Pediatric-onset necrobiosis lipoidica

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A 14-year-old boy with a 9 year history of type 1 diabetes mellitus (DM; HbA1c, 8.0–11.5%) presented with symmetric pre-tibial red-brown plaques with well-defined borders (Fig. 1) that had lasted for 6 months. He denied itch, pain or a previous traumatic event, and noted that the skin lesions had started as small papules (<0.5 cm).

Skin biopsy was performed and histopathology indicated extensive necrotic areas of collagen degeneration in the dermis with palisaded granulomas composed of histiocytes, lymphocytes and fibroblasts (Fig. 2). Thickening of the blood vessel walls was also present.

These findings were consistent with necrobiosis lipoidica (NL), a disorder of collagen degeneration strongly associated with DM but also described in non-diabetic patients.1 Pediatric onset is rare.1,2

The etiology of NL remains uncertain, but it seems to involve metabolic and inflammatory changes such as an increase in tumor necrosis factor-α skin level, deposition of glycoproteins in blood vessel walls, antibody-mediated vasculitis and increased collagen cross-linking.1,3

Lesions are usually asymptomatic and the main complaint is the unpleasant cosmetic appearance.

The patient was treated with 0.1% topical tacrolimus with partial regression of the erythema and size of the lesions in 6 months. Treatment for NL is not very effective, partially because the exact etiology remains unknown.1,4 Glycemic control is important, and topical corticosteroids should be used with caution in order to avoid cumulative atrophic effect.

Fig. 1 Pre-tibial red-brown plaque (60 × 25 mm) with indurated borders and atrophic centers.

Fig. 2 Areas of collagen degeneration with palisaded granulomas involving the subcutaneous tissue and dermis.

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The main complication of the disease is ulceration that could occur after trauma.
Follow up is advised given that squamous cell carcinoma has been reported in some patients.5

References

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