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Lipodystrophia Centrifugalis Sacralis Infantilis

A 15-year Follow-up Observation

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A unique case of Lipodystrophia centrifugalis sacralis infantilis in a caucasian is reported. This case fulfils all the clinical requirements of the centrifugalis lipodystrophy described in Oriental children by Imamura et al. (1) and usually localized on the abdomen. The 15-year follow-up of this case clearly demonstrates the tendency toward spontaneous remission of the disease after puberty.

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In 1971, under the term "Lipodystrophia centrifugalis abdominalis infantilis" Imamura et al. (1) described a peculiar atrophic skin disease characterized by: (*a*) depression of the skin of the abdomen and of the neighbouring regions, due to the loss of subcutaneous fat; (*b*) centrifugal enlargement of the depressed area; (*c*) presence of an inflammatory border surrounding the entire lesion; (*d*) onset before the age of 3 years; (*f*) absence of abnormalities in other organs. Regional lymph node swelling was noted in about 65% of the patients (2, 3). Thirty patients, out of 55 examined, showed complete or partial spontaneous improvement as time passed (4). Up to now, about 90 cases have been reported in Oriental children and only one case in an English caucasian infant (5).

CASE REPORT

In 1973, a 3-year-old Italian girl was seen at our Department of Pediatric Dermatology for a cutaneous depression of the sacrolumbar region, which had appeared at the age of 1 year as a round patch 3 cm in diameter. The lesion quickly lost its inflammatory features and resolved into a centrifugally enlarging depressed area. No trauma or injection had preceded the appearance of the lesion. On clinical examination the patient showed a large oval depressed area with the larger cross diameter of 11 cm and smaller longitudinal diameter of 6 cm on the sacrolumbar region (Fig. 1). A slightly erythematous border surrounded the entire lesion. The underlying blood vessels were visible through the skin. Regional lymphadenopathy was absent. Laboratory findings and X-ray examination of thorax and bones were all within normal limits. The girl's parents refused histological examination and failed to bring her back for further check-ups.

In February 1988, that is to say 15 years after the first observation, the patient returned spontaneously to our Clinic for a mycologic examination. On that occasion, she asked for a check of the lesion in the lumbosacral region. The patient reported that the lesion had continued to enlarge up to the age

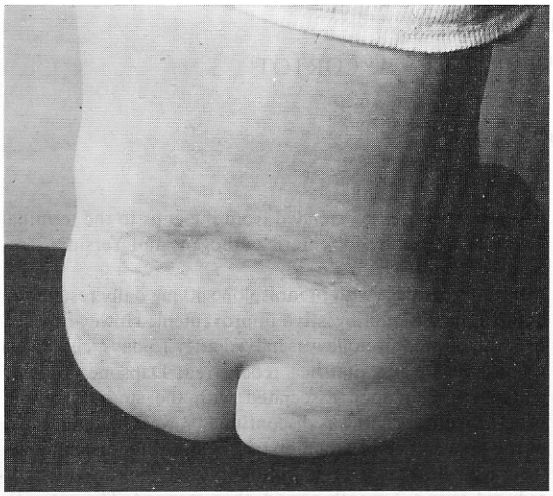

Fig. 1. The patient at the age of 3 years.

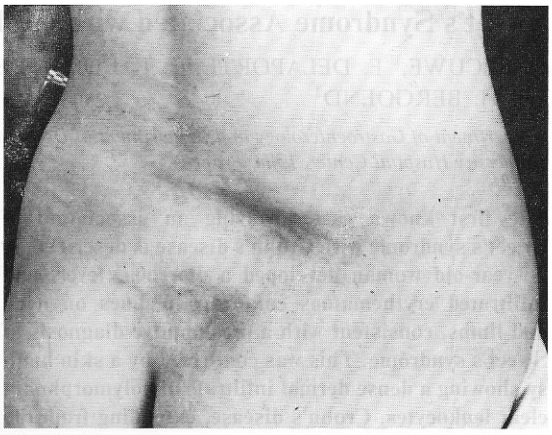

Fig. 2. The patient at the age of 18 years.

of 9 years and that since the age of 15 it had started to improve spontaneously. Clinical examination of the lumbar region showed a triangular slightly depressed area, with no inflammatory margins (Fig. 2) on the right side of the thoraco-lumbar spine. A comparison with the picture taken 15 years earlier (Fig. 1) demonstrated a distinct decrease in size and depth of the lesion. The patient stated that she had never received treatment for the lesion.

COMMENT

The case reported here fulfils all the clinical requirements of the centrifugal lipodystrophy described by Imamura et al. (1, 3, 4), but the lesion appeared on, and remained localized to, the lumbosacral region. The 15-year follow-up of our case clearly demonstrates the tendency toward spontaneous remission of the disease after puberty (4). It should be noted that also in the other case of centrifugalis lipodystrophy described in a caucasian infant (5), the lesion involved the left lumbar region, but extended anteriorly to the abdominal wall.

Our case confirms that the form of centrifugal lipodystrophy described by Imamura et al. (1) is a distinct clinical entity, clearly distinguishable from progressive lipodystrophy forms (1, 5). However, the definition of the disease as Lipodystrophia centrifugalis abdominalis infantilis is questionable, since late onset cases (6) as well as non-abdominal localizations have been described. The cause and pathogenesis of this disease remain unknown. Since the disease shows a predilection for Orientals, the existence of some still obscure racial factor has been hypothesized (4).

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