

Clinicopathologic challenge

**Chronic annular lesions of the cheeks**

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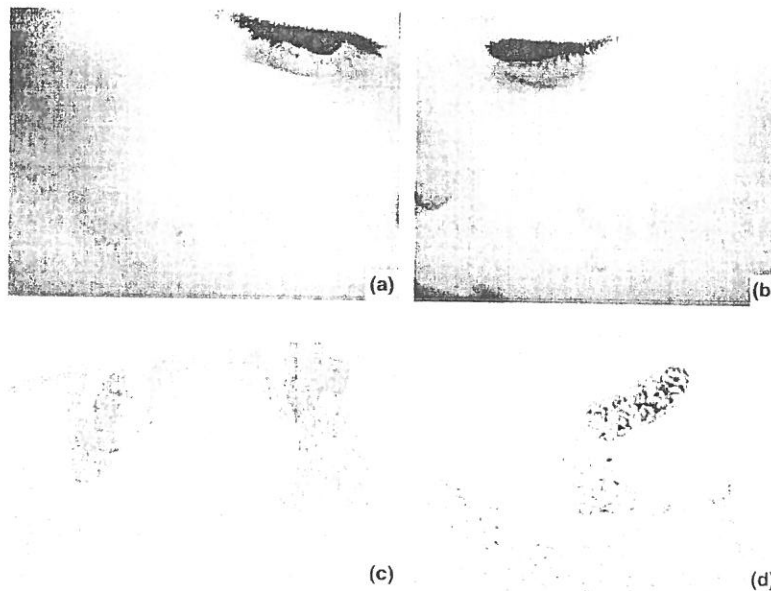
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**History**

One year previously an otherwise healthy 10-year-old girl presented with asymptomatic symmetrical annular lesions involving both cheeks (Fig. 1a,b). Progressively, she developed slightly erythematous and hyperkeratotic papules with small nodular elements. The pattern was linear,

annular and serpiginous. Some lesions revealed central atrophy. No other skin lesions were present. There was no significant medical, allergic and surgical history, and the family history was unremarkable. An extensive work-up excluded the association with other genetic disorders. There was no oral drug intake. The patient was previously treated with emollients, topical corticosteroids,

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**Figure 1** Well-circumscribed erythematous and hyperkeratotic serpiginous papules of the left (a) and right (b) cheeks. (c) Moderately acanthotic epidermis and a transepidermal perforating channel containing elastic fibers (H/E stain, 100×). (d) Orcein staining revealing the transepidermal elimination of altered elastic fibers (Orcein stain, 100×)

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antimycotics and keratolytics without success. This clinical appearance suggested a diagnosis of granuloma annulare or post-traumatic epidermal inclusion cysts. A 2-mm punch-biopsy was performed under local anesthesia (Fig. 1c,d).

**What is your diagnosis?**

Elastosis perforans serpiginosa (EPS).

**Discussion**

EPS is one of the primary perforating dermatoses of dermal origin.<sup>1</sup> Histologically, EPS reveals a moderately acanthotic epidermis and a transepidermal perforating channel containing basophilic material. An inflammatory reaction may be present in the superficial dermis with multinucleated giant cells. Orcein staining illustrates the transepidermal elimination of the elastic fibers, and the increased quantity of elastic fibers in the upper dermis.<sup>2</sup> Focal dermal elastosis is also observed.<sup>3</sup> Histopathology is also capable to eliminate other reactive perforating collagenoses.

EPS usually begins in childhood or early adulthood. Most commonly, the lateral aspects of the neck are involved, followed by the arms and flexural areas. This young girl presented with bilateral EPS lesions on the cheeks, a rather unusual localization for EPS.

The etiology of EPS is unknown,<sup>3</sup> but in 30–40% of the cases it is associated with systemic diseases, such as Down syndrome, Ehlers–Danlos syndrome, acrogeria, osteogenesis imperfecta, pseudoxanthoma elasticum, Rothmund–Thomson syndrome or Marfan syndrome.<sup>4,5</sup> A few reports cite familial occurrence.<sup>6</sup> Besides the idiopathic type, a penicillamine-induced form is described.<sup>7</sup> A thorough work-up is recommended to search for these associations.

Clinically, multiple small, firm keratotic papules and nodules are arranged in annular plaques over the neck, axillae, antecubital fossae and forearms. EPS is frequently misdiagnosed because of its rarity.<sup>8</sup> The clinical differential diagnosis includes tinea, granuloma annulare, sarcoidosis, actinic granuloma, perforating pseudoxanthoma elasticum, porokeratosis and discoid lupus erythematosus.

There is no standard therapy for EPS. Successful treatments have been reported with liquid nitrogen cryotherapy, isotretinoin, tazarotene, imiquimod and CO<sub>2</sub>,

Erbium-YAG and pulsed dye lasers.<sup>9,10</sup> More anecdotal reports mention menthol, sulfur and benzoyl peroxide. Surgical options include local excisions. Electro-dessication or cellophane tape-stripping are other treatment alternatives. Koebner effects should be avoided. Long-term results are variable. Treatment may cause atrophy and scarring. Spontaneous resolution of EPS has occasionally been reported, often after several years.<sup>1</sup>

Topical retinoids were prescribed without success. CO<sub>2</sub> laser resurfacing will be proposed in the future. EPS should be included in the differential diagnosis of annular lesions of the face during childhood.

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