Dermatitis herpetiformis presenting as intertriginous dermatitis

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SUMMARY

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.

KEY WORDS

dermatitis herpetiformis, atypical presentation, eosinophilic spongiosis, intertrigo-like

Dermatitis herpetiformis is a chronic blistering disease, and represents one of the extraintestinal manifestations of celiac disease, and shares a similar genetic background and pathophysiology (1). Typically, lesions are symmetrically distributed over extensor surfaces of extremities, shoulders, sacral areas, buttocks and the nuqual plane. Less frequently, they may appear on the face, scalp and groin (2). When the lesions are confined solely to atypical locations it might cause a significant delay in the diagnosis of dermatitis herpetiformis. Here we describe a patient whose disease had been localized to the groin for more than 3 years.

Case report

A 54-year-old man presented with a long-standing itchy and burning eruption in the groin and inner thigh region. The lesions had first appeared three years earlier as small reddish papules and plaques accompanied by intense pruritus. Gradually, they coalesced to form large plaques in the groin, inner thigh and buttocks. Superficial erosions caused a burning sensation. The lesions never disappeared completely and got worse during the hot summer months. His condition was treated with topical antifungals and occasionally with topical corticosteroids, but apart from a minimal alleviation of symptoms these drugs were not effective. On examination, erythematous plaques were observed in the groin, the inner thighs and buttocks, and especially perianally (Figure 1). Oval erosions were noted throughout the plaques but were more numerous at the edges. The rest of his skin was unaffected. He complained about severe episodic pruritus and burning. He was treated for mild type II diabetes mellitus with dietary

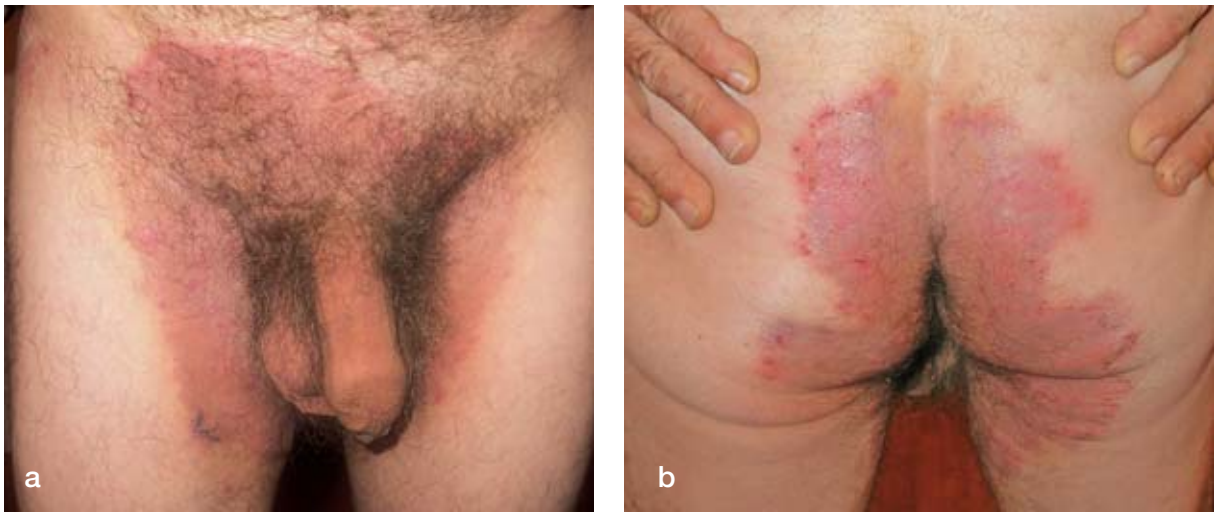

Figure 1. Erythematous plaque rimmed with papules and erosions covering a) the pubic area, groins, inner thighs, and b) perianal region.

