Drug-induced elastosis perforans serpiginosa

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DESCRIPTION

A 42-year-old woman presented with a 5-year history of multiple enlarging skin lesions. Her medical history was significant for Wilson's disease, for which she had taken penicillamine for approximately 20 years.

She had multiple small erythematous keratotic papules grouped in both a circular and arciform pattern. These lesions were present on her neck, in her axillary skin folds, antecubital fossa and groin. She had associated skin laxity (figures 1 and 2).

A skin biopsy showed altered dermal collagen with extrusion of elastic fibres through the epidermis via a hair follicle (figure 3). The diagnosis was penicillamine-induced elastosis perforans serpiginosa.

Lysyl-oxidase is a copper-dependent enzyme required for dermal elastic fibre cross-linking. Penicillamine, a copper chelator, impairs the activity of this enzyme. There have been a number of case reports of elastosis perforans serpiginosa in the setting of penicillamine therapy for both Wilson's disease and systemic sclerosis. The lesions are reported to improve with discontinuation of penicillamine, during pregnancy,¹ and with application of liquid nitrogen, intralesional triamcinolone acetone

Figure 3 Skin biopsy: Extrusion of elastic fibres through the epidermis via a hair follicle.

and topical allium cepa-allantoin-pentaglycan gel.² Alternative agents for treating Wilson's disease include trientine and zinc salts. The former has been reported to be associated with deterioration in skin lesions in one case report.³ Interestingly, there are familial cases reported in the literature, with a presumed autosomal dominant mode of inheritance.⁴

In our case, an attempt was made to stop penicillamine and our patient was switched to trientine. The symptoms of Wilson's disease deteriorated necessitating reintroduction of penicillamine. Her skin is being treated with copper containing creams and a lower dose of penicillamine.

Figure 1 Keratotic papules, axillary fold.

Figure 2 Skin laxity and keratotic papules grouped in serpiginous and arciform pattern, anterior neck.

Learning points

- ▶ There are 3 clinical variants of elastosis perforans serpiginosa:
 - Idiopathic.
 - Reactive (related to systemic disease, eg, Marfan's, Ehlers Danlos, pseudoxanthoma elasticum and Downs syndromes).
 - Drug-induced (D-penicillamine).
- ▶ It is a rare condition characterised by transepidermal elimination of elastic fibre aggregates.
- ▶ Management is challenging and often complicated by a deterioration in Wilson's disease on cessation of penicillamine.

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