Necrotizing Rash Associated with Autoimmune Hepatitis

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Background:
An unfamiliar skin eruption often prompts an immediate referral to dermatology, when more thorough evaluation may be warranted first. We describe a case where an unusual skin lesion was the first observed sign of an underlying gastrointestinal illness.

Clinical Case:
A previously well male presented with a burning skin rash.
Physical examination revealed multiple desquamating lesions. Serologic evaluation showed elevations in liver enzymes, international normalized ratio, antinuclear antibody, anti-smooth muscle antibody and gammaglobulin titers, thrombocytopenia and C4 hypocomplementemia. Skin biopsy revealed epidermal necrosis and diffuse lymphocytic infiltrate. Liver biopsy showed interface hepatitis. Upon initiation of steroids, the skin lesions and abnormal liver enzymes resolved.

Conclusion:
The clinical findings and response to steroids confirmed autoimmune hepatitis. This case highlights the importance and challenge the internist faces in distinguishing between primary dermatoses and cutaneous manifestations of gastrointestinal disease.

Case Presentation

A 49-year-old previously well male presented with a three-week history of multiple burning and peeling skin lesions. These lesions initially appeared on his right palm and fingers and subsequently spread diffusely. His review of systems was otherwise negative. The man denied any recent changes in soaps, lotions or detergents, trauma, known allergies, insect bites, heat or chemical exposure or sick contacts. He did not take any medications, vitamins or supplements. The patient immigrated to the United States from South America ten years ago but had not traveled since. He worked in construction but always wore gloves. He most recently engaged in sexual intercourse four years ago with one female partner, with intermittent condom use. He denied any history of sexually transmitted infection. He admitted to consuming two beers weekly for the past eight years as well as a remote history of marijuana use, but he denied use of tobacco or other recreational drugs.

On admission, the patient was afebrile and hemodynamically stable. His physical examination revealed multiple small, coalescing, desquamating, vesiculopustural lesions with surrounding erythema (Figure 1), in a scattered distribution on the bilateral palms, left elbow, scalp and bilateral ears. The mouth, groin and soles were spared. Nikolsky sign was negative.

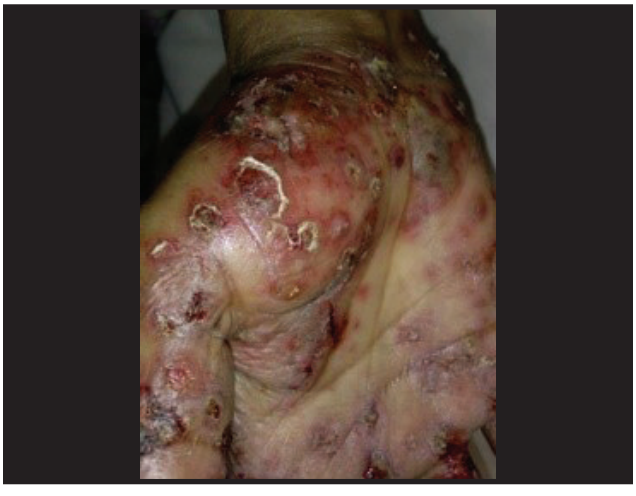

Figure 1. Coalescing desquamative vesiculopustular lesions on the patient's right palm.

Laboratory evaluation showed an elevated

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alanine aminotransferase of 100, aspartate aminotransferase of 571, alkaline phosphatase of 184, hyperbilirubinemia of 1.5, hypoalbuminemia of 2.7, increased international normalized ratio of 1.56 and thrombocytopenia of 18,000. He also had elevated antinuclear antibody (ANA) and anti-smooth muscle antibody (ASMA) titers of 1:320, hypergammaglobulinemia and C4 hypocomplementemia. Titers were negative for the viral hepatidites, rapid plasma reagin, human immunodeficiency virus, human T-cell lymphotropic virus, Babesia, Ehrlichia, double stranded deoxyribonucleic acid, Ro, cryoglobulins, porphyrins, glucose-6-phosphate dehydrogenase, rheumatoid factor, celiac panel and the anti-neutrophil cytoplasmic, cardiolipin, topoisomerase-1, histone and mitochondrial antibodies. The urine drug screen, gonorrhea screen and blood and skin cultures were negative. Skin biopsy revealed confluent epidermal necrosis with increased dermal mucin (Figure 2a) and diffuse perivascular and periadnexal lymphocytic infiltrate (Figure 2b), with no deposition on direct immunofluorescence (DIF). Liver biopsy demonstrated interface hepatitis. The patient was initiated on steroids, and his skin lesions and abnormal liver enzymes resolved after four months.

Case Discussion

The presence of elevated liver transaminases, ANA and ASMA titers and hypergammaglobulinemia in conjunction with liver biopsy finding of interface hepatitis and response to steroids all verify the diagnosis of autoimmune hepatitis (AIH).[1] While other dermatological conditions may be considered, they do not embody the clinicopathologic characteristics of this patient's skin lesions. While the skin's histological findings could suggest Rowell's syndrome,[2] the positive ANA titer was the only major criterion for the condition fulfilled in this case. Also, the lack of deposition on DIF and atypical clinical appearance for any of the subtypes excluded cutaneous lupus erythematosus. Perivascular and periadnexal lymphocytic infiltration can also be observed in polymorphous light eruption (PLE), Jessner's lymphocytic infiltration of the skin and reticular erythematosus mucinosis (REM).[3] However,

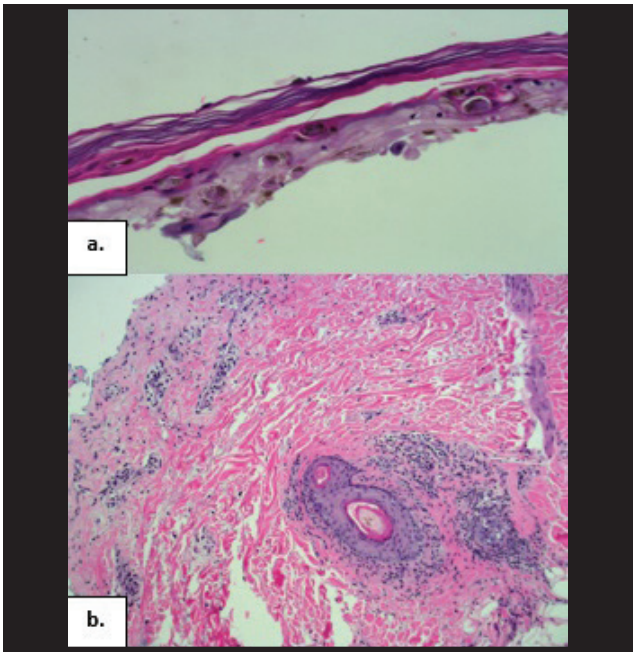

Figure 2. Histopathological specimen of the patient's skin showing confluent epidermal necrosis and increased dermal mucin (a) and diffuse perivascular and periadnexal lymphocytic infiltration (b). H&E stain. 100x magnification.

these differentials were excluded by the atypical clinical appearance, presence of mucin deposition and high ANA titer. The patient's skin eruption was attributed to a cutaneous manifestation of his underlying AIH.

Case Conclusion

When faced with an unusual desquamative diffuse skin rash as described here, the primary care physician may rush to consult dermatology, when instead a more comprehensive systemic evaluation should be pursued. Such an abnormal skin finding can actually be the first indication of something systemic. This case highlights the importance and challenge presented to the primary care physician in distinguishing between primary dermatoses and cutaneous manifestations of gastrointestinal disease. ■

References

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