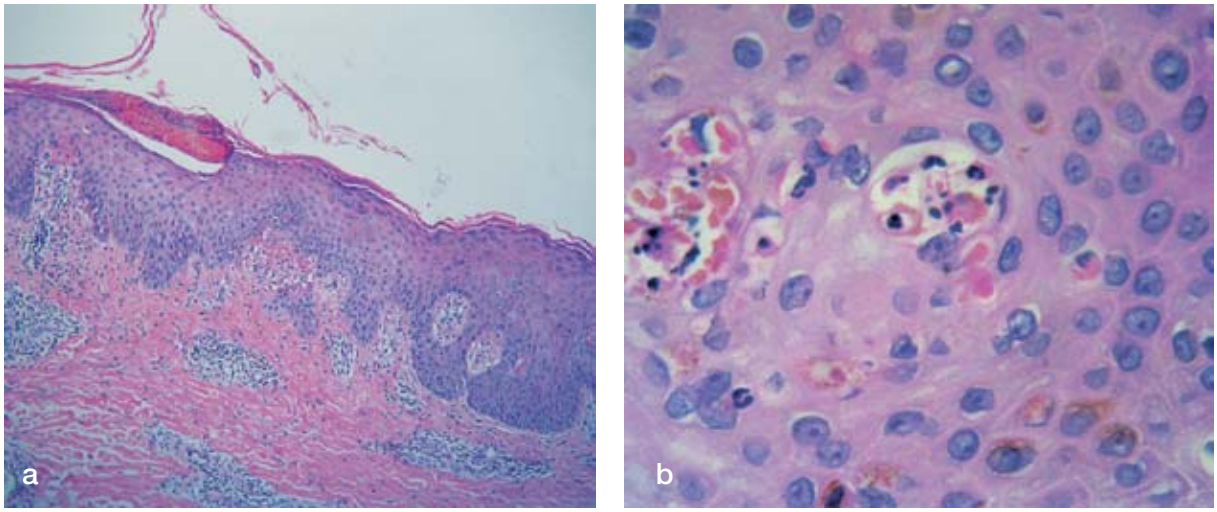

Figure 2. Biopsy specimen from the periphery of the plaque showing a) perivascular lymphocytic infiltrate intermingled with neutrophils and few eosinophils. Note collection of neutrophils at the tip of papillary dermis (original magnification H&E, x100); b) a detail, eosinophilic spongiosis within overlying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

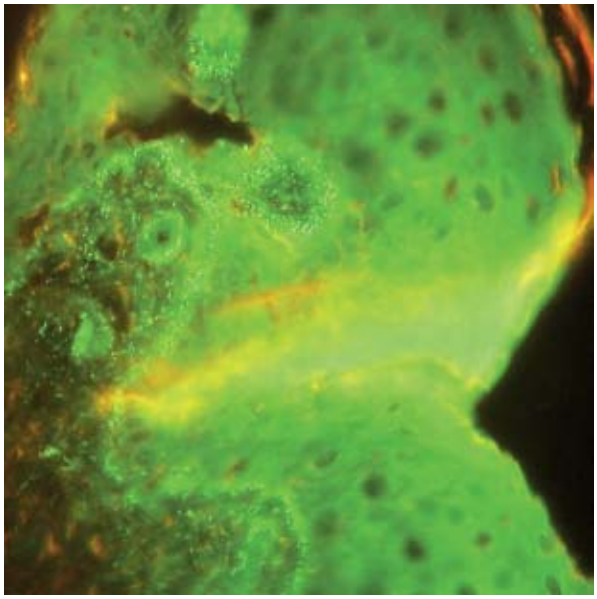

Figure 3. Linear distributed fine granular IgA deposits along the basement membrane zone with accentuation at the tips of dermal papillae (direct immunofluorescence, anti-human IgA-FITC conjugate, original magnification x 200).

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 antimycotics 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 papilae (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

1. Herron MD, Zone JJ. Dermatitis herpetiformis and linear IgA bullous dermatosis. In: Bologna JL, Jorizzo JL, Rapini RP, eds. Dermatology. Mosby, London, 2003, 479-89.

2. Nicholas MEO, Krause PK, Gibson LE, Murrey JA. Dermatitis herpetiformis. Int J Dermatol 2003; 42: 588-600.

3. Collin P, Reunala T, Rasmussen M, Kyronpalo S, Pehkonen E, Laippala P, Maki M. High incidence and prevalence of adult coeliac disease. Augmented diagnostic approach. Scand J Gastroenterol 1997; 32: 1129-33.

4. Bagheri B, Hall RP. Dermatitis herpetiformis. In: Jordon RE. Atlas of bullous disease. Churchill Livingstone: New York, 2000, 99-108.

5. Machado-Pinto J, McCalmont TH, Golitz LE. Eosinophilic and neutrophilic spongiosis: clues to the diagnosis of immunobullous diseases and other inflammatory disorders. Semin Cutan Med Surg 1996; 15: 308-16.

6. Pasricha JS. Eosinophilic spongiosis in vesiculo-bullous diseases. Indian Journal of Dermatology, Venereology and Leprology 1988; 54: 196-8.

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