Blistering Skin Disease Associated with Insulin-dependent Diabetes Mellitus: A Quiz

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A 42-year-old male with life-long well-regulated insulin-dependent diabetes mellitus and no other known co-morbidities was referred to the Department of Dermatology. He had suffered from recurrent papulo-vesicular eruptions for 16 years and had earlier being diagnosed as lichen simplex chronicus.

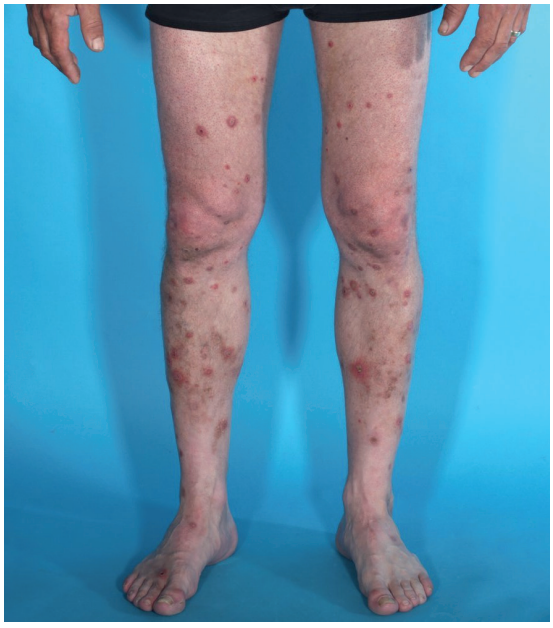

Fig. 1. Clinical picture of the patient`s legs. The patient presented with excoriated papulo-nodules.

Medical examination revealed small blisters, inflamed erythematous skin with perilesional oedema as well as erosions and crusts primarily on the dorsal extremities (Fig. 1). Histology showed subepidermal blistering, inflammatory infiltrate with eosinophils and neutrophils in dermal papillae (Fig. 2). Direct immunofluorescence showed deposition of IgA immunoglobulin in dermal papillae.

What is your diagnosis? See page 17 for answer.

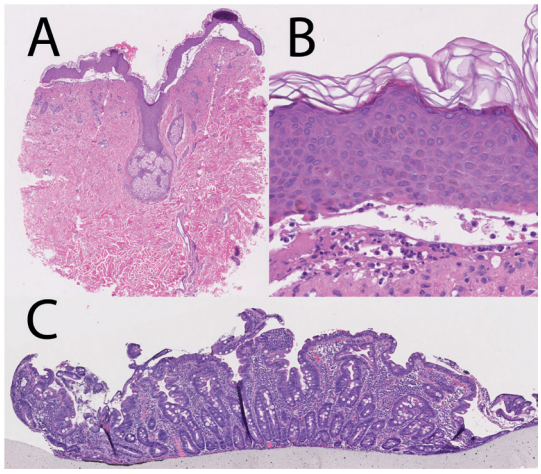

Fig. 2. Skin biopsy with subepidermal blister (A). The blister contains, fibrin, neutrophils and few eosinophils (B). Biopsy from doudenum with villous atrophy (C).

Nodular Eroded Lesions in the Diaper Area of a 16-month-old Girl: A Quiz

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A 16-month-old girl was referred to the Department of Dermatology because of a 6-month history of eroded papular rash in the diaper area (Fig. 1). The condition had been interpreted as diaper dermatitis, and various topical treatments had been used: antifungal agents, moderate-potent topical corticosteroids, fusidic acid and aciclovir. None of them gave significant improvement. The parents had also tried Aloe vera gel on their own initiative. The child was otherwise healthy and not predisposed to any dermatologic diseases.

What is your diagnosis? See next page for answer.

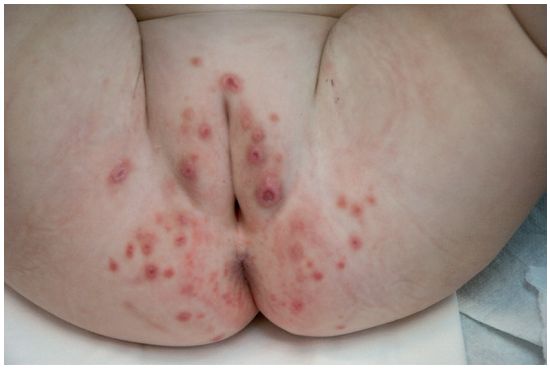

Fig. 1. A 16-month-old girl with nodular erosive lesions in the diaper area.

