Diffuse rash with silvery scales and anasarca

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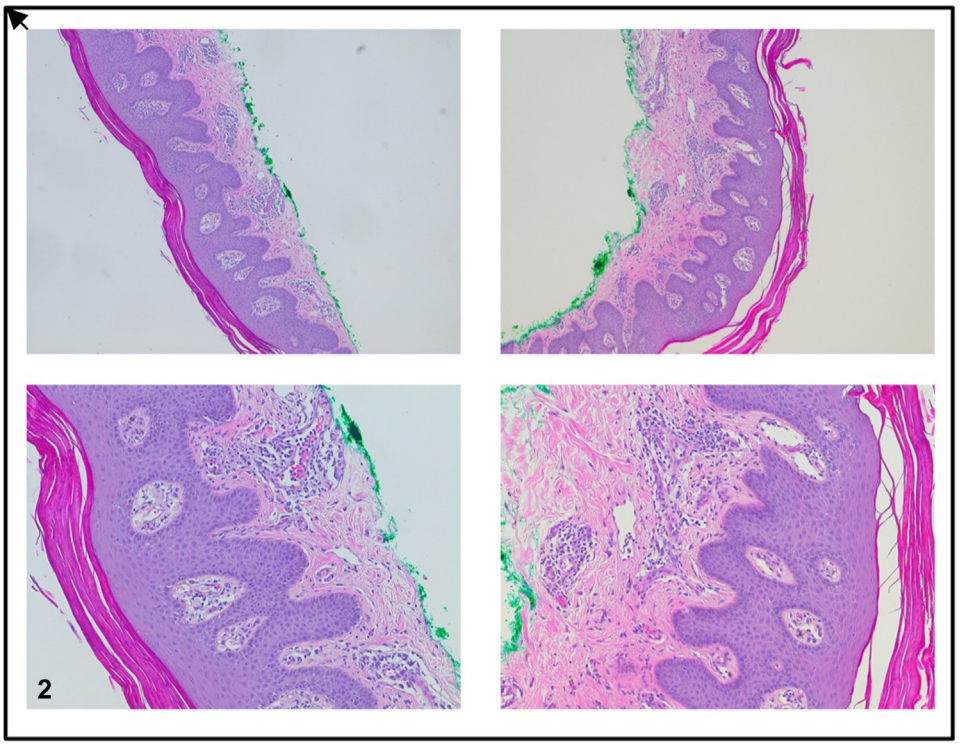

A 67-year-old African American female with a history of psoriasis presented with a rash for 9 months. The rash began on her back and limbs, did not resolve after a short course of oral steroids, and expanded. Review of systems was positive for edema of all limbs and hair loss. She denied any precipitating illness/injury, changes in medication, or substance use. She received topical triamcinolone and intravenous antibiotics with minimal improvement. Exam was significant for diffuse rash with silvery scales, with flakes covering her entire body and anasarca particularly in her limbs (Fig 1). Two shave biopsies were performed (Fig 2).

Question 1: Based on the clinical presentation and histopathologic findings, what is the most likely diagnosis?

A. Erythrodermic psoriasis (EP)

B. Chronic plaque psoriasis

C. Guttate psoriasis

D. Sézary syndrome

E. Toxic epidermal necrolysis

Answer:

A. EP — Correct. In patients with pre-existing psoriasis who present with widespread erythroderma and scaling >80% of body surface area, EP should be considered. Histology shows hyperkeratosis, parakeratosis, or inflammatory infiltrate in the dermis that may penetrate the stratum corneum (Munro's abscesses). In patients with darker skin, erythroderma may be difficult to identify.[1,2]

B. Chronic plaque psoriasis — Incorrect. Chronic plaque psoriasis presents with erythematous, round, desquamated plaques with sharply defined borders most commonly on the face, scalp, elbows, knees, and lower back.[1,2] While the patient has a history of plaque psoriasis, the widespread nature of this episode along with hair loss and anasarca makes EP the more appropriate diagnosis.

