BRIEF ARTICLES

A Rare Case Of Dermatitis Herpetiformis Presenting As Fingertip Petechiae

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ABSTRACT

Dermatitis herpetiformis (DH) is an uncommon inflammatory autoimmune disease that most commonly presents as a pruritic, papulovesicular eruption in young children and adolescents. It follows a chronic and relapsing course and usually involves extensor surfaces of the elbows, forearms, buttocks, and knees and can also involve the scalp. DH is usually accompanied by gluten-sensitive enteropathy (celiac disease). In most patients with DH, the enteropathy is asymptomatic. DH is usually a life-long condition that requires continued treatment, including dapsone and elimination of gluten from the diet. We describe a rare case of a patient who presented with fingertip petechiae as the only initial manifestation of DH. DH should be considered in the differential diagnosis of petechiae of the fingertips, even if it is the only presenting sign.

INTRODUCTION

Dermatitis herpetiformis (DH) is an infrequent inflammatory autoimmune cutaneous disorder usually accompanied by gluten-sensitive enteropathy (celiac disease [CD]). Patients with DH commonly present with pruritic, polymorphic lesions, primarily consisting of grouped erythematous papules and urticarial plaques with vesicles or blisters symmetrically distributed on the extensor surfaces of the elbows, forearms, buttocks, and knees, and sometimes involving the scalp. We describe a rare case of a patient presenting with fingertip petechiae as the only initial manifestation of DH. Only six cases of acral petechial lesions as the sole initial presenting sign of DH in adult patients have been described in the literature.[1-6] Because DH is often the first presenting sign of CD, dermatologists should be aware of this unusual clinical presentation.

CASE REPORT

A 36-year-old well-developed, well-nourished woman presented with a 4-month history of recurrent flares of painful petechiae involving several fingertips. The lesions tended to follow along dermatoglyphic lines (Figure 1A). The remainder of her skin examination

was initially unremarkable, including normal nail units. The patient's remaining physical examination and review of systems were unrevealing. Her extensive laboratory evaluation, including coagulation studies, complete blood count, serum cryoglobulin, plasma cryofibrinogen, and transthoracic echocardiogram, was negative.

Punch biopsy of a petechial lesion on her thumb (Figure 1B and 1C) demonstrated infiltration of neutrophils within the dermal papillae and subepidermal clefting. There was also a perivascular mixed inflammatory cell infiltrate with rare nuclear dusting and few extravasated erythrocytes in the superficial dermis. There was no evidence of vasculitis. Acid-fast bacilli, Fite, Gram, and periodic acid-Schiff stains were negative for mycobacterial, bacterial, or fungal organisms. A perilesional direct immunofluorescence study (Figure 1D) demonstrated granular deposition of Immunoglobulin A (IgA) in the dermal papillae and at the basement membrane zone. Some deposition of IgA was also found surrounding the superficial blood vessels. No deposition of other immunoreactants was found within the epidermis, basement membrane zone, vessels, or dermis. The histologic and immunofluorescence findings were consistent with DH.

The patient subsequently underwent additional testing for antibody markers and genetic testing for CD. Autoantibodies to endomysium, tissue transglutaminase, and deaminated gliadin were not present. Furthermore, the patient demonstrated no heterodimers for HLA-DQ2 or HLA-DQ8. Nevertheless, given the confirmed diagnosis of DH and its close association with CD, the patient underwent esophagogastroduodenoscopy. Duodenal biopsies showed minimal villous blunting with increased intraepithelial lymphocytes within villous tips, consistent with CD.

Six weeks after her initial presentation, the patient presented with grouped pruritic vesiculopapules on both elbows and forearms (Figure 2A). She also presented with erythematous and crusted papules along the sides of her fingers (Figure 2B). Another biopsy sample obtained from the right forearm showed collections of neutrophils demonstrating slight nuclear dusting within dermal papillae, also consistent with DH (Figure 2C).

The patient was started on a gluten-free diet, which has kept her cutaneous symptoms under good control. She declined the standard dapsone regimen treatment but opted for small doses of dapsone when flare-ups occurred.

Figure 1. Dermatitis herpetiformis. Initial presentation. Clinical findings – (A) Fingertip petechiae along dermatoglyphic lines; (B) Light microscopy of a lesional punch biopsy specimen - Subepidermal vesiculation and collections of neutrophils within dermal papillae (40x); (C) Perivascular mixed inflammatory cell infiltrate including neutrophils and rare nuclear dusting. Numerous extravasated erythrocytes (100x); (D) Perilesional direct immunofluorescence study - granular deposition of IgA in dermal papillae and at the basement membrane zone. Deposition of IgA about superficial blood vessels (100x).

