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AN UNUSUAL EXAMPLE OF NECROBIOSIS LIPOIDICA AFFECTING THE FACE

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Necrobiosis lipoidica is a rare skin disease most often seen in diabetics. The diagnosis may be difficult. The lesions are frequently mistaken for granuloma annulare, granuloma multiforme, rheumatoid nodules, xanthelasmata and Miescher's granulomatosis disciformis (Muller and Winkelmann, 1966; Beare and Wilson Jones, 1972). The lesions are usually encountered on the legs and the trunk. This paper reports a patient with recurrent histologically proven necrobiosis lipoidica affecting the eyelids, associated with other typical lesions elsewhere on the body.

CASE REPORT

The patient, a 70-year-old retired farmer, first attended our clinic in 1973 with a history of slowly growing yellowish plaques on the left upper and lower eyelids. On the upper lid there was an irregular ulcerating tumour, $2 \times 1{\cdot}5$ cm in size, and another solid tumour of equal size on the lower lid.

Two years previously, the patient had been treated elsewhere by intra-lesional steroid injections for infiltrating ulcerating pigmented tumour plaques on the pretibial aspect of both legs. The response to treatment was not good and biopsies from the tumours showed the typical changes of necrobiosis lipoidica. He had also a similar tumour presumably of the same aetiology, on the left side of the neck.

The eyelid lesions which concerned us resembled xanthelasmata, but the biopsy appearances were again characteristic of necrobiosis lipoidica. The lesions were excised and the defects skin grafted with a good result. Five years later, in 1978, there was a recurrence of the eyelid lesions which now involved the upper and lower eyelids of both eyes. There was marked palpebral shrinkage causing a closing defect and ectropion, most marked on the left side (Figs. 1 and 2).

In the interim he had again been treated elsewhere, both operatively and by steroid injection, but the response was poor. He complained less about the crural lesions even though they had increased in size, and a new finding was a small lesion of similar appearance to the others on the left arm.

The lesions on the left upper and lower eyelids were again excised and skin grafts applied. The ectropion was also corrected.

HISTOPATHOLOGY

Biopsy of the left leg showed patchy dermal necrosis, with prominent proliferative fibroblasts and intra-cellular lipid deposits.

The appearance of upper and lower left eyelid biopsies were similar, showing obvious collagen necrosis, patchy granulomatous infiltration including inflammatory cells and plasma cells. Repeat biopsies taken before the treatment for recurrence in 1978 showed essentially the same features.

Recurrent lesions of necrobiosis lipoidica at the outer canthus of the left orbit. There is marked ectropion of the lower eyelid.

FIG. 1. Face view.

FIG. 2. Lateral view.

PREVIOUS MEDICAL HISTORY

Laboratory studies in 1971 when the patient was 68 years old had shown an elevated erythrocyte sedimentation rate (ESR) (107 mm/h). Immune electrophoresis had shown extreme elevation in the IgG (M-component). There had been no Bence-Jones protein in the urine, and the bone marrow appearances were normal. The patient was non-diabetic.

At the age of 74, the ESR was still elevated, 130 mm/h, and the patient was markedly anaemic. No definitive diagnosis was made. He was treated by blood transfusions. Subsequent investigations showed consistently elevated ESR (170 mm/h), a pancytopenia, low albumin, and an immune-electrophoretic M-component (IgG, kappa). X-rays of the skull, spine, thorax and pelvis were all normal. The patient refused further investigations and treatment, and died 6 months later in another hospital. Post mortem studies showed multiple myelomata of the kidneys, spleen and bone marrow.

DISCUSSION

The clinical and histological appearances of necrobiosis lipoidica have been recognised for more than 50 years (Oppenheim, 1930). The condition was first seen in patients with diabetes, and was considered to be a complication of the disease. Later, it was seen also in non-diabetic patients.

Necrobiosis lipoidica is rare, occurring in only 3 out of every 1,000 diabetics. It does not bear any consistent relationship to the duration or seriousness of the disease (Wilson Jones, 1971). Seventy per cent of patients with necrobiosis lipoidica have diabetes, which generally precedes the appearance of the skin lesions. Women are primarily affected (ratio 3:1), and the age of onset is usually between 35 and 45 years in diabetics, somewhat later in non-diabetics (Muller and Winkelmann, 1966).

Clinically, the lesions are round or oval indurated plaques, with slightly raised

borders. There may be areas of central depigmentation and atrophy. Yellowish areas are centrally located, while the margins are reddish-brown (Beare and Wilson Jones, 1972). Ulceration occurs in one-third of the patients. The lesions usually appear in the pretibial region but may be seen on the thighs, feet, trunk, arms, hands, neck and scalp. Facial lesions are distinctly uncommon.

It is interesting to note that diabetes is less frequently associated with necrobiosis lipoidica when this occurs on the face or scalp (Wilson Jones, 1971). The histological appearance of the "diabetic" lesions on the legs also differs somewhat from that of the face and the scalp lesions, which display more prominent multinucleate giant cells and epithelioid cells. By contrast necrobiotic debris and palisading granulomas are not so marked in the facial lesions (Rollins and Winkelmann, 1960; Mehregan and Altman, 1973), which also display destruction of the elastic network, but minimal changes in the collagen fibres (Muller and Winkelmann, 1966).

The differential diagnosis of unusual ulcerative lesions of the eyelids should include necrobiosis lipoidica. Histological examination will confirm the diagnosis. The condition responds poorly to steroid therapy and is probably best treated by excision and skin grafting, when indicated. However, because the true cause of the lesion is unknown, recurrence at the site of operation is a distinct possibility. We can find no reference in the literature to indicate a possible link between necrobiosis lipoidica and myelomatosis.

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