

A Familial Case of Scleroderma en Coup de Sabre

Sir,

Linear scleroderma represents a rare disease, more often observed in the pediatric age group. The lower extremities are usually involved; less commonly it affects the frontoparietal area and the anterior scalp. Recently, a large case series of frontoparietal scleroderma en coup de sabre has been reported (1), but in this series, as well as in the English language literature, to the best of our knowledge, no familial cases have ever been reported. We report herein 2 patients, a 14-year-old girl and her grandfather, affected by frontoparietal scleroderma. Both patients present an atrophic lesion on the midline of the forehead extending from the scalp to the medial side of the eyelids, which is the seat of frontoparietal scleroderma most often reported in the literature (2).

CASE REPORTS

Case 1

A 14-year-old girl consulted us for evaluation of a 2-year history of an atrophic, band-like, pigmented lesion, localized on her right forehead just adjacent to the midline. The lesion at onset appeared erythematous and later evolved into a firm, shiny, pigmented band. There was no redness around the lesion and she did not refer systemic symptoms or signs. Results of laboratory and neurological investigations were within the normal limits and, in particular, no immunological abnormalities were found. Anti-*Borrelia burgdorferi* antibodies were negative.

Case 2

The 67-year-old grandfather of Case 1 presented for evaluation of a band-like region of depressed induration involving his right forehead just adjacent to the midline. The lesion appeared longer than that of his granddaughter. He had first noted a pale lesion on his forehead at age 15 years, then the lesion progressed, in the course of a year, from a reddish plaque to a depressed linear scar. Since then, the patient has not developed any other skin lesion, there has been no systemic involvement and the linear scar has remained totally unchanged. A radiograph showed a linear, tinned band on the skull plate under the area of skin abnormality. The patient refused any other investigation.

DISCUSSION

Few familial cases of morphea have been reported in the literature (3–6) and, to our knowledge, no cases of familial

scleroderma en coup de sabre have been described. Although some data suggest that the scleroderma diathesis may be genetically determined (3), linear scleroderma en coup de sabre is still considered a non-hereditary disorder (6). The distribution of frontoparietal scleroderma which often follows Blaschko's lines is common to other skin diseases such as epidermal naevi, Darier's disease, linear porokeratosis, linear lichen planus, and lichen striatus. The cases reported here, showing an identical distribution pattern of very similar clinical lesions in 2 members of the same family, could suggest that the tendency for the development of scleroderma en coup de sabre is due to genetic mosaicism with the formation of a clone of vulnerable cells. The exposure to a trigger factor may develop clinical manifestations in genetically predisposed individuals.

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