

A Case of Localized Cutis Marmorata Teleangiectatica Congenita (van Lohuizen Syndrome)

Sir,

Cutis marmorata teleangiectatica congenita was originally described in 1922 by van Lohuizen (1). It is a rare disease consisting of teleangiectasia and venectasia on livedo skin and may be localized or generalized. The skin involved is erythematous and atrophic and may be ulcerated (2).

We report the case of a 5-year-old girl who was presented at our department because of atopic eczema. The patient's father had a café-au-lait spot with naevocellular naevi on his right hip, but no vascular naevi. Otherwise there was no skin disease in the immediate family. At the dermatological investigation, a teleangiectatic, phlebelectatic area of about 6 × 10 cm, localized on the patient's right hip, was noticed (Fig. 1).

According to the patient's mother, this skin change been present since birth. The girl had no symptoms. There was no pain or ulceration in the area. Histologic examination of a biopsy showed an increased number of and enlarged capillaries and veins. Infrared photography of the lesion showed the phlebelectatic dermal and subcutaneous vessels (Fig. 2).

The histologic findings of the van Lohuizen syndrome are not specific but support the clinical findings. In some patients the lesions may be painful, in accordance with the histological finding of an increased number of nerve fibres (3). The cause is unknown. A genetic predisposition has been discussed, as some authors have found similar lesions in family members (4, 5). An association with other abnormalities has been found in 27–50% (6, 7), hypo- or hyperplasia of the affected limb being the most common defect. Other defects include aplasia cutis, heman-

giomas, and phlebelectatic veins, the latter having earlier been revealed by means of near-infrared spectroscopy (3).

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Fig. 1. Teleangiectatica, phlebelectatic lesion on the patient's right hip.



Fig. 2. Demonstration of the enlarged subcutaneous venous vessels by means of infra-red photography.