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CASE FOR DIAGNOSIS

Unilateral Linear Hyperpigmentation in a Girl

Pigmentación lineal hemicorporal en una niña

Medical History

A 12-year-old girl with no past history of interest was seen for pigmented lesions that had started to appeared 3 years earlier on the trunk, right arm, and right leg. The lesions were asymptomatic and had progressed gradually in a cephalad direction. She reported no previous episodes of inflammation or induration, and there had been no significant changes in color.

Physical Examination

On physical examination, hyperpigmented lesions with a Blaschkoid distribution were observed on the right side along the full length of the limbs and the anterior and posterior surfaces of the trunk (Figures 1 and 2). The lesions were not indurated and there was no alopecia, but some affected areas were slightly depressed.

Histopathology

Histology revealed marked hyperpigmentation of the basal layer of the epidermis, thickened, homogeneous collagen in the reticular dermis, and a mild perivascular chronic inflammatory infiltrate formed of lymphocytes, histiocytes, and occasional plasma cells (Figure 3). The skin adnexa and eccrine sweat glands were completely normal, and staining with orcein excluded alterations in the elastin fibers.

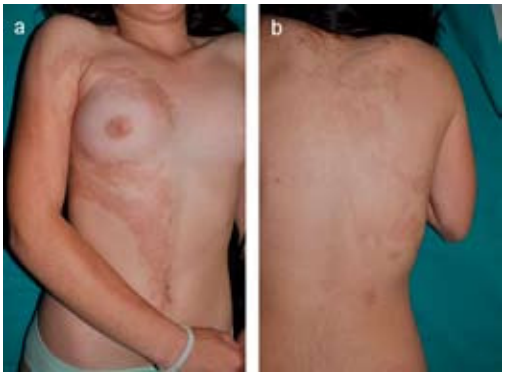

Figure 1

Additional Tests

Complete blood count and routine biochemistry were normal, and antinuclear antigens were negative.

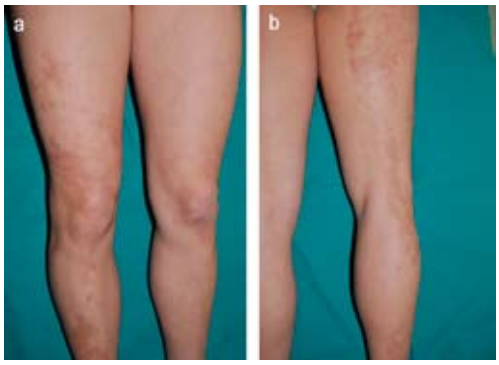

Figure 2

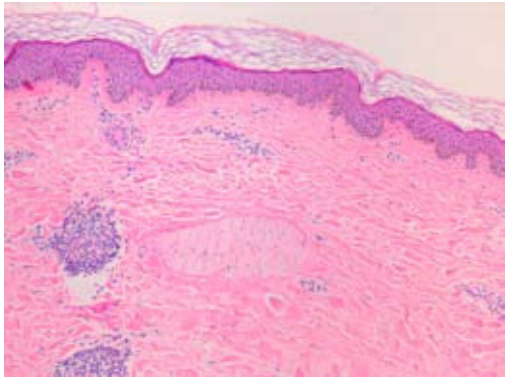

Figure 2 Hematoxylin-eosin stain, original magnification ×40.

What Was Your Diagnosis?

Diagnosis

Linear atrophoderma of Moulin.

Clinical Course and Treatment

As there was a clinical suspicion of morphea, the patient was treated with conventional ultraviolet A phototherapy (3 sessions per week for 2 months) and oral cystine for 1 month, with no improvement. At the time of writing, the lesions were stable.

Discussion

Linear atrophoderma of Moulin is a rare condition of unknown origin characterized by the appearance of linear hyperpigmented lesions along the Blaschko lines. In the majority of cases published, the condition develops during childhood and affects a small area of the body[1,2]; a completely unilateral distribution, as in our patient, is very rare. To date only sporadic cases have been reported and, as occurs with other diseases of blaschkoid distribution, it is believed to develop as the result of a postzygotic mosaicism that affects an unknown gene.[1] The lesions are asymptomatic and are only occasionally preceded by an inflammatory phase.[1,3] There have been no reports of systemic diseases associated with linear atrophoderma of Moulin, although positive antinuclear antibodies have been detected in a single case.[4] Histological findings are variable; no dermal sclerosis or inflammatory infiltrate was described in the early reported cases,[5] but a variable degree of dermal collagenization and the presence of perivascular inflammatory infiltrates were subsequently observed.[1,2] There is hyperpigmentation of the basal layer and, despite the suggestion of its name, there is no dermal atrophy; the slight depression of the lesions that is observed occurs due to a reduction in the subcutaneous cellular tissue.[5] The principal differential diagnoses include atrophoderma of Pasini and Pierini and linear morphea. Atrophoderma of Pasini and Pierini is histologically indistinguishable from linear atrophoderma of Moulin, but the lesions are patchy rather than linear and are typically found on the back. Linear morphea lesions have a pearly appearance, though they are occasionally pigmented, and there is alopecia and induration; in late phases they lose the induration. Histologically there is much greater sclerosis than in linear atrophoderma of Moulin, loss of the skin adnexa, and occasional involvement of the deep tissues. In view of the clinical and histological similarities, some authors consider that linear atrophoderma of Moulin could be a final stage of linear morphea; however, others believe there to be sufficient evidence to differentiate the 2 conditions.[1]

Linear atrophoderma of Moulin has a chronic course and there is no known effective treatment for the disease. In a single case, potassium aminobenzoate (Potaba) proved useful in stabilizing the disease at an early stage.[6]

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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