Scaly erythematous papules and plaques in a teenager

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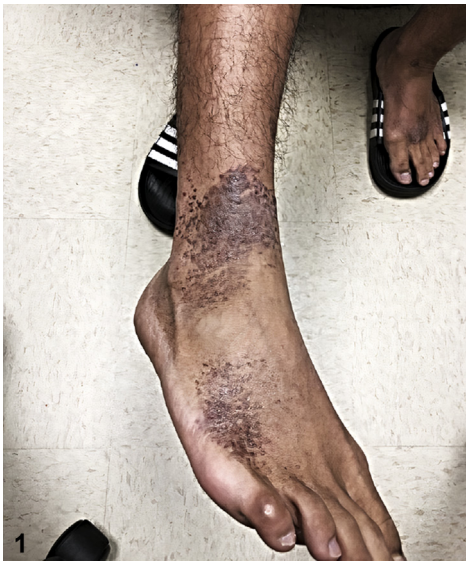

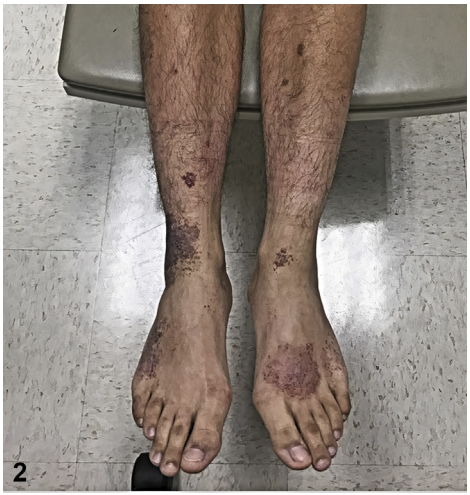

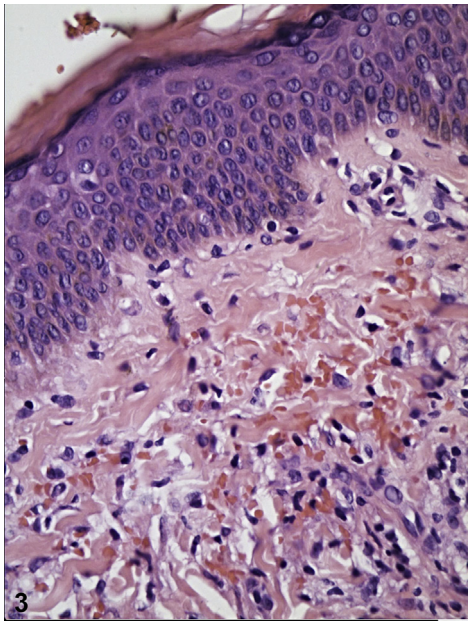

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A 13-year-old healthy Hispanic boy presented to the dermatology clinic with a 10-month history of a pruritic rash on his trunk and extremities. He was previously treated with fluocinolone 0.01% oil, triamcinolone 0.1% cream, and hydroxyzine 10 mg with no relief. The patient denied pain or a similar rash in the past. He had no relevant family or social history. Physical examination demonstrated non-blanching, scaly erythematous papules and plaques on his legs, right arm, and trunk (Figs 1 and 2). A punch biopsy from lower portion of his right leg was performed (Fig 3).

Question 1: Based on the clinical presentation, history, and histologic findings, what is your diagnosis?

A. Nummular eczema

B. Lichen planus

C. Erythema elevatum diutinum (EED)

D. Eczematid-like purpura of Doucas and Kapetanakis

E. Contact dermatitis

Answers:

A. Nummular eczema—Incorrect. Although clinically consistent with nummular eczema, biopsy revealed considerable perivascular dermal infiltrates and extravasated red blood cells, which are absent in eczema.

B. Lichen planus—Incorrect. While lichen planus can present with pruritic violaceous papules and plaques on the extremities, the histologic findings of the disease were absent in this patient's biopsy.

C. EED—Incorrect. EED is a necrotizing, small-vessel vasculitis characterized by erythematous plaques on the dorsal aspects of the hands or extensor surfaces. Histology would demonstrate a leukocytoclastic vasculitis with fibrinoid necrosis of the dermal vessel wall.

