

Dermatitis herpetiformis presenting as pseudovasculitis



Malan Kern, MD,^a Kevin H. Kim, MD, PhD,^b Gina Johnson, MD,^c
Spencer D. Hawkins, MD,^d and Henry K. Wong, MD, PhD^b
Little Rock, Arkansas and Winston-Salem, North Carolina

Key words: acral petechiae; dermatitis herpetiformis; leukocytoclastic vasculitis; pseudovasculitis.

INTRODUCTION

Dermatitis herpetiformis (DH) is a cutaneous manifestation of gluten intolerance characterized by an intensely pruritic, papulovesicular eruption with a predilection for the extensor extremities, scalp, and buttocks.¹ Because of its variable clinical presentation, DH can be a challenging clinical diagnosis. However, histopathology and immunofluorescence can provide vital data in confirming the diagnosis.² We report a case of a 47-year-old patient who presented with clinical features resembling vasculitis but with classic histopathology and immunofluorescence findings for DH.

CASE REPORT

A 47-year-old white man with a 7-year history of DH presented to the emergency department for the evaluation of a generalized petechial rash and tender swelling of the left arm. He first noticed the rash about a month prior after discontinuing dapsone, but it had progressively worsened over the preceding week. The rash was different than his typical DH presentation. In addition to the pruritic papulovesicular rash on the scalp and arms, he had flat, bright red lesions associated with severe burning pain. The patient's DH was previously well controlled with a gluten-free diet and dapsone, 100 mg daily. He reported that dapsone was discontinued a month prior by his primary care physician to avoid possible exacerbation of acute kidney injury secondary to severe dehydration. During this period, he did not adhere to a strict gluten-free diet. No new medications were recently started before presentation, and the patient's long-term medications included omeprazole for

Abbreviations used:

DH: dermatitis herpetiformis
DIF: direct immunofluorescence
EED: erythema elevatum diutinum

gastroesophageal reflux disease and lisinopril-hydrochlorothiazide for hypertension.

Physical examination of the skin found erythematous papulovesicles on the scalp, upper arms, and elbows and well-demarcated erythematous macules coalescing into larger patches distributed on the upper back, buttock, and trunk (Fig 1). In addition, purpuric macules and patches were noted in the periphery of erythematous patches on the anterior shins, knees, and anterior thighs (Fig 2). Scattered petechiae were also noted on the distal fingertips. Additionally, a large tender ulcer with purulent discharge and thick yellow crusting in the periphery was present in the left extensor forearm.

Laboratory findings were significant for an elevated white blood cell count (28.1 K/ μ L), and a left forearm wound culture was positive for methicillin-resistant *Staphylococcus aureus*. Biopsy of the left anterior thigh found a subepidermal blister with aggregates of neutrophils within the papillary dermal tips (Fig 3). There was pronounced dermal hemorrhage, but no evidence of leukocytoclastic vasculitis was identified (Fig 4). Direct immunofluorescence (DIF) showed granular IgA deposition in the papillary dermal tips (Fig 5). Given the overall clinical, histopathologic, and DIF findings, a diagnosis of DH was favored.

From the College of Medicine^a and the Departments of Dermatology^b and Pathology,^c University of Arkansas for Medical Sciences; and the School of Medicine, Wake Forest University.^d

Funding sources: None.

Conflicts of interest: None declared.

Correspondence to: Kevin H. Kim, MD, PhD, 4301 W. Markham Street, Slot 576, Little Rock, AR 72205. E-mail: khkim@uams.edu.

JAAD Case Reports 2017;3:444-7.

2352-5126

© 2017 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<http://dx.doi.org/10.1016/j.jidcr.2017.06.033>



Fig 1. Well-demarcated distributed erythematous macules coalescing into larger patches with geographic borders on the upper back.



Fig 2. Purpuric macules and patches in the periphery of erythematous patches on the anterior shins, knees, and thighs.

The patient was restarted on 100 mg daily of dapsone, topical corticosteroids, and a gluten-free diet for management of DH. The general surgery department evaluated the patient for the draining ulcer on the left forearm for possible debridement, but with rapid improvement

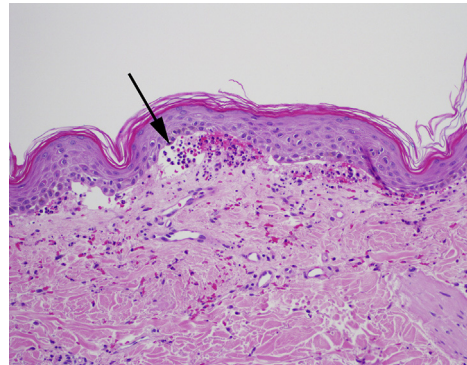


Fig 3. Subepidermal blister with aggregates of neutrophils within the papillary dermal tips (arrow) with pronounced dermal hemorrhage but no evidence of leukocytoclastic vasculitis. (Hematoxylin-eosin stain; original magnification: $\times 200$.)

