

Fig. 8. Morphea tuberosa.

Meeting at University Hospital, Uppsala, April 17, 1970

Morphea Tuberosa-lichen Sclerosus

Presented by Lennart Juhlin

In 1967 this 50-year-old housewife developed a tender swelling in the scapular region which was diagnosed by her local doctor as an infected atheroma. It was treated by forcibly pressing out its grey-white contents. The lesion healed primarily, but after a few months it was replaced by a hard, whitish infiltrate with a diameter of 8–10 cm. Its centre tended to ulcerate and was covered by a fine crust. The patient was then referred to the dermatology clinic.

Upon treatment with 0.2% fluocinolone under occlusion, there was some decrease in its volume. Local application of a gel with dimethylsulfoxide (DMSO) had no definite effect. The lesion does not disturb the patient except for some itching at its periphery.

Histological examination of repeated biopsies showed changes of the type seen in lichen sclerosus et atrophicus rather than sclerodermia. There was also a marked sclerosis of the deeper localized connective tissue. There were no signs of keloid.

Discussion

N. Thyresson: When I first saw the patient 2 years ago I thought she might have dermatofibrosarcoma protuberans, but the histology was that seen in lichen sclerosus et atrophicus. There had

been a tendency to blistering in the centre. Since blisters can occur in both morphea and lichen sclerosus, this does not help us with the diagnosis.

L. Juhlin: The biopsies showed follicular hyperkeratosis and a subepidermal homogenization with some vesiculation as we have seen in lichen sclerosus. In the middle cutis down in the subcutis, there is a pronounced sclerosis with thick irregular collagen fibres giving the impression of morphea. Morphea and lichen sclerosus have been described in the same patient, but it has been doubted by other authors (2). There is no keloid tissue, but macroscopically I would favour a diagnosis of keloid-like scleroderma or morphea tuberosa (1). I should like to hear Dr Lagerholm's view on the histology.

B. Lagerholm: The histological slides fit best with a diagnosis of lichen sclerosus.

References

1. Korting, G. W.: Über keloidartige Sklerodermie. Arch Derm Syph 198: 306, 1954.
2. Steigleder, G. & Raab, F.: Lichen sclerosus et atrophicus. Arch Derm (Chicago) 84: 219, 1961.

Erythrokeratoderma Figurata Variabilis

Presented by Sven Öhman

A 17-year-old boy whose family history revealed no skin diseases. His present lesions first devel-

oped when he was 2 months old. They consisted of sharply demarcated, thin, red plaques with annular, serpinginous outlines. Some had a tendency to central healing. Most of the plaques had a peripheral border of brownish-yellow crusty scales (Fig. 9). The lesions were symmetrically distributed, mainly on the outer side of his upper arms, buttocks, and dorsal parts of his thighs. From time to time there had been some change in extension, size, shape and number. There was usually a slight tendency to improvement during the summertime. He had never noticed any vesicles or bullae, but on a few occasions some oozing. No or very little itching had been present. Hair, nails and teeth appeared normal.

Laboratory studies: Routine analysis of the blood and urine were normal. KOH preparations as well as mycologic cultures were negative.

Biopsy report: Biopsies from arm and thigh show a slight infiltration of lymphocytes and histiocytes subepidermally and some sclerosis of the corium. The changes were non-characteristic.

Treatment: A steroid ointment and urea 10% in hydrophilic ointment have been tried with some temporary benefit.

Discussion

G. Swanbeck: The case does not agree completely with what is described in the literature. Have you stained for fats?

S. Öhman: No.

G. Swanbeck: Could it be angiokeratoma corporis diffusum?

G. Rajka: Usually the changes are more erythematous than is shown here, but this variability as described is exactly characteristic of the disease, which therefore favors Dr Öhman's diagnosis.

Erythro-keratodermia Congenitalis Symmetrica Progressiva?

Presented by Sture Lidén

A 20-month-old girl who, since the age of one month, has had hyperkeratotic skin lesions which

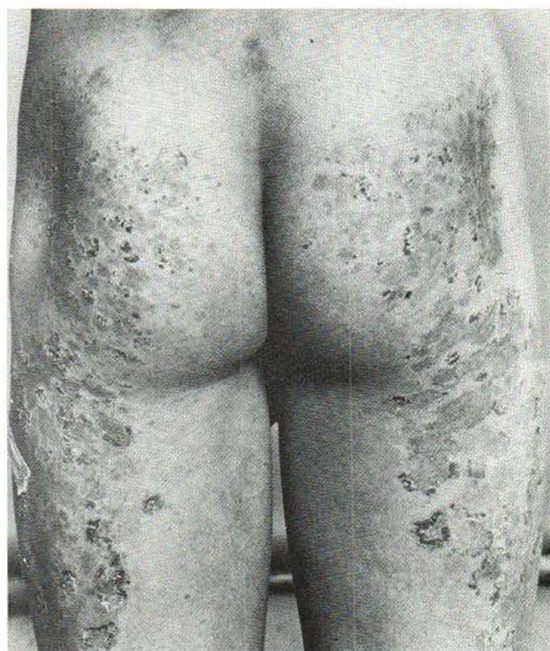


Fig. 9. Erythrokeratodermia figurata variabilis.

have progressively increased in extent. Her grandmother had pronounced thickening of her palmar skin. The child now has partly follicular, acuminate papules surrounded by erythema, and partly grey-brown hyperkeratoses, symmetrically localized, and surrounded by an erythematous zone. Lesions localized to the extremities and head (Fig. 10). The trunk is essentially free.

Extensive hematologic examinations and urine analyses were normal as were the serologic tests for syphilis, metabolic screening, vitamin A determination in serum, and skeletal age determination. Motor age tests revealed a retarded development in the upper extremities. EEG showed increased occurrence of slow rhythms, but not to a definitely pathological extent. Histological examination showed Verrucose epithelial hyperplasia with significant hyperkeratosis and slight spot-shaped interspersions of parakeratosis. In the corium the blood vessels were dilated and, in some places, surrounded by a slight infiltration of lymphocytes. Histologically, the picture cannot be distinguished from verucca plana or epidermodysplasia.