Naevus Lipomatosus Cutaneus Superficialis: Overlap with Connective Tissue Naevi

Sir,

Naevus lipomatosus cutaneus superficialis (NLCS) is a rare hamartomatous lesion, first described by Hoffmann & Zurhelle in 1921 (1); to date there have been approximately 65 cases reported in the literature. Lesions are characterised by the presence of mature adipose tissue within the dermis. The sex incidence is equal, there is no familial tendency and usually no associated abnormalities. The naevi are usually soft, non-tender, skin-coloured or yellowish papules or nodules, often occurring in a band-like or zosteriform distribution, and with a predilection for the pelvic girdle, particularly the gluteal region. They may be single, multiple or very rarely occur in a generalised form (2). Our case is unusual in that it is, to our knowledge, only the second reported case to involve the knee (3). In addition, it was unusually indurated, had recently increased in size and become symptomatic.

A 49-year-old woman presented with a warty lesion behind the right knee, present since birth but which had gradually increased in size and become painful over a 2-year period (Fig. 1). Previous medical history included longstanding obesity, non-insulin dependent diabetes mellitus and type IV hyperlipidaemia. On examination the lesion was verrucous with surrounding indurated erythema. Investigations revealed a normal full blood count, renal, liver and thyroid function tests. Fasting lipids were raised (total cholesterol 7.9 mmol/l, triglycerides 7.8 mmol/l), as was glycated haemoglobin (8.4% normal range: 3.5–5.5). Histology of the erythematous indurated portion of the lesion showed basket weave hyperkeratosis and mild compact orthokeratosis and acanthosis of the epidermis. There was a marked increase in mature adipose cells throughout the reticular dermis, arranged in clusters and

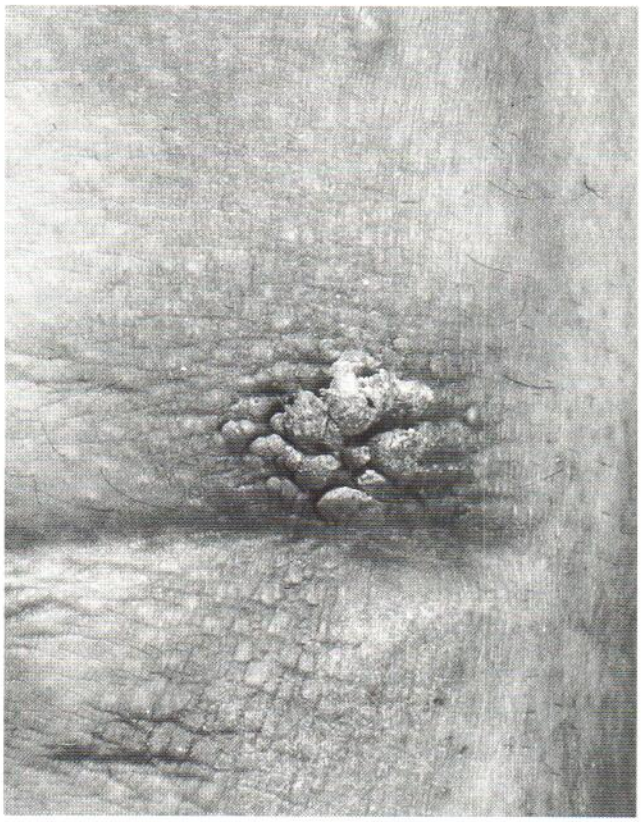

Fig. 1. Warty lesion behind right knee.

