

# Acral petechial eruptions without gastrointestinal symptoms: Three cases of dermatitis herpetiformis



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## INTRODUCTION

Dermatitis herpetiformis classically presents with intensely pruritic vesiculopapules on the extensor surfaces and is associated with gluten sensitivity and celiac disease. Acral petechiae and purpura are an uncommon but important presentation of dermatitis herpetiformis that can help guide the astute diagnostician, especially because they may be the only sign of gluten sensitivity. We report 3 cases of dermatitis herpetiformis presenting with acral petechiae in the absence of gastrointestinal symptoms.

## REPORT OF 3 CASES

### Patient 1

A 48-year-old white man with a history of mild plaque psoriasis presented with petechiae that had been present for 12 months on the bilateral aspect of his index fingers and 1 palm. He endorsed occasional pruritus unresponsive to topical steroids. He denied trauma, preceding vesicles, or pustules. A previous evaluation result for celiac disease with bowel biopsy was inconclusive. He consumed gluten and denied bloating, abdominal pain, or abnormal stooling. He was not receiving treatment for psoriasis, which had previously been controlled with topicals.

On examination, there were numerous petechiae scattered on the lateral aspect of the fingers of both hands, with a few on the left palm (Fig 1, A). The result for the remainder of his skin examination was

unremarkable except for xerosis with faint erythema on the bilateral aspect of the elbows.

Tissue transglutaminase 3 IgG and IgA levels were elevated (IgG 20 U/mL [normal <5 U/mL]; IgA >100 U/mL [normal <3 U/mL]). He elected to pursue a gluten-free diet trial in lieu of biopsies and reported that the petechiae resolved after 2 weeks of gluten avoidance.

### Patient 2

A 31-year-old white man with a history of atopic dermatitis presented with a recurrent, pruritic rash on the arms and legs since adolescence. His medical history was notable for 3 hospitalizations for pneumonia. He denied gastrointestinal symptoms.

On examination, 1- to 3-mm skin-colored to yellow-pink vesicles on an erythematous base were clustered and configured individually on the elbows, knees, posterior portion of the neck, and scalp. He also had asymptomatic, linear petechiae following the dermatoglyphs on the fingers (Fig 1, B), which had appeared without preceding hemorrhagic vesicles or pustules.

A punch biopsy and direct immunofluorescence of intact vesicles from the neck revealed papillary microabscesses on hematoxylin-eosin staining and a negative direct immunofluorescence result. Tissue transglutaminase 3 IgA level was elevated (>40 U/mL). Skin lesions resolved with a gluten-free diet and dapsone, but recurred a day after

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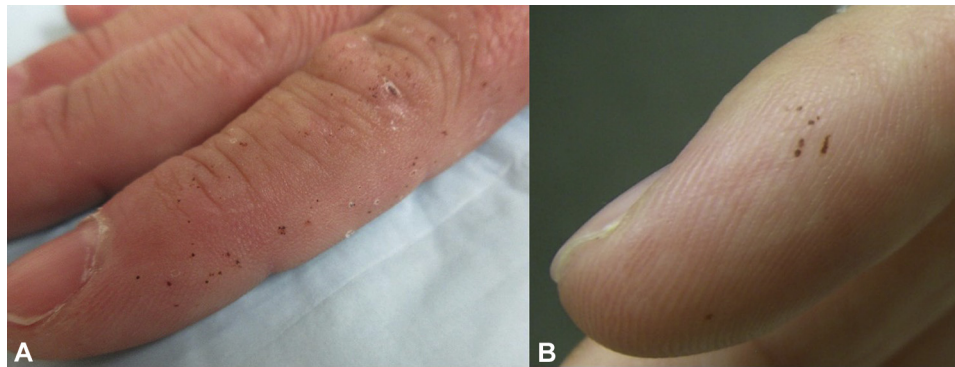
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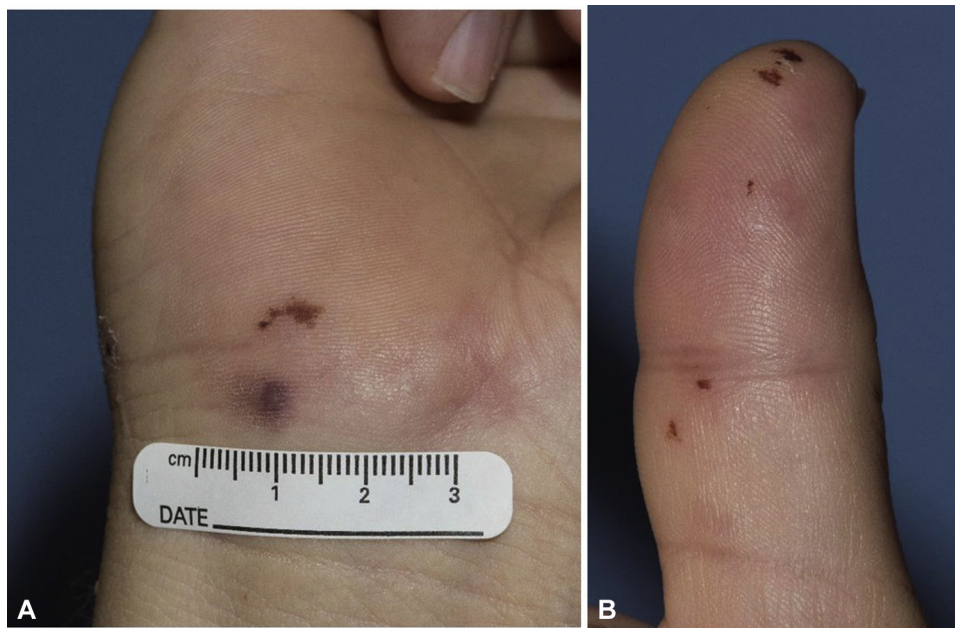
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**Fig 1.** Acral petechiae on the lateral aspect of the right index finger were present for approximately 1 year in patient 1 (A). Petechiae following dermatoglyphs on the lateral aspect of the right index finger in patient 2 (B).



