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CASE REPORTS

LINEAR SCLERODERMA IN A FAMILY

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A rare occurrence of localized scleroderma in a mother and daughter is reported. This is perhaps the first reported family in Indian literature.

Key Words : Scleroderma, Morphoea

Introduction

Localized scleroderma is a disorder of unknown aetiology. It's familial occurrence is extremely rare. We report a family in which mother had scleroderma en coup-de-sabre and daughter had linear scleroderma.

Case Reports

Mother : A 35-year old woman noticed slight hyperpigmentation on the right side of the face since the age of 15 years. Later skin over the affected area became firm. A linear groove appeared on the sides of the forehead extending into the scalp. Hair over this area were gradually lost (Fig. 1). These changes did not progress after the initial 5 years.

Examination revealed bound down skin over the right cheek including alae nasi and the frontoparietal region. There was a linear band of alopecia. No other abnormality was found.

Daughter : A 13-year-old girl presented with an area of dried up and shiny skin over her right leg and thigh for the last 3 years.

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On examination, skin over the right shin and thigh was hyperpigmented and bound down. Hair over the affected area were lost. There were areas of patchy hyperkeratosis (Fig. 2). No other abnormality was detected. Histopathological examination of the affected skin in both subjects confirmed the diagnosis of scleroderma.

Comments

Scleroderma in more than one member of a family is very rare. Reports of such cases are only few.

Fig. 1. Localized scleroderma of en coup de sabre type in mother

Rees and Burnett[1] reported localized scleroderma in a father and daughter. Christianson et al[2] studied 235 patients of localized scleroderma and found only 2

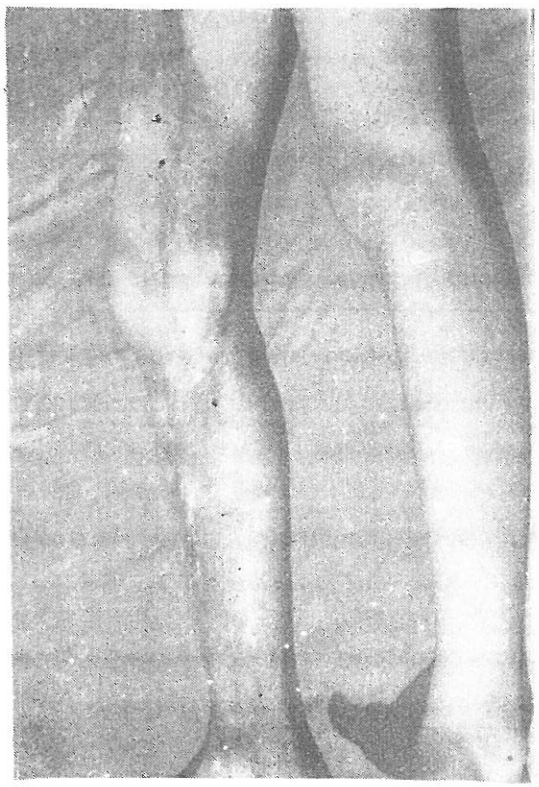

Fig. 2. Linear scleroderma in right leg and thigh of daughter.

instances in which 2 family members were involved. In a review of literature, Burge et al[3] found 13 familial instances of scleroderma occurring in diverse forms and combinations. Wuthrich et al[4] added 2 more families to literature, one having 3 children and the other 2 with localized scleroderma.

The present family is perhaps the first being reported in Indian literature. Localized scleroderma in one member is of en-coup-de sabre type and in the other is of linear type. These reports suggest that there is some genetic predisposition to develop sclerodermatous process.

References

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