Answers to Quiz

Blistering skin disease associated with insulin-dependent diabetes mellitus:

Diagnosis: **Dermatitis herpetiformis**

The patient had been seen for the first time 16 years earlier, when he was referred to our department with a year-long history of recurrent pruritic vesiculo-papular skin eruptions. He had no gastrointestinal complaints. The medical examination revealed chronic excoriations and ulcers on hyperpigmented skin on the face and dorsal lower extremities. Lesional skin punch biopsies for light microscopy showed superficial lymphocytic dermatitis and for direct immunofluorescence showed no immunoglobulin deposition. A diagnosis of lichen simplex chronicus was suggested, and he was prescribed a potent topical steroid and discharged. Six years later he was re-referred presenting with unchanged symptoms on his face and extremities. The medical examination showed excoriations. No skin biopsy was performed. The patient was discharged with the diagnosis lichen simplex chronicus, after one consultation.

Eight years later, the patient was re-referred with unchanged symptomatology. Medical examination revealed small blisters, inflammed skin with slight oedema, erosions, and crusts primarily on the dorsal extremities. The diagnoses bullous pemphigoid or diabetic bullae were suggested. New perilesional skin biopsies from non-excoriated fresh lesions were performed. Histology showed subepidermal blistering, inflammatory eosinophils and neutrophils in dermal papillae. Direct immunofluorescence showed deposition of IgA immunoglobulin in dermal papillae. Dermatitis herpetiformis was diagnosed. Even though the patient had no gastrointestinal symptoms, coeliac disease was suspected, and he was referred to the Department of Gastrointestinal Medicine where a duodenal biopsy revealed villus atrophy, and a blood sample showed an increased level of plasma transglutaminase antibodies (>80 au./l). Treatment with Dapsone 100 mg daily was initiated, and the patient's symptoms cleared within two weeks. He was also referred to a dietician for a gluten-free diet.

Dermatitis herpetiformis is a cutaneous manifestation of coeliac disease (1). It is more common in Northern Europe and USA with an estimated prevalence about 10 per 100,000 inhabitants (2–3). Although coeliac disease is more common in females, dermatitis herpetiformis seemingly has a higher prevalence in males (3–5). The common age at presentation is 30–40 years.

The most common clinical signs of dermatitis herpetiformis include intense pruritic papules, urticarial plaques and herpetiform blisters, preferably on the extensor surfaces (6). Erosions, hyperpigmentation, and excoriations may be present.

The histopathological features include subepidermal blisters with microabscesses predominantly infiltrated by neutro-

phils (7–8). Direct immunofluorescence microscopy from perilesional skin reveals granular deposition of IgA within the papillary dermis.

Dermatitis herpetiformis has a chronic course. Besides gluten-sensitive enteropathy, dermatitis herpetiformis can also be associated with other autoimmune diseases, especially thyroid disease and insulin-dependent diabetes mellitus.

Dapsone is the drug of choice and relieves pruritus within a few days. A strict gluten-free diet reduces intestinal inflammation and IgA deposition of the skin, eventually allowing patients to withdraw from Dapsone treatment. The course of dermatitis herpetiformis is unpredictable as the disease waxes and waines.

In conclusion, dermatitis herpetiformis should be considered in patients with a history of autoimmune disease and recurrent pruritic skin lesions. If the patient presents with chronic, excoriated skin lesions, we strongly suggest not to discharge the patient, but offer a new clinical examination when acute clinical findings are present.

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Nodular eroded lesions in the diaper area of a 16-month-old girl: Comment

Diagnosis: **Granuloma gluteale infantum**

This is a rare but re-emerging condition, related to the increasing use of reusable diapers (1). The literature on this topic is sparse, but the condition is described to be a complication to irritative diaper dermatitis. The toxic skin reaction can result from diaper occlusion, prolonged use of topical corticosteroids, irritant contact dermatitis and candidiasis (1–4). The condition presents overlapping features with Jacquet erosive diaper dermatitis, which is associated with prolonged exposure to urine and feces under occlusion and is more often seen in elderly people. In the presented case, long-term use of topical steroids combined with use of very dense reusable diapers (Fig. 2) might have been precipitating factors. To exclude other differential diagnoses, such as Langerhans cell histiocytosis, a biopsy was performed revealing mild epidermal hyperkeratosis. No signs of fungal or viral infections were detected. Despite the name, there are no granulomas in the skin lesions (4).

We advised to change treatment with topical corticosteroids to barrier creams and a pasta containing ichthammol and zinc oxide. The nodules resolved within three months.

We present this case to remind clinicians to consider the diagnosis granuloma gluteale infantum in patients with persistent diaper dermatitis.

Fig. 2. The diaper type that in this case had been used since birth with rice paper lining.

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