**Fig 2.** Arcuate, red and brown macules superior to a violaceous, dermal papule on the right palm and volar aspect of the wrist (A) in patient 3. The same patient had petechiae on the thumb, with 2 superior lesions tracking along dermatoglyphs (B).

self-challenging with gluten. A small-bowel biopsy confirmed celiac disease.

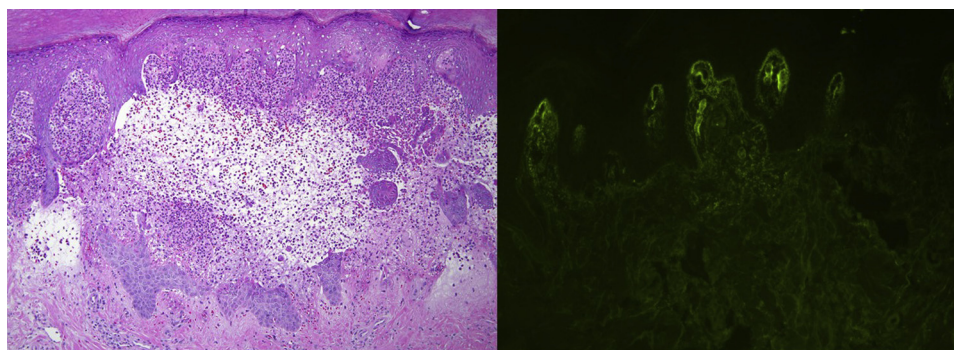
### Patient 3

A 60-year-old white man with history of type 2 diabetes was referred for a recurrent, purpuric eruption with oral ulcers and night sweats for the past year after negative infectious, allergic, hematologic, and rheumatologic evaluation results. Painful, purpuric lesions began on the hands and progressed to involve the elbows and feet; a few 2- to 4-mm ulcers affected the tongue and buccal portion of the cheeks. Results for previous skin biopsies of the fingers were not diagnostic and direct immunofluorescence results had been nondiagnostic.

Physical examination was notable for petechiae and deep-seated hemorrhagic papules on the fingers, palms, wrists, and suprapubic skin (Fig 2). Additionally, he had a discrete 3-mm tongue ulcer. Skin biopsy and direct immunofluorescence results from the hand were characteristic of dermatitis herpetiformis (Fig 3). He began receiving dapsone and a gluten-free diet, with resolution of his symptoms in 5 weeks. Subsequent gastrointestinal testing did not show evidence of celiac disease.

### DISCUSSION

Petechiae and purpura as primary acral lesions are a recognized cutaneous manifestation in dermatitis herpetiformis and celiac disease, but may be



**Fig 3.** Punch biopsy from the finger of patient 3 demonstrated dense neutrophilic aggregates in the dermal papillae, superficial dermal edema, and dermal hemorrhage on hematoxylin-eosin staining (left). Direct immunofluorescence demonstrated granular IgA deposits in the dermal papillae, consistent with dermatitis herpetiformis (right).