D. Eczematid-like purpura of Doucas and Kapetanakis—Correct. Eczematid-like purpura of Doucas and Kapetanakis often presents as an eczematous, non-blanching, red-brown purpuric or petechial eruption in the lower extremities. Epidermal spongiosis with perivascular lymphocytic infiltrates and extravasation of red blood cells are seen histologically.[1] Therefore, the clinical and histologic picture of our patient best reflect eczematid-like purpura of Doucas and Kapetanakis.

E. Contact dermatitis—Incorrect. Contact dermatitis could be a plausible diagnosis for our patient, as it presents with a well-demarcated erythematous papule or plaque confined to the site of contact with an allergen or irritant. However, the histologic features of contact dermatitis were absent in this patient.

Question 2: What is the best initial treatment for the management of eczematid-like purpura of Doucas and Kapetanakis?

A. Non-pharmacological interventions and topical corticosteroids

B. Narrow-band ultraviolet B phototherapy

C. Cyclosporine

D. Rutoside and oral ascorbic acid

E. Pentoxifylline

Answers:

A. Non-pharmacological interventions and topical corticosteroids—Correct. Eczematid-like purpura of Doucas and Kapetanakis can be a therapeutic challenge for dermatologists. The disease has a relapsing-remitting course and spontaneous improvement is common; therefore, the first-line initial management of the condition is medium-to-high-strength topical corticosteroids and compression stockings, as it is theorized that capillary dermal fragility and venous hypertension contribute to disease presentation.[2]

B. Narrow-band ultraviolet B phototherapy—Incorrect. Research on narrow-band ultraviolet B phototherapy and its effects on pigmented purpuric dermatoses (PPD) lesion size and pigmentation have demonstrated promising results. However, it is not a first-line management option in patients with PPD.

C. Cyclosporine—Incorrect. Immunosuppressants, such as cyclosporine, have demonstrated promising results in the treatment of PPD; however, the medication's adverse side effect profile makes it an undesirable treatment option.

D. Rutoside and oral ascorbic acid—Incorrect. While a retrospective review from 2014 and a handful of case reports demonstrated improvement of PPD lesions with 100 mg rutoside and 1000 mg vitamin C, the treatment is considered second-line.[3]

E. Pentoxifylline—Incorrect. In 2004, a single-blinded, randomized trial of adults with Schamberg disease compared a treatment regimen of 400 mg

pentoxifylline three times daily with 0.05% topical betamethasone two times daily. While patients demonstrated improvement in their lesion pigmentation on pentoxifylline therapy, it is considered a second-line treatment option and is not well-studied in pediatric patients.[4]

Question 3: Which of the following is true regarding eczematid-like purpura of Doucas and Kapetanakis?

A. The disease was first described by Doucas and Kapetanakis in 1953 as an eruption of red, blanching, hemorrhagic papules.

B. The disease is most often found in pediatric-aged boys.

C. In addition to the characteristic histological findings of exuberant perivascular lymphocytic infiltrates, eczematid-like purpura of Doucas and Kapetanakis also presents with epidermal spongiosis.

D. The initial lesion is often found in the upper extremities and can spread to the face or trunk.

E. Thrombocytopenia is the most common laboratory abnormality associated with the disease.

Answers:

A. The disease was first described by Doucas and Kapetanakis in 1953 as an eruption of red, blanching, hemorrhagic papules—Incorrect. In 1953, Doucas and Kapetanakis first described the clinical presentation of the disease as non-blanching hemorrhagic petechiae, which often coalesced to form dark-red plaques.[5]

B. The disease is most often found in pediatric-aged boys—Incorrect. To date, there are 2 reported cases describing eczematid-like purpura of Doucas and Kapetanakis in pediatric patients.

C. In addition to the characteristic histological findings of exuberant perivascular lymphocytic infiltrates, eczematid-like purpura of Doucas and Kapetanakis also presents with epidermal spongiosis—Correct. All PPD are characterized by the classic histological finding of perivascular lymphocytic infiltrates with extravasation of red blood cells. In eczematid-like purpura of Doucas and Kapetanakis, there is also epidermal spongiosis.

D. The initial lesion is often found in the upper extremities and can spread to the face or trunk—Incorrect. In most cases, the disease manifests first in the lower extremities and can spread to the trunk or upper extremities.

E. Thrombocytopenia is the most common laboratory abnormality associated with the disease—Incorrect. No laboratory abnormalities have been associated with eczematid-like purpura of Doucas and Kapetanakis.

Abbreviations used:

EED: erythema elevatum diutinum
PPD: pigmented purpuric dermatoses

Conflicts of interest

None declared.

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