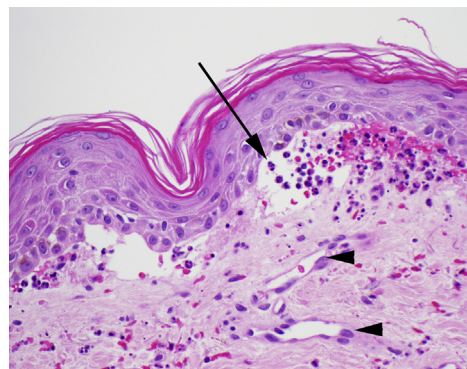


Fig 4. Higher magnification highlights aggregates of neutrophils within the papillary dermal tips (arrow) and no evidence of leukocytoclastic vasculitis (arrow head). (Hematoxylin-eosin stain; original magnification: $\times 400$.)

on intravenous vancomycin, they recommended continued antibiotic treatment for cellulitis and local wound care. On his 2-week postdischarge follow-up visit, the patient's skin lesions were dramatically improved with no new lesions and diffuse pink to tan patches on the extensor extremities consistent with postinflammatory hyperpigmentation.

DISCUSSION

The pleomorphic clinical appearance of DH can at times make diagnosis challenging. The classic findings of a pruritic grouped papulovesicular eruption favoring the extensor surfaces can clinically support the diagnosis; however, the dermatologist must be aware of the clinical variability of DH even in a clinical presentation mimicking eruption from other etiologies, such as vasculitis in this patient's presentation. This variability can be present in both the distribution—isolated scalp or facial

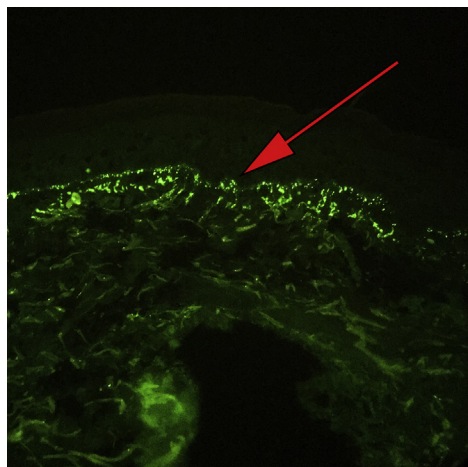


Fig 5. Direct immunofluorescence shows granular IgA deposition in the papillary dermal tips (arrow). (Original magnification: $\times 400$.)

involvement—and in the primary lesion—urticarial plaques or isolated excoriations secondary to pruritus.¹

Additionally, palmoplantar petechiae is increasingly reported in the literature as a component in the clinical presentation of DH with a significant predilection for the pediatric patient.²⁻⁴ Clinically, petechiae are only present on the palmoplantar surfaces with no involvement of the dorsal hands or feet. Although the specific pathophysiologic mechanism for this phenomenon is still unknown, the petechial lesions are present more often on the dominant hand, suggesting a possible role for trauma as an etiologic factor.

Our case of a 47-year-old white man with diffuse confluent erythematous macules and patches in addition to remarkable purpuric changes is an unusual presentation of DH. Although a vasculitic process was initially favored in the differential diagnosis because of the exuberant clinical appearance, no evidence of leukocytoclastic vasculitis was seen on histopathology. We find it intriguing that Naylor et al⁵ reported a similar presentation with diffuse petechiae in a 58-year-old man; however, histopathology findings in their case were consistent with both DH and leukocytoclastic vasculitis. Our case is unusual in that no such changes consistent with small-vessel inflammation were present on histopathology results but rather findings favoring pseudovasculitis with pronounced extravasated red blood cells, which can simulate vasculitis clinically.⁶ Further, our patient with no known history of thrombophilia also had petechiae on the distal fingertips. Although acral petechiae can be a common DH presentation in the pediatric population as previously mentioned, it is rarely

reported in an adult.^{3,4} The constellation of clinical findings in this patient show the pleomorphic manifestations of DH.

