CLINICAL SNAPSHOT

Scleroderma en Coup de Sabre

A 3-year-old girl was brought to us with facial cutaneous lesions. The initial manifestation was an erythematous macula on the nose, followed 18 months later by formation of a hypopigmented atrophic scar extending from the hairline to the jaw. This was accompanied by craniofacial asymmetry and malposition of the jaw. Overall, the clinical and histological findings pointed to linear circumscribed scleroderma (scleroderma en coup de sabre). This is a sclerosing connective-tissue disease of the skin that may also affect other tissues such as bone. Scleroderma en coup de sabre can result in neurological symptoms such as headache or seizures and may even cause structural alterations in the brain. In our patient, gliosis in the area of the basal ganglia was diagnosed with the aid of magnetic resonance imaging. The differential diagnosis includes, for example, hemiatrophia faciei progressiva. An attempt at treatment with systemic glucocorticoids and methotrexate is planned.

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