ONLINE EXCLUSIVE

Diffuse erythematous rash resistant to treatment

Our patient's rash had spread to most of her body and returned after initial treatment attempts. A skin biopsy helped us to figure out why.

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The authors reported no potential conflict of interest relevant to this article.

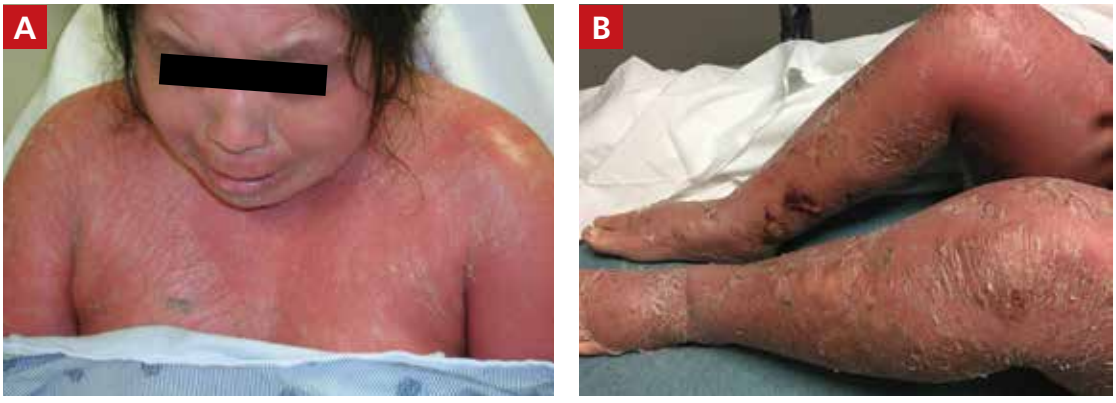
A 39-YEAR-OLD WOMAN presented to the emergency department for evaluation of diffuse redness, itching, and tenderness of her skin. The patient said the eruption began 4 months earlier as localized plaques on her scalp, elbows, and beneath both breasts. Over the course of a few days, the redness became more diffuse, affecting most of her body. She also noticed swelling and skin desquamation on her lower extremities.

The patient had visited multiple urgent care clinics and underwent several courses of prednisone with initial improvement of symptoms, but experienced recurrence shortly after finishing the tapers.

On physical examination, more than 95% of the patient's skin was bright red and tender to the touch, with associated exfoliation (**FIGURES 1A-1B**). Her lower extremities had pitting edema with superficial erosions that were weeping serous fluid. She was afebrile and normotensive, but had shaking chills and was tachycardic, with a heart rate of 115 bpm. There was no nail pitting, pustules, or lymphadenopathy. Lab tests revealed a low albumin level of 2.2 g/dL (normal: 3.5-5.5 g/dL), an elevated white blood cell count of 14,700 cells/mcL (normal: 4500-11,000 cells/mcL), and normocytic anemia (low hemoglobin of 8.7 g/dL; normal: 12-15.5 g/dL). The patient was admitted.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE 1
An erythematous rash, with exfoliation, covers 95% of the patient's body



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Erythroderma is usually a clinical presentation of an existing dermatosis, such as psoriasis or atopic dermatitis.

Diagnosis: Erythroderma

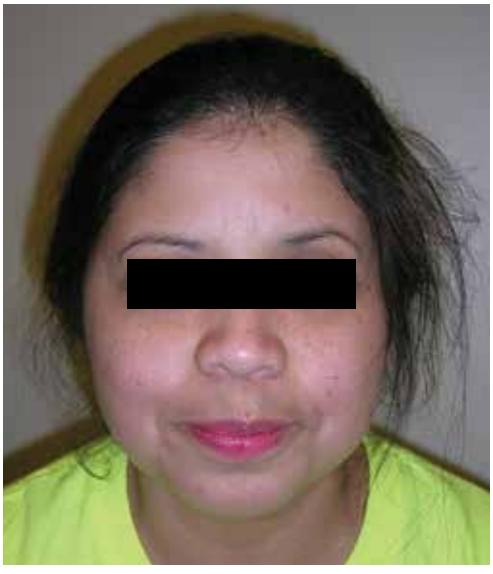
Based on the patient's clinical presentation, we diagnosed severe erythroderma secondary to psoriasis. A punch biopsy was performed, and pathology demonstrated subacute spongiotic dermatitis with superficial neutrophilic infiltrates, consistent with psoriasis.

