Coalescing plaques on the face, trunk, and upper extremities

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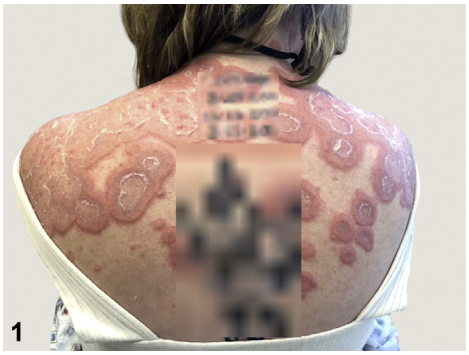

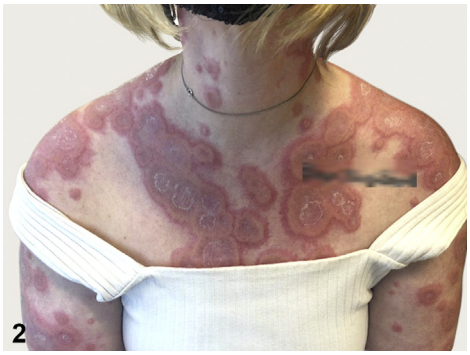

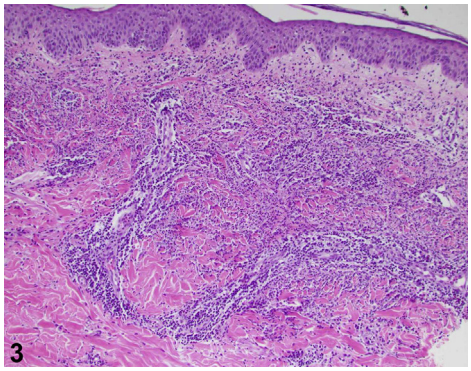

A 38-year-old woman from Southern Arizona with no significant past medical history presented with a 6-day history of a painful, pruritic rash. Examination showed pink, coalescing, annular plaques with central scale scattered across the face, upper extremities, back (Fig 1), and chest (Fig 2). She reported fatigue and migratory arthralgias but denied fevers, respiratory symptoms, new medications, pregnancy, or recent travel. Biopsy demonstrated papillary dermal edema with a dermal neutrophilic interstitial infiltrate, scattered eosinophils, and an associated perivascular lymphoid infiltrate (Fig 3). Autoimmune serology was negative for antinuclear and anti-Ro antibodies. Complete blood count with differential demonstrated neutrophilia, and the rash rapidly improved with topical 0.1% triamcinolone administration.

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Question 1: What is the most likely diagnosis?

A. Erythema nodosum (EN)

B. Sweet syndrome (SS)

C. Urticarial vasculitis

D. Neutrophilic eccrine hidradenitis

E. Erythema multiforme (EM)

Answers:

A. EN — Incorrect. EN is a delayed hypersensitivity reaction that presents with painful erythematous nodules, frequently involving the lower extremities. Histology reveals septal panniculitis with mixed inflammatory infiltrate and granulomatous inflammation without vasculitis.

B. SS — Correct. SS (acute febrile neutrophilic dermatosis) is an inflammatory disorder that presents with rapid onset, painful, edematous papules or plaques commonly involving the face, arms, and upper trunk. Typically, patients also present with neutrophilic leukocytosis and fever, though not all patients are febrile. SS may be associated with infection, malignancy, inflammatory disorders, pregnancy, or vaccination, and histopathology shows a dense dermal neutrophilic infiltrate with dermal edema and occasionally scattered eosinophils.[1,2]

C. Urticarial vasculitis — Incorrect. Urticarial vasculitis may present with urticarial or targetoid lesions, which persist for more than 24 hours but typically resolve within 72 hours. Histology reveals leukocytoclastic vasculitis of small vessels.[3]

D. Neutrophilic eccrine hidradenitis — Incorrect. Though also a neutrophilic dermatosis, neutrophilic eccrine hidradenitis predominantly occurs in patients with acute myeloid leukemia receiving chemotherapy. Neutrophilic eccrine hidradenitis presents with erythematous edematous plaques most commonly on acral surfaces, and histology reveals eccrine gland neutrophilic infiltrates with epithelial necrosis.[1]

E. EM — Incorrect. EM presents with targetoid lesions with a dusky center. Lesions typically involve the extensor acral extremities, and involvement of the trunk is less common. EM is generally asymptomatic and self-resolving, and histology reveals dyskeratosis of the basal keratinocytes and a dermal lymphohistiocytic infiltrate.[3]

Question 2: Of the following systemic fungal infections, which would most likely be detected in this patient?

