Congenital Ichthyosiform Erythroderma with *Trichophyton rubrum* Infection

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Ichthyosis is the name given to a group of disorders characterized by a generalized persistent non-inflammatory scaling disorders of the skin surface. There are various forms of ichthyoses classified according to their mode of inheritance, clinical and pathologic features, and the underlying defect. Ichthyosis comprises three conditions according to their mode of inheritance: autosomal dominant ichthyosis, sex-linked recessive ichthyosis, and autosomal recessive ichthyosis(1).

In many instances it is possible to subdivide recessive ichthyosis into two types. A less severe type, congenital ichthyosiform erythroderma, shows fine white scales with fairly pronounced erythroderma and has a tendency to improve at the time of puberty. The more severe type, lamellar ichthyosis, shows large, plate-like scales and severe ectropion but only slight erythroderma. In both forms the flexural surfaces and the palms and soles are involved(2). We report a case of congenital ichthyosiform erythroderma with *Trichophyton rubrum* infection.

Case Report

A 10-year-old caucasian girl born of a non-consanguineous marriage was referred to the skin clinic for itching, scaling, redness and dryness of the skin which started during the first months of life. She had no history of recurrent skin blisters or dental abnormalities. Two months ago she was hospitalized with a generalized erythroderma and treated with corticosteroids and antihistaminics. Skin examination showed erythroderma, generalized fine micaceous scaling involving all flexures, scalp, palmar and plantar areas, and facial involvement with mild ectropion, palmar and plantar hyperkeratoses, and thickening of both toenails and right fingernails (Figs. 1 & 2). Hair and buccal mucous membranes were not affected. Examination of other systems were normal.

A hematoxylin-eosin-stained section of a skin specimen from lesion of the trunk showed hyperkeratosis with focal parakeratosis, irregular increase in granular layer, regular acanthosis and dermal superficial perivascular infiltrate. A diagnosis of non-
bullous congenital ichthyosiform erythroderma (NCIE) was made (Fig. 3). Examination of the scrappings from the scalp, trunk, arms and thickened nails after lysis with potassium hydroxide showed a dense mat of intertwining hyphae, fungal culture showed evidence of infection with Trichophyton rubrum. The patient was treated with oral ketoconazole (100 mg daily) with marked improvement within 5 days and virtual clearing of the erythroderma after one month. The underlying NCIE became evident. Nail lesions cleared more slowly over the next four months.

Discussion

The relationship of fungal infections and various forms of ichthyosis is infrequently reported(3). Koblenzer and Miller(4) first reported a patient with lamellar ichthyosis and fungus infections caused by Trichophyton rubrum; subsequently cases with various forms of ichthyosis associated with fungal infections caused by Trichophyton rubrum, Epidermophyton floccosum, Candida spp, and Trichophyton mentagrophytes have been described(3,5,6).

In ichthyosis of the common type, desquamation of keratin is retarded and this may be the factor responsible for the persistence of ringworm infections(7); moreover, ichthyosis is often associated with an atopic state, and the depressed cell-mediated immunity present in some of these patients could further predispose to superficial fungal infections(6-8). We did not find any immunologic alterations in our patient,
which makes us think that topical treatment and characteristics of the ichthyotic skin facilitate the infection as Moreno-Gimenez thought(3).

The present case and review of the literature suggests that cases with ichthyosis and dermatophytosis are more frequent than realized, but difficult to diagnose, so patient with unexplained exacerbation of ichthyosis or erythroderma must be evaluated for superimposed fungal infection.

REFERENCES


6. Moreno-Gimenez JC, Garcia-Perez A. Dermatofitosis en distintas formas de-
Resolution of Cyclic Neutropenia by Intramuscular Gamma Globulin in a Case of Common Variable Immunodeficiency with Predominantly Antibody Deficiency

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Cyclic neutropenia is a rare disorder and as the name suggests, neutropenia occurs cyclically and neutropenic nadirs are seen every 21 ± 2 days. It often presents by 5 years of age and is accompanied by recurrent illnesses that coincide with the agranulocytic period(I). Although the etiology is unknown, it has been associated with dysgammaglobulinemia(2). Therapy with granulocyte-colony stimulating factor (GCSF) has been successful(3).

We present a case of common variable immunodeficiency (CVI) with mainly antibody deficiency, along with a typical cyclic neutropenia. The patient was successfully treated with intramuscular gammaglobulin.

Case Report

A 1½-year-old boy was referred for fever every 15 to 20 days since the age of 11 months. He was free of fever only for 2 months when he was 14 months old.

Three weeks prior to the present illness he had a purulent ear discharge and pneumonia with right upper lobe consolidation. His Hb was 11.1 g/dl, Hct 33.6%, WBC 9.2 × 10^9/L, polymorphonuclear leukocytes (PMN) 51% and nonsegmented neutrophils (NS) 3%, IgG 302 mg/dl, TgA 22 mg/dl and IgM 49 mg/dl (normal range for TgG: 423-1184 mg/dl, TgA: 35-222 mg/dl, IgM: 22-131 mg/dl).

On examination, the child was afebrile but irritable and had a swollen, red, left middle finger following an injury. He weighed 10 kg. His investigations were as follows: Hb 10.1 g/dl, Hct 33.6%, WBC 5.6 × 10^9/L, PMN 1%, NS 1%. The following day he had a temperature of 103°F. His absolute neutrophil count (ANC) was zero, at this stage (Fig. 1).