

Dermatology Online Journal

Volume 17 Number 10

October 2011

[DOJ](#)

[Contents](#)

Erythroderma of unknown etiology

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Dermatology Online Journal 17 (10): 6

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Abstract

We present a 43- year-old man with a greater than 15-year history of erythroderma. A definitive diagnosis has not been established. The differential diagnosis is discussed.

History



Figure 1



Figure 2

A 46-year-old man presented to Bellevue Hospital Center with a widespread, erythematous, pruritic eruption that involved more than 80 percent of his body. The eruption started on his shoulder when he was in his 20s and, over the course of a one-year, spread to involve his face, trunk, and limbs. He was treated by various doctors in China and New York City with pills and creams with minimal benefit. He was not on any medications at the onset of the eruption and was working at a clothing manufacturing company. He has since filed for disability and does not currently work. A review of systems was negative for fevers, chills, arthralgias, fatigue, weight-loss, worsening of eruption in the sun, enlargement of lymph nodes, oral ulcers, and personal or family history of atopy and psoriasis.

Physical examination

Confluent, erythematous plaques with fine scale involved the face, neck, trunk, and limbs. Areas of sparing at the antecubital and popliteal fossae were noted. There was thinning of the eyebrows centrally and thickened plaques with more hyperkeratotic scale on the dorsal aspect of the hands. No ectropion was noted. Hypertrophic periungium with loss of cuticles in several fingers also

was noted.

Laboratory data

A complete blood count and a comprehensive metabolic panel were normal. Eosinophilia was 6 percent. Serum IgE level was 173 IU/mL. Peripheral blood flow cytometry showed a normal CD4/CD8 ratio and no atypical lymphocytes. T-cell receptor gene rearrangement studies were negative.

Histopathology

There is a perivascular and band-like inflammatory infiltrate that is comprised predominantly of lymphocytes, histiocytes, and eosinophils. Some lymphocytes extend to an overlying hyperplastic epidermis where there is spongiosis, rare necrotic keratinocytes, and focal parakeratosis.

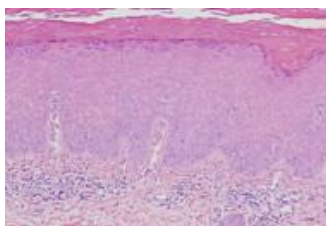


Figure 3

Discussion

Erythroderma is a term that is used to describe a variety of dermatologic diagnoses that present as a scaling, erythematous dermatitis, which involve greater than 80 percent of body surface area. Commonly, erythroderma is a presentation of an already established diagnosis such as psoriasis or atopic dermatitis. However, in about 25 percent of erythrodermic cases, no definitive etiology is found [1].

A recent study in China that surveyed 82 patients with erythroderma found psoriasis to be the most common cause (30%) [2]. There also were cases of hypereosinophilic syndrome, sarcoidosis, and dermatomyositis that led to generalized erythema and scale. Among drug-induced cases of erythroderma (17%), Chinese herbal remedies were the leading cause (67%). Another case series in which 67 patients with erythroderma were examined, the top two culprits were psoriasis and adverse drug reactions, followed by cutaneous T-cell lymphoma [3]. This study also found a non-causative association between elevated levels of lactic acid dehydrogenase and peripheral eosinophilia and paraneoplastic erythroderma related to an underlying malignant condition. Hereditary ichthyoses also should be considered in the differential diagnosis if the age of the individual is appropriate.

Our patient's case highlights an erythroderma that started in his late 20s as a localized patch on a sun-exposed area (neck/shoulder) and then progressed to involve more than 90 percent of his body. Clinically, the most striking finding is the sparing of the antecubital and popliteal fossae, which is suggestive of the deck-chair sign of papuloerythroderma of Ofuji (PEO) [4].

This clinical entity, which was first described in 1984, is comprised of intensely pruritic erythroderma as a result of coalescing papules, with sparing of the flexural fossae and with a peripheral eosinophilia, with or without elevated serum IgE levels. Although a distinct clinical entity, PEO is not a homogenous disease with well-understood pathophysiology. A PubMed search yielded 66 articles that were dedicated to this condition, with most of them highlighting a unique underlying association of PEO with gastric cancer (most common solid tumor), Sézary syndrome, and disseminated strongyloidiasis [5, 6]. With the exception of cutaneous T-cell lymphoma (CTCL) leading to PEO, histopathologic findings are varied and non-specific [7].

One of the most worrisome diagnoses in the differential for erythroderma is CTCL. The histopathologic confirmation of CTCL can be quite difficult and often necessitates multiple biopsies separated in time. In a recent retrospective study that analyzed 97 patients with erythrodermia, mycosis fungoides was found to have the best clinicopathologic correlation [8]. Nonetheless, it is imperative that CTCL remain high on the differential diagnosis until blood flow cytometry and gene rearrangement studies are demonstrated to be negative in addition to histopathologic features.

The widespread eruption, the corrugated appearance on the dorsal aspect of the hands, and the sparing of flexural areas also invoke diagnoses of various ichthyoses and erythrokeratodermas. Although most genodermatoses are present at birth or early infancy, later presentation in childhood can be seen. Since Netherton syndrome was considered, our patient has had a trichogram, which was normal. He is waiting for an ophthalmologic examination.

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