A. *Histoplasma capsulatum*

B. *Cryptococcus neoformans*

C. *Coccidioides immitis/posadasii*

D. *Paracoccidioides brasiliensis*

E. *Blastomyces dermatitidis*

Answers:

A. *Histoplasma capsulatum* — Incorrect. *Histoplasma capsulatum* is endemic in the Mississippi and Ohio River valleys and Central and South America. In healthy patients, EN and EM are the most common dermatologic manifestations. Disseminated histoplasmosis, most common in immunocompromised or elderly individuals, presents with mucosal ulcerations and papules, plaques, or ulcers on the face and trunk.[4]

B. *Cryptococcus neoformans* — Incorrect. Cryptococcosis is seen in immunocompromised patients. Dissemination can present with subcutaneous papules, plaques, and nodules.

C. *Coccidioides immitis/posadasii* — Correct. *Coccidioides* is endemic in the Southwestern United States and Central and South America. Immunologic reactions such as SS, EN, and EM have been described. In this case, serologic testing for anti-*Coccidioides* IgM/IgG antibodies was positive. Extrapulmonary dissemination of *Coccidioides* may present with papules, ulcers, plaques, nodules, and abscesses.[4]

D. *Paracoccidioides* — Incorrect. *Paracoccidioides* is endemic in southern Mexico and Central and South America. Mucocutaneous fungal dissemination may present with mucosal ulceration and verrucous, necrotic, or ulcerative lesions around the mouth and nose.[5] There are very few reports of cutaneous immunologic reactions.

E. *Blastomyces* — Incorrect. *Blastomyces* is endemic in the Eastern and Central United States. Immunologic reactions such as EN can occur but are very uncommon. SS has not been associated with blastomycosis. Disseminated infection may present with painful ulcers, verrucous lesions, plaques, and subcutaneous nodules.[4]

Question 3: Which of the following statements is true regarding SS secondary to coccidioidomycosis?

A. SS is common in patients with coccidioidomycosis

B. *Coccidioides* spherules are commonly detected in skin lesions

C. Systemic corticosteroids are the mainstay of treatment

D. This phenomenon most frequently occurs in immunocompetent patients

E. Enzyme immunoassay (EIA)-based detection of anti-*Coccidioides* IgM/IgG antibodies is the most specific serologic test for diagnosis of coccidioidomycoses

Answers:

A. SS is common in patients with coccidioidomycosis — Incorrect. SS secondary to coccidioidomycosis is rare. EN and EM are much more common, occurring in approximately 25% of patients with pulmonary coccidioidomycosis.[4]

B. *Coccidioides* spherules are commonly detected in skin lesions — Incorrect. SS is an immunologic reaction associated with *Coccidioides* infection. Cutaneous lesions in disseminated disease may contain *Coccidioides* spherules.

C. Systemic corticosteroids are the mainstay of treatment — Incorrect. Though systemic corticosteroids are the first-line therapy for SS, in this situation, systemic treatment may worsen coccidioidomycosis infection and should be avoided.[2] Topical corticosteroids are indicated in this context. In our case, topical corticosteroids significantly improved lesions within 2 weeks.

D. This phenomenon most frequently occurs in immunocompetent patients — Correct. SS secondary to coccidioidomycosis occurs most often in immunocompetent patients and has even been described as a positive prognostic sign.[4] Immunosuppressed patients are at risk for extrapulmonary disseminated coccidioidomycosis with cutaneous involvement.

E. EIA-based detection of anti-*Coccidioides* IgM/IgG antibodies is the most specific serologic test for diagnosis of coccidioidomycoses — Incorrect. EIA-based serologic examination is useful for rapid assessment of anti-*Coccidioides* IgM and IgG antibodies. However, immunodiffusion is more specific for diagnosis, and positive serology as determined by EIA should be confirmed.[2] In our case, fungal serology showed the presence of anti-*Coccidioides* IgM and IgG antibodies as measured by immunodiffusion and positive anti-*Coccidioides* IgM antibodies as measured by EIA.

Abbreviations used:

EIA: enzyme immunoassay
EM: erythema multiforme
EN: erythema nodosum
SS: Sweet syndrome

Conflicts of interest

None disclosed.

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