Erythroderma is widespread reddening of the skin associated with desquamation, typically involving more than 90% of the body's surface area.[1] In most instances, erythroderma is a clinical presentation of an existing dermatosis. The most common causative conditions include primary skin disorders (such as psoriasis or atopic dermatitis), idiopathic erythroderma, and drug eruptions. Less common causes include cutaneous T-cell lymphoma, pityriasis rubra pilaris, and contact dermatitis.[1]

It's unclear why some skin diseases progress to erythroderma; the pathogenesis is complicated and involves keratinocytes and lymphocytes interacting with adhesion molecules and cytokines. Erythroderma can arise at any age and occurs in all races, but is more common in males and older adults, with a mean age of 42 to 61 years.[2] The annual incidence of erythroderma is estimated to be one per 100,000 adults.[3]

FIGURE 2

One month after treatment began...



...the erythroderma was resolved.

A complete picture of the patient is essential to making the diagnosis

Diagnosis can be difficult and hinges on historical and physical exam findings, as well as lab evaluations and skin biopsies. The history should focus on current and former medications, while the physical exam should hone in on clinical manifestations of existing dermatoses. The most common extracutaneous finding is generalized lymphadenopathy, which if prominent, may warrant lymph node biopsy, with studies for evaluation of underlying lymphoma.

Tachycardia develops in 40% of patients, secondary to increased blood flow to the skin and fluid loss, with risk of high-output cardiac failure.[2] Patients often have chills because their skin is not able to regulate their body temperature normally.[4]

The lab evaluation should include a complete blood count with differential and a comprehensive metabolic panel, as well as blood, skin, and urine cultures if infection is suspected as an inciting factor. Typical findings include mild anemia, leukocytosis, eosinophilia, and an elevated erythrocyte sedimentation rate.[5] In addition, patients with chronic erythroderma commonly have low albumin.[6] Unfortunately, lab studies don't always reveal the underlying cause of the erythroderma.

Biopsies are commonly performed. However, the underlying etiology is often not clearly reflected in the result. Histology is typically nonspecific; findings frequently include hyperkeratosis, acanthosis, spongiosis, and perivascular inflammatory infiltrate. Additionally, the prominence of histologic features may vary depending on the stage of disease and the severity of inflammation. More specific findings may become evident later in the disease as the erythroderma clears, so repeated skin biopsies over time may be needed for diagnosis.[7]

Consider these conditions, which can lead to erythroderma

First and foremost, it is important to get a thorough history, particularly about prior skin conditions and symptoms that may indicate the presence of undiagnosed skin conditions.

Psoriasis is one of the most common causes of erythroderma. A history of preexisting psoriasis is very helpful, but when this is not present, a biopsy can help confirm a clinical suspicion for psoriasis. It also helps to look for clues of psoriasis like nail changes or a history of plaques over the elbows and knees.

Atopic dermatitis is another common cause of erythroderma, and the history might include scaling and erythematous patches or plaques involving flexural surfaces before erythroderma occurs. Patients may have a history of atopic dermatitis from childhood and/or a history of other atopic conditions such as asthma and allergic rhinitis.

Drug eruptions occur following the administration of a new medication and can mimic a myriad of dermatoses.

Cutaneous T-cell lymphoma can lead to erythroderma and be differentiated with skin biopsy; pathology may show atypical lymphocytes, and Pautrier's microabscesses may be seen.[8]

Pityriasis rubra pilaris is a relatively rare condition that presents with red-orange scaling patches and thickened yellowish palms and soles.[9]

Tx targets underlying etiology and associated complications

When treating a patient with erythroderma, it's important to prevent hypothermia and secondary infections. If symptoms are severe, hospitalization should be considered. Nutrition should be assessed, and any fluid or electrolyte imbalances should be corrected.

Oral antihistamines are commonly administered to suppress associated pruritus. Topical treatment usually consists of corticosteroids under occlusion with bland emollients. Depending upon the underlying disease, the following systemic medications may be started: methotrexate 7.5 to 15 mg once/week; acitretin 10 to 25 mg/d; or cyclosporine 2.5 to 5 mg/kg/d; in addition to topical treatment.[4]

Our patient. Pathology for our patient was indicative of psoriasis. She was started on a regimen of cyclosporine 4 to 5 mg/kg/d, diphenhydramine 25 to 50 mg as needed for itching, triamcinolone 0.1% ointment under wet wraps to her trunk and extremities, and hydrocortisone 2.5% ointment to be applied to her face daily. She was released after 5 days in the hospital. At outpatient follow-up one week later, her erythroderma was resolving. One month later, her erythroderma was resolved (**FIGURE 2**), although she did have psoriatic plaques on her lower legs. JFP

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Hospitalization should be considered to prevent hypothermia and secondary infections, as well as to assess nutrition and correct any fluid or electrolyte imbalances.