**Table I.** Differential diagnosis for primary acral purpura and petechiae

Category	Cause	Evaluation	Further considerations
Inflammatory	Dermatitis herpetiformis	Skin biopsy, DIF Anti-tTG3 antibody Antiendomysial antibody	Usually personal or family history of gluten intolerance or celiac disease
Vasculitis	Small vessel Leukocytoclastic vasculitis IgA vasculitis Medium vessel Anti-ANCA antibody Connective tissue related (RA, SLE, DM) Polyarteritis nodosa Mixed cryoglobulinemia	Skin biopsy, DIF ANA Rheumatoid factor CBC CMP Urinalysis Hepatitis panel ASO Cryoglobulins	Primary hypercoagulable states,* thrombocytopenia, thrombocytosis, and calciphylaxis can also cause acral purpura. These may present as widely distributed or retiform purpura.
Embolic	Bacterial endocarditis Atherosclerosis Postprocedural cholesterol emboli Left-sided atrial myxoma	Skin biopsy ECHO CBC, CMP Blood cultures	Admit for endocarditis evaluation Ophthalmologic examination
Infectious	Gonococcemia Meningococcemia Sepsis Rocky Mountain spotted fever Atypical hand, foot, and mouth disease (A6)	Skin biopsy Urine and blood cultures CBC, CMP Coagulation panel Lumbar puncture RMSF IFA <sup>†</sup>	Admit for IV antibiotics supportive care
Trauma	Trauma	Thorough history Full skin examination	Chemotherapy, thrombocytopenia, anticoagulant use can predispose

ANA, Anti-nuclear antibody; ANCA, anti-neutrophil cytoplasmic antibody; ASO, antistreptolysin O; CBC, complete blood count; CMP, complete metabolic panel; DIF, direct immunofluorescence; DM, dermatomyositis; ECHO, echocardiogram; IV, intravenous; RA, rheumatoid arthritis; RMSF IFA, Rocky Mountain spotted fever indirect immunofluorescence; SLE, systemic lupus erythematosus; tTG3, tissue transglutaminase 3. \*Primary hypercoagulable states include factor V Leiden, prothrombin gene mutation; protein C/S, antithrombin III deficiency; and antiphospholipid antibody, hyperhomocysteinemia.

<sup>†</sup>Rocky Mountain spotted fever indirect immunofluorescence result is typically not positive until antibodies develop weeks after the onset of symptoms, so empirical treatment initiation is based on clinical diagnosis.

overlooked in the absence of classic pruritic, vesiculopapular lesions on extensor surfaces and in individuals without overt gastrointestinal symptoms. Acral petechiae in dermatitis herpetiformis have been reported several times but are likely underreported; the differential diagnosis for acral

purpura is broad (Table I).<sup>1-5</sup> Petechiae may be the sole presenting sign or may be present in conjunction with classic dermatitis herpetiformis skin findings.<sup>6</sup> Because acral stratum corneum is thick, edema and hemorrhage become trapped in the papillary dermis, forming “dots” instead of the

more common, excoriated erosions on nonacral skin. Petechiae may appear linear or curvilinear as they follow dermatoglyphics on acral skin. Linear petechiae may be referred to as vibices.

Dermatitis herpetiformis can be diagnosed with hematoxylin-eosin–stained skin biopsy, which demonstrates collections of neutrophils in the dermal papillae; and direct immunofluorescence, which demonstrates granular IgA deposition in the papillary dermis. In equivocal cases, detection of elevated serum tissue levels of transglutaminase 3 IgA or antiendomysial antibodies, or genetic testing for the presence of *HLA-DQ2* and *HLA-DQ8*, can help confirm diagnostic suspicion.<sup>7</sup> Biopsy site selection is important: punch biopsy of an acute vesicle is best for capturing intact papillary microabscesses on hematoxylin-eosin staining, whereas normal-appearing perilesional skin has a higher yield for direct immunofluorescence.<sup>8</sup>

A gluten-free diet is essential for symptom management and decreasing the long-term risk for mucosal-associated T-cell lymphoma. Dapsone is first-line treatment for dermatitis herpetiformis, but will not decrease malignancy risk or improve celiac disease. Before initiation of dapsone, patients need to be screened for glucose-6-phosphate dehydrogenase deficiency, complete blood cell count, and basic metabolic panel and counseled on the risk of agranulocytosis, methemoglobinemia, and peripheral motor neuropathy. Compliance with a gluten-free diet can be monitored by following tissue

transglutaminase 3 IgA titers (normal <5 U/mL), the level of which will decrease with gluten avoidance. Physicians should consider vaccinating patients with a new diagnosis of dermatitis herpetiformis with the pneumococcal vaccine because there is evidence of increased risk of bacterial pneumonia, especially in proximity to their diagnosis.<sup>9</sup>

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