also resolved. Interestingly, she also had complete desquamation and virtual clearance of the EHK and keratoderma of the palms and soles. However, typical lesions of EHK and keratoderma eventually recurred. She was maintained on infliximab with initial excellent control and later switched to adalimumab. The co- occurrence of two uncommon to rare skin diseases, bullous congenital ichthyosis/EHK and generalized pustular psoriasis is rare.

Genetic factors in the development of pustular psoriasis have not been well-characterized. Possible factors associated with the development of pustular psoriasis in this patient include a disrupted epidermal barrier due to EHK or rapid corticosteroid taper. EHK may also show dramatic improvement as part of the inflammatory response and treatment with anti-TNF α agents. These findings suggest that skin diseases with strong genetic components may co-operatively interact to trigger or ameliorate the severity of a co-existing genodermatosis.

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