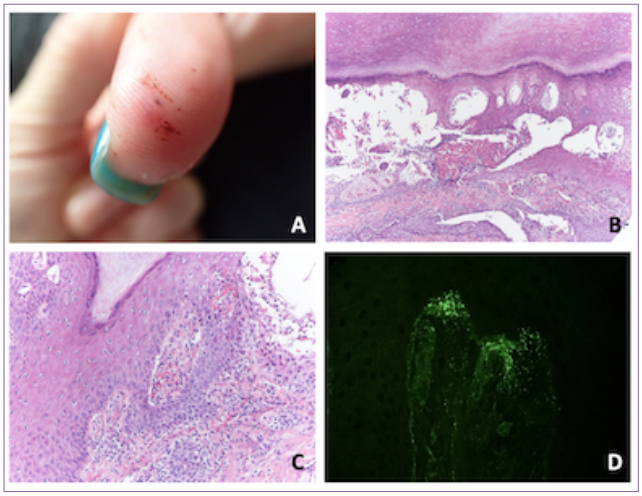
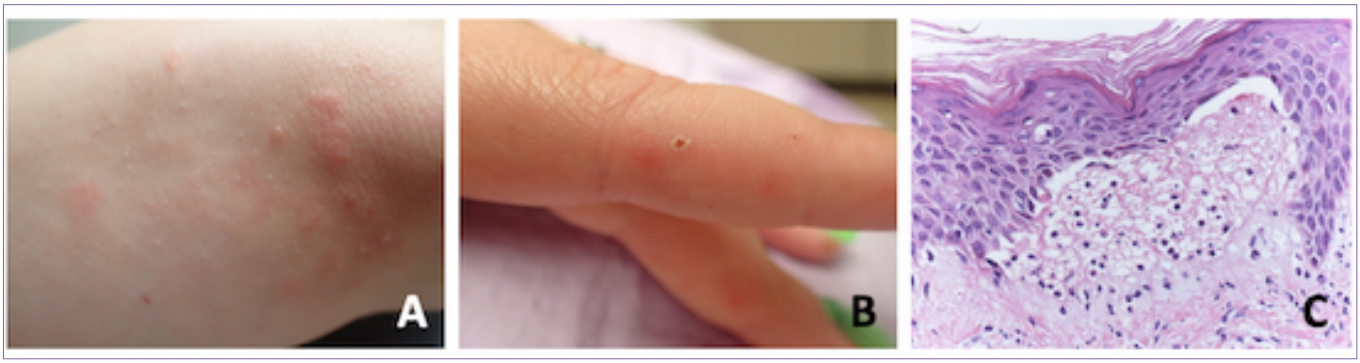


Figure 2. Dermatitis herpetiformis. Six weeks after initial presentation. (A) Grouped pruritic vesiculopapules on bilateral elbows and forearms; (B) Erythematous and crusted papules along the sides of the fingers; (C) Light microscopy of a specimen obtained from the right forearm - collections of neutrophils demonstrating slight nuclear dusting within dermal papillae (200x).



DISCUSSION

DH is an autoimmune vesiculobullous disorder that classically presents with intensely pruritic grouped erythematous papules and urticarial plaques with vesicles or blisters. Often the initial lesions turn into erosions and excoriations due to chronic pruritus. Typically, the lesions are symmetrically distributed on the extensor surfaces of the elbows, knees, shoulders, midline of back, buttocks, and sacral region, although they can also involve the face, scalp, nuchal area, and groin.[7] Petechiae or purpuric macules on the fingers or palmoplantar surfaces are uncommon presentations of DH and are more likely to occur in children.[7] Only six adult cases of DH presenting with acral petechiae or purpura as the sole initial presenting sign have been described in the literature.[1-6] In our case, the initial and only manifestation was petechiae involving the fingertips. More classic-appearing lesions of DH developed in our patient's elbows and forearms at the 6-week follow-up visit.

DH is histologically characterized by collections of neutrophils in the dermal papillae with subepidermal blister formation. On direct immunofluorescence, there are granular depositions of IgA within the dermal papillae and/or along the dermo-epidermal junction. These histologic and immunofluorescence findings were also observed in cases presenting with acral petechiae or purpuric lesions, with some cases demonstrating extravasated red blood cells in the upper dermis.[1,3,8-10] While one case presented with leukocytoclastic vasculitis on light microscopic examination, no deposition of immunoreactants was found along the blood vessels on direct immunofluorescence.[11] In our case, numerous extravasated erythrocytes were found in the papillary dermis that accounted for the petechial appearance. While some IgA deposition was noted along the small blood vessels on direct immunofluorescence in our case, it was interpreted as secondary to DH rather than to a vasculitic process, as no light microscopic evidence of vasculitis was detected and the patient soon proceeded to present with classic, non-purpuric lesions of DH.

DH is usually accompanied by CD, an autoimmune response against gliadin, tissue transglutaminase, and epidermal transglutaminase induced by gluten ingestion.[4] The typical presentation of CD is characterized by gastrointestinal signs and symptoms, and typical diagnostic testing

includes serologic screening (e.g., anti-tissue transglutaminase and anti-endomysial IgA antibodies) and genetic testing for human leukocyte antigens (HLA-DQ2 and HLA-DQ8) that are strongly associated with CD. When extraintestinal manifestations of CD exist, as occurs in patients with DH, minimal or absent gastrointestinal signs/symptoms may occur,[12] such as in our patient. This case had an atypical presentation of DH; she lacked clinical signs/symptoms of CD and tested negatively for screening serologies and for the HLA heterodimers, HLA-DQ2, and HLA-DQ8. Without the confirmation of her DH diagnosis by histologic and immunofluorescence evaluation, the patient probably would not have undergone a small bowel biopsy, and detection of her underlying CD would likely have been delayed.

The diagnosis of DH can be difficult without the classic presentation. A rapid diagnosis of DH can vastly improve the quality of life of patients with undiagnosed CD, because gastrointestinal and cutaneous symptoms can be resolved with a gluten-free diet. This case strongly emphasizes that DH should be included in the differential diagnosis of petechial involvement of the hands and fingers.

KEYWORDS: dermatitis herpetiformis, celiac disease, gluten-sensitive enteropathy; autoimmune disease, direct immunofluorescence.

ABBREVIATIONS: DH - Dermatitis herpetiformis; CD - Celiac disease; IgA - Immunoglobulin A

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