Dermatitis herpetiformis presenting as intertriginous dermatitis

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Case report

A 54-year-old man with atypical dermatitis herpetiformis Duhring lasting about three years is reported. The lesions were located exclusively in the pubic area, groins, inner thighs and perianal regions. Routine histopathology and the direct as well as the indirect immunofluorescence supported the diagnosis. Application of a gluten-free diet and dapsone alleviated the symptoms.
Figure 1. Erythematous plaque rimmed with papules and erosions covering a) the pubic area, groins, inner thighs, and b) perianal region.

Figure 2. Biopsy specimen from the periphery of the plaque showing a) perivascular lymphocytic infiltrate intermingled with neutrophils and few eosinophils. Note collection of neutrophils at the tip of papillary dermis (original magnification H&E, x100); b) a detail, eosinophilic spongiosis within overlying epidermis (original magnification H&E, x1000, immersion).

Figure 3. Linear distributed fine granular IgA deposits along the basement membrane zone with accentuation at the tips of dermal papillae (direct immunofluorescence, anti-human IgA-FITC conjugate, original magnification x 200).
measures. Otherwise he was healthy with no apparent gastrointestinal disturbances. Laboratory work-up was normal with only marginally elevated fasting serum glucose (6.9 mmol/L). Considering the long history of the eruption together with the poor response to topical antymycotics and corticosteroids, differential diagnoses included autoimmune bullous diseases and benign familial pemphigus. A biopsy was taken from the margin of a plaque situated on the thigh including clinically uninvolved skin. The specimen was processed for routine histology and direct immunofluorescence (DIF) testing. HE stained skin sections revealed perivascular lymphocytic infiltrate with neutrophils and sparse eosinophils, edema of the papillary dermis with extravasated erythrocytes and fibrin as well as a few foci of eosinophilic spongiosis (Figure 2). A serum sample was screened for a panel of autoantibodies by indirect immunofluorescence (IIF) on several tissue substrates. DIF revealed finely granular IgA deposits along the basement membrane with marked accumulation at the tips of dermal papillae (Figure 3). Antiendomysial antibodies of the IgA isotype were found in the serum (titer 1:40, substrate: monkey esophagus). The diagnosis of dermatitis herpetiformis was confirmed and the patient was put on a dapsone and gluten-free diet.

**Discussion**

Despite the fact that almost a quarter of patients with celiac disease might be affected with dermatitis herpetiformis, the vast majority of adult patients with Duhring’s disease do not have gastrointestinal symptoms (3). When clinical presentation of dermatitis herpetiformis is atypical, such a diagnosis may be hard to entertain. Although classical textbooks, monographs and review papers (1, 2, 4) mention non-standard sites of affection such as the face and inguinal folds, we were not able to retrieve a single case description of dermatitis herpetiformis confined exclusively to an atypical localization. In our patient, extension of the eruption to the buttocks perianally may be considered typical, but the continuum of the affected area with the inner thigh, groin and pubic lesions made it an unusually extensive sole location for Duhring disease. Eosinophilic spongiosis is a histologic pattern seen in numerous skin diseases although most commonly described in autoimmune bullous disorders (5). Among them, pemphigus, and especially its herpetiformis variety, has frequently been linked to this histologic pattern. Though eosinophilic spongiosis has only rarely been described in the setting of dermatitis herpetiformis (no modern dermatology textbook mentions it within the description of the histopathology), in an earlier report Pasricha observed eosinophilic spongiosis in 3 out of 6 (50%) patients with Duhring disease (6).

Any long-standing itchy, inflammatory and vesicular/erosive eruption in skin folds that is non-responsive to topical antifungals and corticosteroids should include dermatitis herpetiformis in its differential diagnostic considerations and be accompanied by a prompt skin biopsy with appropriate histopathologic and immunofluorescence analysis.

**References**


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