

Eosinophilic Spongiosis in Early Pemphigus Foliaceus

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Abstract. A case of widespread non-specific, itchy exanthema in a 74-year-old female is presented. An early skin biopsy showed eosinophilic spongiosis. Later a dermatitis herpetiformis-like eruption developed. However, immunofluorescence microscopy showed intercellular IgG deposits characteristic of pemphigus. Recognition of eosinophilic spongiosis may be helpful in differentiating early pemphigus from other bullous diseases.

Key words: Pemphigus foliaceus; Dermatitis herpetiformis; Eosinophilic spongiosis

Pemphigus foliaceus in the early preacantholytic stage may present as dermatitis herpetiformis and the differential diagnosis may be difficult. Occurrence of epidermal spongiosis with infiltration of eosinophils in early pemphigus has sometimes been helpful in the diagnosis (1, 2, 3, 4). We here report a case presenting as dermatitis herpetiformis. A biopsy in the preacantholytic stage showed eosinophilic spongiosis. Later the demonstration of IgG deposits intercellularly by the immunofluorescence technique confirmed the diagnosis Pemphigus foliaceus.

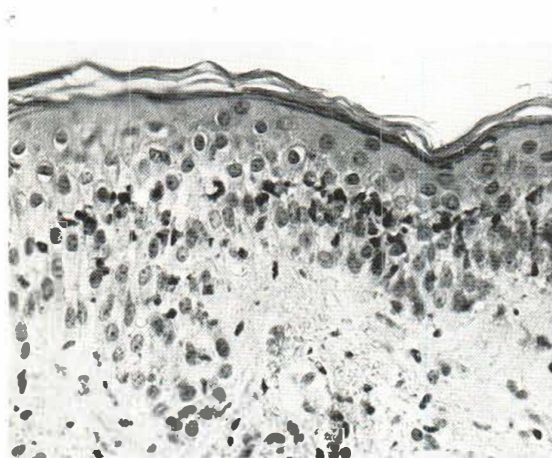


Fig. 1. Early biopsy showing intercellular edema of epidermal keratinocytes and infiltration with eosinophilic leukocytes.

CASE REPORT

A 74-year-old woman had, since the age of 40, suffered from diabetes mellitus and been treated with insulin. During recent years she had had diabetic retinopathy, neuropathy and nephropathy and was treated with propylthio-uracil for hyperthyroidism. In February 1977 she noticed an itching exanthema on both legs. Gradually the exanthema became universal. The first clinical impression was that of atypical pityriasis rosea. A skin biopsy on April 6, 1977, at the local hospital was interpreted as an allergic vasculitis. However, revision of this early biopsy revealed intercellular edema of epidermal keratinocytes and profuse infiltration with eosinophilic leukocytes, i.e. the characteristic features of eosinophilic spongiosis (Fig. 1).

She was referred to us on May 27. She displayed a dark red, polycyclic edematous eruption with partly confluent elements, located on the trunk and proximal upper and lower extremities. Numerous itch marks but no bullae were noticed (Fig. 2).

On June 20 she presented a bulla on the right elbow. A tentative diagnosis of dermatitis herpetiformis was made and treatment with Diaphenylsulfon (Avlosulfon®, ICI) 50 mg a day was started. A biopsy from the back was now interpreted as compatible with subcorneal pustulosis.

The eruption progressed despite Avlosulfon® with the



Fig. 2. Skin eruption at its peak, displaying areate and annular lesions, several excoriations, but no bullae.

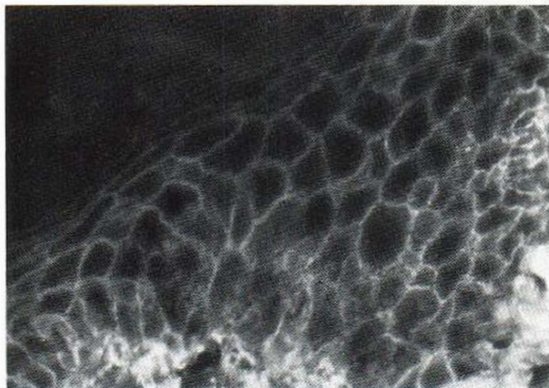


Fig. 3. Direct immunofluorescence demonstrating intercellular IgG deposits.

appearance of bullae on the palms and soles. Histologic examination of a bulla from the palm was not helpful, but by the immunofluorescence technique, IgG deposits intercellularly in the epidermis were demonstrated, thus revealing Pemphigus foliaceus (Fig. 3). Avlosulfon® was discontinued, prednisone 60 mg a day was started and the eruption subsided rapidly. Prior to prednisone treatment the patient had an eosinophilia of 51% of a total of 8000 leukocytes per microlitre. Following 2 weeks of prednisone treatment that was gradually reduced to 40 mg a day, the eosinophilic count dropped to 2% of a total of 6900 leukocytes per microlitre. Azathioprin (Imurel® Wellcome) 50 mg a day was added and prednisone further reduced to 25 mg a day. This combination was successful in preventing recurrence within the observation period of 3 months.

DISCUSSION

Clinically our patient presented as a case of dermatitis herpetiformis, yet she did not respond to Avlosulfon®. The skin biopsy in this preacantholytic stage showed eosinophilic spongiosis as first reported to be the presenting sign in Pemphigus by Emmerson & Wilson-Jones (2), later by others (1, 3, 4). In addition, our patient had a pronounced eosinophilia with 51% eosinophilic leukocytes in the peripheral blood. Osteen et al. (4) and Sneddon & Church (5) also found eosinophilia amounting to 15–24% in peripheral blood.

The combination of a dermatitis herpetiformis-like eruption and eosinophilic spongiosis in an early skin biopsy should suggest Pemphigus. The demonstration of intercellular IgG deposits in the epidermis by direct immunofluorescence microscopy, however, is essential for the establishment of the diagnosis of Pemphigus foliaceus.

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Delayed Cold Urticaria

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Abstract. Five patients with delayed cold urticaria are described. The urticarial skin response was present between 24 and 72 hours after ice challenge. In two of the patients the cold sensitivity was of clinical relevance. Some of the patients displayed low alpha₁-antitrypsin and increased C4 levels in their serum. Our findings may justify the introduction of cold provocation as a routine procedure in the investigation of a patient with chronic urticaria.

Key words Cold urticaria; Delayed cold urticaria; Chronic urticaria.

In cold urticaria the symptoms usually develop within minutes after cold exposure and subsequent warming. In most cases the clinician is able to establish the diagnosis in his office. There are, however, a few reports of delayed cold urticaria