The histopathologic diagnostic criteria for active cutaneous vasculitis include angiocentric or angioinvasive inflammatory infiltrates, fibrinoid necrosis, and infiltration of vessel wall by an inflammatory infiltrate.⁷ Secondary changes such as red blood cell extravasation (hemorrhage) and subepidermal or papillary dermal neutrophils, can be suggestive, but not diagnostic, for active vasculitis. These secondary changes in the absence of histopathologic evidence of vasculitis should direct one to include pseudovasculitis in the differential diagnosis.⁶ Although we recognize that the histopathologic findings in vasculitis may exhibit a dynamic process as vasculitis progresses from an active acute to a chronic state, the clinically active vasculitis-like lesions in our patient did not have the histopathologic evidence required for the diagnosis of active cutaneous vasculitis.

Erythema elevatum diutinum (EED) is a chronic dermatosis that presents with symmetrical violaceous papules and plaques with a predilection for the extensor surfaces.⁸ Two articles in the literature report on EED in association with DH.^{9,10} In both cases, histopathologic findings showed changes consistent with a fibrosing leukocytoclastic vasculitis; specifically, early lesions demonstrating a dermal neutrophilic infiltrate with eosinophils, whereas late lesions showed perivascular fibrosis, fibrinoid necrosis, and intracellular lipidosis. Additionally, DIF showed linear IgA deposits.^{10,11} Although the morphology and the distribution of our patient's cutaneous findings may be similar to that of EED, the lesions in EED tend to be asymptomatic unlike those in our patient. Further, the histopathologic and DIF findings argue against EED, as no such evidence of leukocytoclasia was found in our patient other than a nonspecific dermal hemorrhage. DIF findings in our patient of granular IgA deposits in papillary dermal tips further argue against a diagnosis of EED. Although dapsone can be a treatment option for EED independent of DH, in those cases of concomitant disease, EED can be refractory to dapsone.⁹ Additionally, we note that dapsone can be an effective treatment for chronic small-vessel vasculitis; however, the overall clinicopathologic correlation in our case does not favor a vasculitic process. Therefore, we find it more likely that dapsone in this case was effective in treating the patient's DH.

Our case highlights that the pleomorphic clinical appearance of DH can present a challenge in the diagnosis of DH. Awareness of its clinical

variability including vasculitis-like changes can help avoid misdiagnosis and delay in appropriate treatment.

REFERENCES

1. Hull CM, Zone JJ. Dermatitis herpetiformis and linear IgA bullous dermatosis. In: Bologna JL, Jorizzo JL, Schaffer JV, eds. *Dermatology*. Philadelphia: Elsevier Saunders; 2012:491-500.
2. Bolotin D, Petronic-rosic V. Dermatitis herpetiformis. Part I. Epidemiology, pathogenesis, and clinical presentation. *J Am Acad Dermatol*. 2011;64(6):1017-1024.
3. Tu H, Parmentier L, Stieger M, et al. Acral purpura as leading clinical manifestation of dermatitis herpetiformis: report of two adult cases with a review of the literature. *Dermatology*. 2013;227(1):1-4.
4. Zaghi D, Witheiler D, Menter AM. Petechial eruption on fingers. Dermatitis herpetiformis. *JAMA Dermatol*. 2014; 150(12):1353-1354.
5. Naylor E, Atwater A, Selim MA, Hall R, Puri PK. Leukocytoclastic vasculitis as the presenting feature of dermatitis herpetiformis. *Arch Dermatol*. 2011;147(11):1313-1316.
6. Carlson JA, Chen KR. Cutaneous pseudovasculitis. *Am J Dermatopathol*. 2007;29(1):44-55.
7. Carlson JA. The histological assessment of cutaneous vasculitis. *Histopathology*. 2010;56(1):3-23.
8. Bologna JL, Jorizzo JL, Schaffer JV. *Dermatology*. 3rd ed. Philadelphia: Elsevier; 2012.
9. Aftab MN, Dee A, Helm TN. Erythema elevatum diutinum arising in the setting of dermatitis herpetiformis. *Cutis*. 2006; 78(2):129-132.
10. Chandrasekaran SS, Rai R, Vedachalam S, Dorairaj L, Palaniraman S. Erythema elevatum diutinum in association with dermatitis herpetiformis. *Indian Dermatol Online J*. 2014; 5(1):48-50.
11. Farella V, Lotti T, Difonzo EM, Panconesi EX. Erythema elevatum diutinum. *Int J Dermatol*. 1994;33(9):638-640.