Periorbital necrobiosis lipoidica

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Abstract

Necrobiosis lipoidica is a granulomatous skin condition typically occurring on the legs. A patient with this condition presented with gross bilateral induration of the eyelids sufficient to close both eyes.

Necrobiosis lipoidica is a rare granulomatous dermatopathy of uncertain aetiology. In about 70% of cases it is associated with diabetes.¹ Its typical presentation is of yellow, waxy, atrophic patches with purple margins on the shins. Less commonly it presents in sites away from the legs and very rarely on the face. It has been reported involving the periorbital tissues as small yellowish plaques.² This report is of a patient with necrobiosis lipoidica affecting all four eyelids with gross infiltration causing closure of both eyes.

Case report

The patient, a 66-year-old diabetic male, presented with both eyes closed by massive

swelling and induration of the eyelids (Fig 1). Two weeks previously he had noticed small, painless, cutaneous nodules on his chin, neck, back, and shoulders. Over the next few days these lesions grew in size, and similar small nodules were noticed in the eyelids. From then on until the time of presentation all the lesions grew progressively larger and the lumps in the eyelids began to coalesce. Initially the eyelid lesions were 70-80 mm in diameter and the other lesions 30-40 mm in diameter. All the lesions (six in total) were well circumscribed, firm, non tender, prominent nodules with a typical waxy appearance and with telangiectasia round the edges. There was a slight purulent discharge from the surface of lesions round the eyes.

The patient was otherwise well. He was taking chlorpropamide 300 mg once daily for his diabetes and no other medication. A general medical examination gave normal results, as did the following investigations: full blood count, erythrocyte sedimentation rate, serum urea and electrolytes, random blood glucose, chest x-ray, electrocardiogram, and liver function tests with the exception of a serum alkaline phosphatase level of 199 IU/l (normal: 35-125 IU/l). Biopsy gave the diagnosis of necrobiosis lipoidica.

The patient was admitted to a medical ward and started on intravenous flucloxacillin and fucidin and oral prednisolone 40 mg once a day. Treatment of his diabetes was changed to an insulin (Actraphane) adjustable regimen. Topical chloramphenicol was applied to the eyelids.

Intravenous antibiotics were continued for five days and the prednisolone was reduced over two weeks to 20 mg on alternate days. All the lesions began to resolve noticeably by the second day of treatment; two weeks later they were all still present but considerably better. Both eyes were now open and found to be normal.

Histopathology. The specimen revealed scattered areas of collagen necrosis. Histiocytes were diffusely intermingled throughout, and multinucleate giant cell granuloma formation was prominent. A minor degree of vascular obliteration was present (Fig 2).

Discussion

Necrobiosis lipoidica affects 0.3% of diabetics, but about 70% of cases have diabetes. It involves sites away from the legs in 15% of patients but in only 2% are the legs uninvolved. Its occurrence is unrelated to either the duration or the control of the diabetes, and women are more commonly affected than men, usually in the 35 to 45 age group.³

Sarcoid, foreign body reactions, and infection by Mycobacteria or fungi are other important causes of granulomatous inflammation in the

Figure 1 Appearance at presentation showing the marked lid infiltration together with other skin lesions on the chin and in the suprasternal notch.

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Figure 2 Skin biopsy. A=epidermis; <<=giant cell; B=granulomatous inflammation; C=collagen necrosis (magnified $\times 125$, H and E stain).

skin, and they may also have responded to steroid/antibiotic therapy. Histological assessment in our case, though, indicated a necrobiotic process.

Other necrobiotic skin conditions which may have a similar clinical appearance include granuloma annulare, rheumatoid and pseudo-rheumatoid nodules. The former is also associated with diabetes.⁴

These conditions can again be differentiated histologically, principally by the pattern of collagen necrosis, histiocyte distribution, and the extent of granuloma formation.

Usually treatment for necrobiosis lipoidica is required only for cosmetic reasons or for ulceration with secondary infection. Intralesional steroid injections and local excision have been tried with varying success.^{2,5} The response to systemic steroids in our patient was dramatic, but five months after presentation the lesions are still detectable. Steroid treatment has now been stopped.

Most cases of necrobiosis lipoidica are slowly progressive and less dramatic in presentation.⁶ To our knowledge such extensive involvement of the eyelids has not been reported before, and the condition should be included in the differential diagnosis of any nodular or diffuse induration of the eyelids, particularly in diabetic patients.

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