Chapter 17
A 50-Year-Old Man with Itchy, Polymorphic Lesions on the Scalp

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A 50-year-old man was admitted to the Department of Dermatology with a one-month history of widespread skin lesions with coexisting itch. The lesions started on his buttocks and then spread into the scalp, elbows, knees and forearms. No other family member was affected. The patient denied having any gastroenterological symptoms.

A physical examination revealed erosions with crusts and single vesicles on the occipital area of the scalp (Fig. 17.1). Moreover, erythematous areas with vesicles and numerous erosions with crusts were present on the elbows, forearms, buttocks and knees.

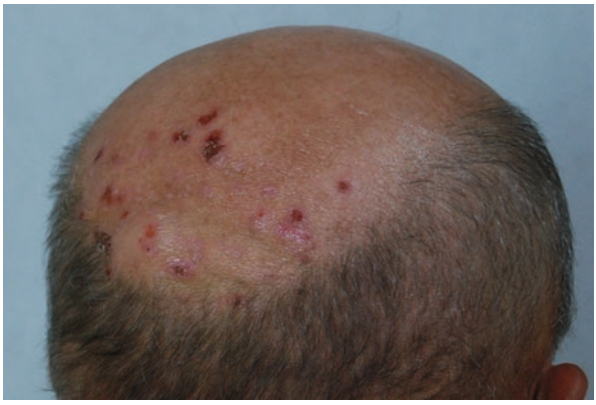

Fig. 17.1 A 50-year-old man with erosions covered by crusts and single vesicles on the occipital area

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Laboratory tests were normal. A direct immunofluorescence test from the perilesional skin of the buttock revealed granular IgA (++) and IgM (+) deposits in the dermal papillae. An indirect immunofluorescence showed the presence of IgA antiendomysial antibodies (IgA-EMA).

Based on the case description and the photographs, what is your diagnosis?

Differential Diagnoses

1. Pemphigus foliaceus.
2. Dermatitis herpetiformis.
3. Bullous pemphigoid.
4. Linear IgA dermatosis.

Diagnosis

Dermatitis herpetiformis.

Discussion

Dermatitis herpetiformis is a cutaneous manifestation of coeliac disease, in which gluten induces development of skin lesions in genetically susceptible individuals with the human leucocyte antigen (HLA) DQ2 or DQ8 haplotypes [1]. In both, celiac disease and dermatitis herpetiformis, the development of IgA autoantibodies against transglutaminases is observed. In the case of dermatitis herpetiformis, IgA autoantibodies are deposited in the superficial papillary dermis [2]. The disease is more common in men compared to women and typically occurs in the fourth decade of life [3]. In dermatitis herpetiformis, polymorphic skin lesions with the presence of vesicles, papules and macules with coexisted itch are observed. However, because of scratching, only erosions and crusts may be detected. The predilection sites for the dermatitis herpetiformis rash are the elbows, knees and buttocks [1]. Scalp involvement is observed in 30% of cases. Hair loss is rarely observed [2, 4]. 20% of patients with dermatitis herpetiformis exhibit gastrointestinal symptoms at time of initial diagnosis [3]. The diagnosis of dermatitis herpetiformis is based on typical clinical picture and direct immunofluorescence test. In direct immunofluorescence test, granular IgA in the papillary dermis are observed. Moreover, the presence of circulating anti-transglutaminase 2 antibodies supports the diagnosis, but their absence does not exclude dermatitis herpetiformis [1]. Therapy of dermatitis herpetiformis consists of a strict life-long gluten-free diet and sulphonamide drugs (dapsone). Moreover, short term use of potent topical corticosteroids may be helpful to decrease the itch [3].

Differential diagnoses for the presented patient included pemphigus foliaceous, bullous pemphigoid and linear IgA dermatosis.

Pemphigus foliaceus is a form of an autoimmune bullous disease affecting the skin. The disease most commonly affects women between 50 and 60 years of age [2]. Skin involvement is characterized by flaccid blisters and erosions localized mainly on the face, scalp, trunk and proximal extremities [5].

Bullous pemphigoid is the most frequent autoimmune bullous disease that mainly affects elderly individuals, usually above 70 years [6]. It presents as itchy, tense blisters over normal skin or over erythematous and edematous background [6]. Mucosal involvement is rarely reported (10–30% of the cases) [6].

Linear IgA bullous dermatosis is a subepidermal vesiculobullous disease that occurs in both adults and children. In children it presents as annular or polycyclic plaques and papules with blistering around the edges, primarily around the mouth and eyes, lower abdomen, thighs, buttocks, genitals, wrists and ankles. In contrast, the adult-onset is characterized by the lesions on the trunk, head and extremities [7].

Based on clinical features and immunofluorescence tests, the patient was diagnosed with dermatitis herpetiformis. Dapsone (100 mg daily) was initiated. Gluten-free diet and gastroenterological consultation was recommended.

Key Points

- Dermatitis herpetiformis presents as itchy, polymorphic skin lesions mainly localized on the elbows, knees and buttocks areas.
- Scalp involvement is observed in 30% of patients with dermatitis herpetiformis; hair loss is rarely observed.

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