

CHILDHOOD PEMPHIGUS FOLIACEUS. HERPETIFORM VARIANT.

Case report. R.C., an 8-year-old girl, presented erythematous, papulo-pomphoid, circinate lesions, mainly on the back (Fig. 1) from two months. On the peripheral border of the lesions there were intensely itchy, 2-4 mm in size erosions. These features led to diagnose an autoimmune bullous disorder, probably IgA linear dermatitis and to carry out laboratory examinations in order to specify the diagnosis. Anti-EMA and antitransglutaminase antibodies and the immunoenzymatic test for antidesmogleins 1 and 3 were negative. The histological examination showed superficial spongiosis with lymphocytic and eosinophilic exocytosis (Fig. 2). The direct immunofluorescence with IgG e C showing the pemphigus phenomenon (Fig. 3), and the indirect one with the same phenomenon at a titre 1/50 in absence of junctional and dermal deposits were a clue to the diagnosis of **pemphigus herpetiformis**. Prednisone 0.5 mg/kg/die was very effective.



Fig. 1

Clinical features. The herpetiform variant of pemphigus foliaceus is characterized, besides the lack of mucosal lesions, by intense itching, circinate, papulo-pomphoid lesions, vesicles and crusts, good response to oral corticosteroids and benign clinical course. In children pemphigus foliaceus is rare and the herpetiform variant exceptional (1, 5).

The **histological examination** (1, 3) shows superficial eosinophilic spongiosis. The immunofluorescence shows deposits of IgG e C3 in the intercellular space of epidermis.

The **differential diagnosis** should rule out dermatitis herpetiformis and IgA linear dermatitis (2, 4). The direct and indirect immunofluorescence, showing the typical intercellular deposits, is essential to rule out other disorders and confirm the diagnosis of pemphigus herpetiformis.

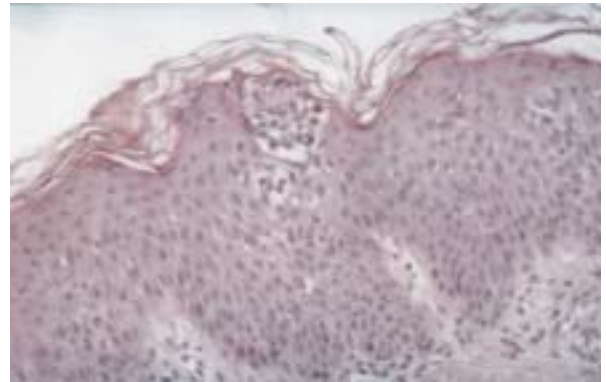


Fig. 2

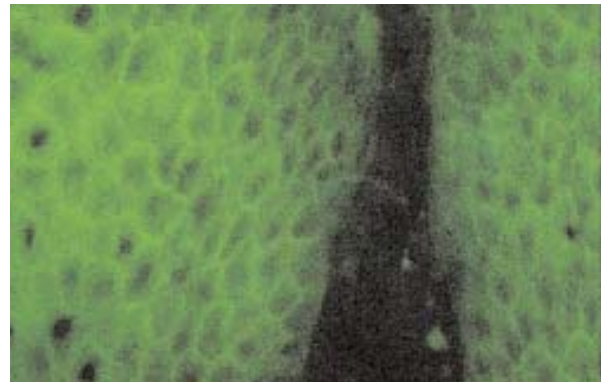


Fig. 3

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CHILDHOOD PEMPHIGUS FOLIACEUS. SEBORRHEIC VARIANT.

Case report. M. Serena, a 4-year-old little girl presented superficial bullous lesions, with clear-cut borders, in various evolution phase, mainly crusted, of the scalp, retroauricular region, trunk and around the mouth (Fig. 3). The eruption did not respond to antibiotics. The histological examination showed (Fig. 1, 2) superficial acantholysis with isolated keratinocytes on the surface of the epidermis. The direct immunofluorescence showed high intercellular and nuclear positivity in the epidermis and a slight positivity at the dermal epidermal junction both with IgG and C3. A plaster applied in site of biopsy on the right armpit (Fig. 3) showed a clear Nikolsky sign, confirming the diagnosis of **seborrheic pemphigus**.

A topical corticosteroid and antibiotic treatment improved significantly the disorder. No new blisters appeared 45 days later. The patient was free of disease in the subsequent 12 years.

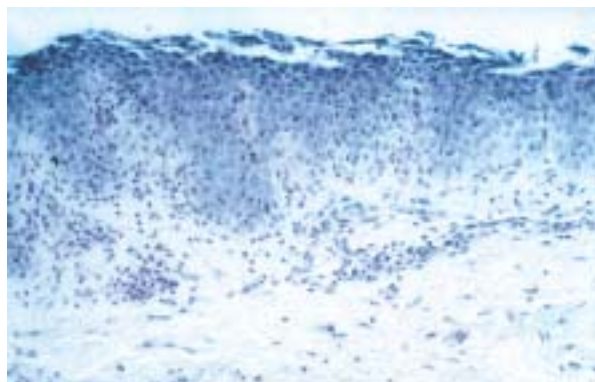


Fig. 1

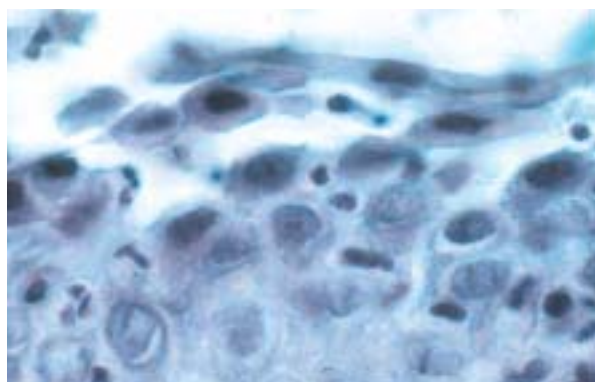


Fig. 2

Clinical features. The seborrheic variant of pemphigus foliaceus affects the seborrheic regions, mainly the scalp (1, 3), in absence of mucosal lesions. The erythematous variant is characterized by lesions on photoexposed areas and by IgG and C3 deposits both interkeratinocytic and at the dermal-epidermal junction. Overlap forms do exist as shown by our case.

Histologically, there is superficial acantholysis with isolated keratinocytes level with the granular layer (2). The **immunofluorescence** shows a high pemphigus phenomenon and, sometimes, lupus erythematosus-like findings (4).

The clinical **differential diagnosis** from impetigo and eczema is particularly difficult in children. The prognosis is usually benign and the disorder can be sometimes controlled by topical corticosteroids.



Fig. 3

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