C. Guttate psoriasis — Incorrect. Guttate psoriasis presents as eruptions of small drop-like lesions, mainly on the trunk, and is most common in children/young adults with a family history of psoriasis. Lesions are typically preceded by a streptococcal throat infection and can be similar in appearance to pityriasis rosacea.[1,2]

D. Sézary syndrome — Incorrect. Sézary syndrome causes erythroderma due to cutaneous T-cell lymphoma. Symptoms additionally can show lymphadenopathy with an elevated CD4-CD8 ratio due to malignant proliferation. Skin biopsy shows T cells infiltrating the epidermis (Pautrier's microabscesses).[1]

E. Toxic epidermal necrolysis — Incorrect. Toxic epidermal necrolysis is characterized by "intense" erythema with full epidermal skin separation >30% of body surface area, typically with involvement of mucosal surfaces or hemorrhagic crusting of the lips. There may be a drug-trigger preceding onset, commonly with initiation of antibiotics. Skin biopsy shows perivascular inflammatory infiltrate, possibly eosinophils, and full epidermal separation.[1]

Question 2: Which of the following is considered a first-line treatment option for the acute management of this patient's condition?

A. Systemic corticosteroids

B. Cyclosporine

C. Topical steroids

D. Methotrexate

E. UV light

Answer:

A. Systemic corticosteroids — Incorrect. Systemic corticosteroids should generally be avoided due to a possibility of withdrawal precipitating erythrodermic flares or rebound erythroderma.[3]

B. .Cyclosporine — Correct. Cyclosporine inhibits calcineurin, impairing transcription of interleukin 2 (IL-2) and other cytokines in T-lymphocytes.[3] It's considered first-line for acute management of EP, particularly acute, severe, unstable EP, along with infliximab.[3] Cyclosporine is well tolerated but should be used cautiously in patients with hypertension or impaired renal function, as it may worsen these conditions or cause acute kidney injury.[3] This recommendation is largely based on an open-label, multicenter study of patients with EP where 67% of patients achieved remission and 27% had significant improvement at 2-4 months.[3]

C. Topical steroids — Incorrect. Medium-potency topical steroids should be utilized with systemic therapy as an adjuvant, not as monotherapy.[3] Supportive care with moisturizers, wet dressings, and oatmeal baths can also be considered.[3]

D. Methotrexate — Incorrect. Methotrexate is approved as a first-line therapy for patients with EP who have contraindications to cyclosporine or infliximab and/or present with less acute or severe disease.[3] Because our patient has no contraindication to cyclosporine and has an acute, severe presentation of EP, methotrexate would not be the most appropriate. Of note, acitretin is also approved with the same indications.[3]

E. UV light — Incorrect. The use of UV light is generally not advised as patients are typically photosensitive, and it's therefore challenging to administer suberythrogenic doses.[3] Photosensitivity can potentially lead to Koebnerization, where new psoriasiform lesions develop along sites of skin injury.[3]

.Question 3: Which of the following medications/biologic treatment options have shown efficacy and promising results in terms of chronic management of this patient's condition?

A. IL-17 inhibitor

B. Systemic corticosteroids

C. Coenzyme Q10

D. C-X-C motif chemokine ligand 10 inhibitor

E. Interferon-stimulated gene 15 (ISG15) inhibitor

Answer:

A. IL-17 inhibitor — Correct. IL-17A is a proinflammatory cytokine implicated in psoriasis and released by Th17 cells.[4] Secukinumab (anti-IL-17A monoclonal antibody), ixekizumab (anti-IL-17A monoclonal antibody), and brodalumab (anti-IL-17 receptor A monoclonal antibody) are approved as therapeutic options for psoriasis and are useful in treating EP.[4]

B. Systemic corticosteroids — Incorrect. Systemic corticosteroids should generally be avoided due to a possibility of withdrawal precipitating erythrodermic flares or rebound erythroderma.[3]

C. Coenzyme Q10 — Incorrect. Only 1 randomized controlled trial suggested benefit of coenzyme Q10, vitamin E, and selenium supplementation in the management of EP after treatment for 30 days.[5] Given the lack of further studies and evidence supporting the use of antioxidants, this is incorrect.

D. C-X-C motif chemokine ligand 10 inhibitor — Incorrect. C-X-C motif chemokine ligand 10 is increased in organ-specific autoimmune diseases, like Graves' disease, type 1 diabetes, and autoimmune thyroiditis, or in rheumatologic disorders such as rheumatoid arthritis, systemic sclerosis, lupus, and cryoglobulinemia, but not in EP.[5]

E. ISG15 inhibitor — Incorrect. ISG15 is a type 1 interferon-dependent transcript and is part of the ISG protein family that encodes a ubiquitin-like protein.[5] ISG15 is thought to play a major role in

innate immune responses and antitumor reactions, but not in EP.[5]

Abbreviations used:

EP: erythrodermic psoriasis
IL: interleukin
ISG15: interferon-stimulated gene 15

Conflicts of interest

None disclosed.

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