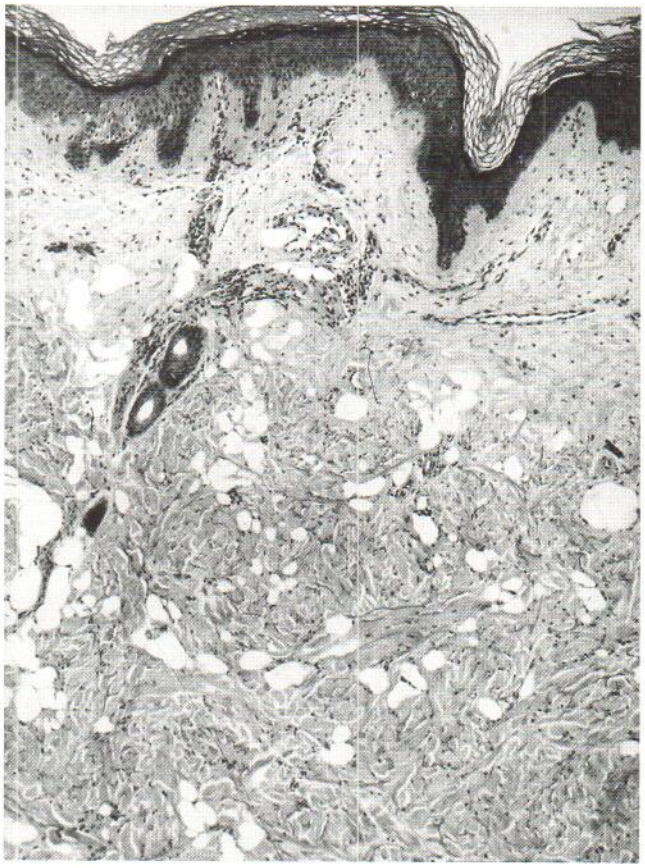

Fig. 2. Naevus lipomatosis: histologic section. Increased amounts of mature adipose cells and blood vessels with a perivascular mononuclear cell infiltrate in the dermis (haematoxylin and eosin).

interspersed by broad interwoven collagen bundles. In addition, increased numbers of blood vessels with a perivascular mononuclear cell infiltrate were present in the subpapillary and reticular dermis, as well as within the foci of ectopic adipocytes (Fig. 2). An elastic Van-Gieson stain showed a reduction in elastic fibres in the superficial part of the lesion and elastic fibre hyperplasia in its deeper portion, as seen in 50% of other reported cases. The verrucous portion showed similar, but less marked changes. These features are all consistent with a diagnosis of NLCS.

The clinical appearance of NLCS is varied. The wide range of clinical diagnoses suspected prior to biopsy includes accessory nipples, lipomata, cellular naevi, connective tissue naevi, naevus sebaceus and warty naevi with comedones (2). Individual lesions may be dome-shaped, sessile or pedunculated, with a smooth, wrinkled, warty, or peau d'orange surface, and there may be follicular plugging or comedones. Lesions develop insidiously from birth or within the first two decades and once formed often remain static, although they have been reported to develop new lobules over the course of many years.

Histologically, in addition to the marked increase in mature adipose cells within the dermis (which can constitute from 10 to 70% of the lesions), there are abnormalities of other connective tissue components. These include thickening of collagen bundles, superficial reduction and deeper increase in elastic fibres, and increased numbers of fibroblasts, mononuclear cells and blood vessels. These features are more suggestive of connective tissue naevi, and Weedon places NLCS within this category (4).

It has also been suggested, however, that NLCS is a hamartomatous lesion arising from blood vessels. This is supported by the fact that in those lesions containing only small amounts of fat, this tends to be localised around subpapillary vessels. In addition there is an inverse relationship between the number of mononuclear cells and mature fat cells around the vessels. Some of these mononuclear cells have been thought to be differentiating lipoblasts (2) and although mature lipocytes have been found perivascularly on electron microscopy, there was no definite evidence of differentiation of vascular cells into adipocytes.

The true origin of this rare naevus remains unclear. Although the most striking abnormality is an excess of dermal fat, which may originate either from vascular endothelial cells or perivascular mesenchymal cells, the presence of varying amounts of other connective tissue components suggests that the lesions of NLCS lie within a spectrum, and that there is considerable overlap with connective tissue naevi.

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CH Orteu, JR Hughes and MHA Rustin
Department of Dermatology, The Royal Free Hospital, Pond Street, London NW3 2QG, United Kingdom.