

during the past 2 years. They were annular areas with raised erythematous, palpable border, the centres were slightly depigmented and atrophic without telangiectasis or hair loss (Fig. 1). There was no history of previous serious illness. Physical examination apart from the skin disclosed no abnormalities. The tuberculin test was positive to 10 TU. The Kveim test was negative. X-rays of the chest, hands and feet were normal. Laboratory studies including the glucose tolerance test, serum cholesterol and triglycerides were normal. Biopsies from the forehead showed large, bizarre giant cells scattered between the collagen bundles, generally in small groups. Some of the giant cells contained beautiful, large asteroid bodies (Fig. 2). Lymphocytes surrounded the giant cells in small quantities. Necrosis and necrobiosis were absent.

DISCUSSION

Atypical necrobiosis lipoidica of the face is not particularly rare. It is frequently confused with annular sarcoid (1, 5). Miescher's granulomatosis disciformis chronica et progressiva presented as a separate entity in 1948 by Miescher & Leder (3) is nowadays considered as a variant of the necrobiosis lipoidica (2). The fact is that diabetes seems to be a rare occurrence in patients in whom the lesions are present in areas exclusive of the legs. Wilson Jones (5) found diabetes in only one man out of 21 patients with lesions of the face and scalp (of these only two were men). Tappeiner (4) published under the diagnosis of granulomatosis disciformis chronica et progressiva the case of a patient with a lesion limited to the scalp margin which histologically was of the same granulomatous type containing prominent giant cells with asteroid bodies lying loose in the connective tissue. We think that this condition should be designated necrobiosis lipoidica, instead.

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Pemphigus Vulgaris in a 15-year-old Girl

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Received November 22, 1977

Abstract. Pemphigus vulgaris is uncommon in adolescence and only ten well documented cases in this age group were found in a recent review (5). Because the condition is often more severe in the younger age group it is important to consider it in the differential diagnosis of bullous eruptions of childhood and to perform direct and indirect immunofluorescence studies.

CASE REPORT

This female patient was 15 years old when she was referred in March 1976 with a 6-week history of blisters and ulcers in the mouth and a 2-week history of similar lesions



Fig. 1. Flaccid bullae and extensive ulceration on the back.



Fig. 2. Extensive intra-epidermal bulla with acantholysis and little inflammatory reaction in the dermis.



Fig. 3. Direct immunofluorescence of involved skin showing intercellular IgG deposition. $\times 50$.

on the backs of the hands. There was no history of preceding illness or drug ingestion.

On examination she had severe ulceration of the mouth and vulva. Scattered over the trunk and limbs there were flaccid bullae and extensive areas of ulceration (Fig. 1).

Histological examination of a biopsy of a blister from the back revealed an extensive intra-epidermal bulla with acantholysis and virtually no inflammatory reaction in the dermis (Fig. 2). Desmosome antibody was present in the serum at a titre of 1 in 20. Direct immunofluorescence showed IgG and C3 deposition in the intercellular spaces of the epidermis in both involved (Fig. 3) and uninvolved skin taken distant from a lesion.

Therapy was commenced with prednisolone 100 mg daily and Azathioprine 50 mg twice a day. Within 3 days the mucous membrane and skin lesions had improved and no new blisters appeared. The condition remained controlled on reducing doses of prednisolone and Azathioprine.

In September 1976, 6 months after diagnosis, while the patient was taking prednisolone 12.5 mg daily and Azathioprine 50 mg daily, the immunological investigations were repeated. Desmosome antibody was absent and no IgG or C3 was present in the skin. These negative findings prompted a reduction in therapy to prednisolone 5 mg daily and the Azathioprine was stopped. However, within 3 weeks blisters recurred in the mouth and on the skin.

The dose of prednisolone was increased and Azathioprine re-introduced. The patient has now remained free of blisters for 6 months on prednisolone 10 mg daily and Azathioprine 50 mg daily.

DISCUSSION

This case serves as a reminder that pemphigus vulgaris should be considered in the differential diagnosis of bullous dermatoses in adolescence. In addition to showing the classical immunofluorescent changes of pemphigus vulgaris (3) some unusual features have been demonstrated. Intercellular IgG and C3 were found in clinically normal skin distant from the lesions. Usually IgG is demonstrated only in skin adjacent to the lesions (1, 8, 9). C3 has previously been shown only in lesional skin prior to steroid therapy (4). The finding of intercellular IgG in uninvolved skin stresses the importance of performing direct immunofluorescence on it before it is used as a substrate for indirect immunofluorescence with the patient's serum.

The intercellular IgG disappeared from the skin 6 months after therapy was commenced. When this phenomenon has been studied in a few other cases, positive intercellular staining has been shown to persist after therapeutic remission. The accepted indication of remission in pemphigus vulgaris is a fall in titre of desmosome antibody (2, 6, 7) but in this patient the disappearance of the antibody did not denote remission, since a reduction in therapy at this time precipitated a relapse.

Detailed and repeated immunofluorescence investigations have not previously been reported in childhood pemphigus vulgaris and the unusual findings in this patient suggest that the disease in adolescents may behave differently from that occurring in adults.

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Palpebral cellulitis

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Received August 8, 1977

Abstract. Patients with palpebral cellulitis may turn in the first place to a skin clinic. The clinical picture and the course of disease are illustrated here by four case histories. The importance of differentiating between collateral orbital edema, palpebral cellulitis and orbital cellulitis is stressed.

Palpebral cellulitis is an uncommon infection. It can make its entry through an erosion or puncture or via the blood stream, or arise from an inflammation in the skin of the eyelids, the conjunctiva, the orbit, the lacrimal passages, the nasal sinuses, or even the teeth.

Palpebral cellulitis usually occurs unilaterally and is associated with swelling and redness of the involved area. The pre-auricular and mandibular lymph nodes usually become swollen, and some fever may be present. In the early stages the skin is intact, but subsequently erosions, pustules, or even gangrene may develop.

The bulbus and the orbit are usually not involved. The eye can therefore be moved freely, and protrusion is rare.

The condition must be distinguished from allergic contact dermatitis, angioneurotic edema, collateral edema due to ethmoiditis and orbital cellulitis.

Patients with palpebral cellulitis may turn first to a skin clinic, and we therefore think it worthwhile to to present some cases and to discuss the differential diagnoses.

CASE REPORTS

Case 1

Female, 24 years old, developed in the course of about 6 weeks massive swelling and erythema of the eyelids on the left side. Her general condition was well, but she had bouts of low grade fever. On admission, there was massive swelling of the eyelids and the skin looked rather moist (fig. 1). Vision was unaffected, the eye could be moved freely, there was no exophthalmus, but conjunctival chemosis. ESR was 63 mm/h per hour; ear-nose-throat examination was normal. X-ray examination of the nasal sinuses showed a normal picture, but an inflammatory