

A brand-like plaque

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Clinical findings

A 51-year-old woman presented with a 1-year history of an asymptomatic figurate skin lesion that had gradually developed on the nape of her neck. The patient's personal medical history was unremarkable. Examination revealed a 6-cm diameter, pink-red, geometrically delimited, embossed 'C'-shaped plaque resembling a cattle brand (Fig. 1). An irregular hypopigmented macular area measuring 2 cm across arose from the tips of the C-shaped outline and extended outwards. A punch biopsy of the rim of the lesion was performed.

Histopathological findings

Haematoxylin & eosin stained sections revealed a granulomatous inflammation with many foreign body giant cells (Fig. 2). Orcein staining showed prominent elastolysis in the centre of the granuloma (Fig. 3) with elastoclasia (Fig. 4).

What is your diagnosis?



Figure 1 A 6 cm in diameter, figurate, erythematous C-shaped plaque with raised borders and a peripheral hypopigmented area involving the nape of a 51-year-old woman.

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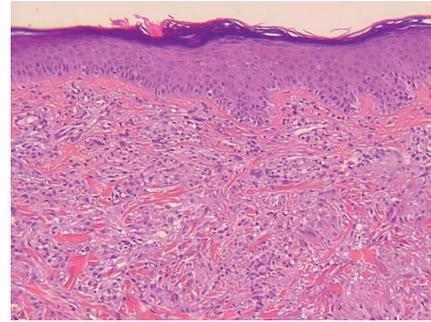


Figure 2 Haematoxylin & eosin staining revealed a granulomatous inflammation with many foreign-body giant cells (original magnification $\times 40$).

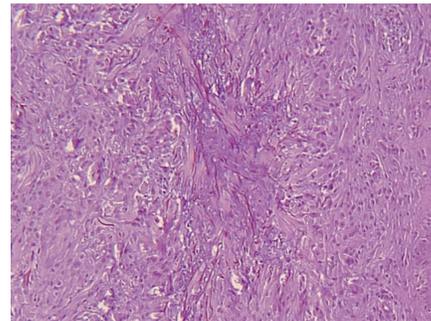


Figure 3 Orcein staining shows prominent elastolytic changes in the central area of the granuloma. Elastoclasia can be seen peripherally (original magnification $\times 60$).

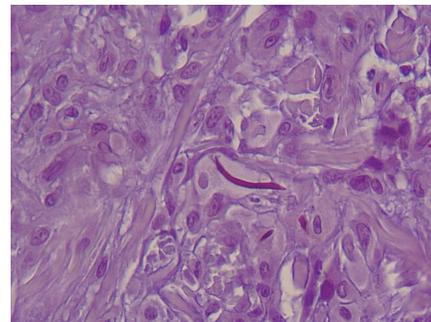


Figure 4 Elastoclasia or elastophagocytosis. Observe foreign-body giant cells engulfing elastotic fibres (orcein, original magnification $\times 100$).

Diagnosis

Annular elastolytic giant cell granuloma.

Discussion

Annular elastolytic giant cell granuloma (AEGCG) is a term coined by Hanke and coworkers¹ in 1979 to encompass three similar conditions previously described: namely, O'Brien's actinic granuloma, granulomatosis disciformis of Miescher and atypical necrobiosis lipoidica in a single and distinct clinicopathological entity. AEGCG affects middle-aged women (male to female ratio, 2 : 1) frequently over the age of 40 years. It presents as a single or grouped flesh or pink coloured papules which gradually coalesce into annular erythematous plaques with raised borders and slightly atrophic, hypopigmented central areas. Sun-exposed regions of the face, neck, arms and chest are the most common localizations involved. Annular lesions vary from 1 to 10 cm in diameter. A solitary case of systemic AEGCG with cutaneous, ocular, lymph node and intestinal involvement has been reported.² Histopathological findings at the rim of the lesion differ from those in the centre. In the mid to the upper dermis of the rim there is an infiltrate composed mainly of histiocytes with many foreign body giant cells arranged around elastotic fibres. Elastic tissue fragments can be identified adjacent to and engulfed by the giant cells (elastoclasia or elastophagocytosis). Occasionally asteroid bodies (degenerated elastic fibres) can be observed. The absence of altered collagen, necrobiosis, increased dermal mucin and epithelioid cell tubercles can distinguish most AEGCG cases from granuloma annulare (GA), necrobiosis lipoidica and cutaneous sarcoidosis. In the central zone of the lesion there are very low quantities or even absence of both elastotic and elastic fibres.³

The pathogenesis of AEGCG remains unclear. There is controversy about whether AEGCG is a distinct disorder or a variant of GA on sun-exposed areas. O'Brien⁴ proposed that actinic damage to elastic fibres would precipitate the granulomatous reaction. However cases of AEGCG involving nonexposed skin have been reported.³ Furthermore, elastolysis and elastoclasia have been reported to occur in GA as well, raising the possibility that the elastolytic event would be secondary to the granulomatous inflammation rather than an initiating mechanism.² Fujimoto *et al.*⁵ have recently found that elastin fragments induced factor XIIIa+ cells and CD68+

macrophages to form granulomas or multinucleated giant cells which expressed the 67-kDa elastin receptor. Proliferative CD68+ and factor XIIIa+ cells along with epithelioid and multinucleated giant cells expressing the 67-kDa elastin receptor were seen in skin biopsies from patients affected by AEGCG.

Annular elastolytic giant cell granuloma is a persistent condition that responds poorly to topical or intralesional corticosteroids. There are single case reports of successful treatment with cyclosporin or chloroquine. Spontaneous resolution may occur over months to years without scarring or mottled dyspigmentation.²

Learning points

- AEGCG encompasses O'Brien's actinic granuloma, granulomatosis disciformis of Miescher and atypical necrobiosis lipoidica.
- It presents as solitary or multiple annular erythematous plaques with raised borders on sun-exposed areas.
- Histologically, AEGCG is characterized by elastolysis, elastoclasia and multinucleated giant cells in the rim of the lesion, while in the centre there is a lack of both elastotic and elastic fibres. Necrobiosis, increased dermal mucin and epithelioid cell tubercles are absent.
- There is controversy about its pathogenesis, although elastin fragments may contribute to granuloma formation.
- There is no satisfactory treatment for this condition but spontaneous resolution may occur.

References

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