Acrodermatitis Chronica Atrophicans in an Italian Child

Sir,

Acrodermatitis chronica atrophicans (ACA) is a late manifestation by Lyme disease (1), characterized by bluish-red discoloration and skin atrophy mainly located on acral parts of the body. It is frequently described in elderly and more rarely in children living in Central or Northern Europe (2). This is a report on a child living in Friuli Venezia Giulia, an Italian region where Lyme disease is endemic.

CASE REPORT

An 11-year-old boy presented the following clinical picture: large areas of white pearly sclero-atrophic skin on the back of hands and feet; on the dorsal aspect of the legs brownish, indurated and hardly extensible skin; large erythematous areas with mildly thinned skin on wrists, right scapular region and neck. The cutaneous manifestations had been present for 4 months, beginning with a mild oedema on the dorsal aspect of the feet. No history of past erythema chronicum migrans was reported. The investigation of IgG antibodies against Borrelia burgdorferi (BB) revealed a positive titre of 1:256. The search for BB-specific DNA in tissue by polymerase chain reaction was positive. A treatment with intravenous penicillin G (20 millions units daily for 20 days) was performed. The erythematous atrophic lesions healed shortly after therapy, whereas large pimentero- atrophic areas still persisted after 6 months. The IgG titre declined.

DISCUSSION

In Italy there are some geographic areas where Lyme disease is endemic: Liguria, Friuli Venezia Giulia and Trentino Alto Adige (3). In the last years a few cases of ACA have been described in these regions, but no cases have been reported in childhood. Only Trevisan et al. (4) observed a peculiar picture in a 6-year-old child, who showed an atrophodermic lesion shortly after erythema chronicum migrans localized on the mammary area.

The differences in the clinical pictures observed in different regions where Lyme disease is endemic may be related to the unequal distribution of BB genospecies among different areas. In Italy Borrelia Garinii seems to be more frequently encountered than Borrelia Afzelii, usually found in subjects affected by ACA.

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Naevus Lipomatosus Cutaneus Superficialis: Overlap with Connective Tissue Naevi

Sir,

Naevus lipomatosus cutaneus superficialis (NLCS) is a rare hamartomatic 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 glycaated 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

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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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Multiple Symmetrical Lipomatosis in a Patient on Long-term Corticosteroid Therapy

Sir,

Multiple symmetrical lipomatosis (MSL) is an uncommon disorder of unknown aetiology, characterised by large non-encapsulated lipomas distributed around the neck, shoulders and other axial regions (1).

CASE REPORT

A 56-year-old male presented with a 20-year history of diffuse lipomatous growths around the shoulders, arms, upper trunk and thighs. The fatty accumulations had been rapidly progressive for the first 4 years and gradually increased over the next 16 years. There was no family history of a similar disease. The patient was a non-alcoholic and had no known diabetes. In addition, the patient had been suffering from chronic actinic dermatitis for the last 30 years and had received oral corticosteroids in doses up to 40 mg of prednisolone equivalent daily. The patient was normotensive. Cutaneous examination revealed diffuse, non-tender, lipomatous growths restricted to the shoulders, upper chest, arms, trunk and thighs (Figs. 1, 2). The face and distal part of extremities were conspicuously spared. Side-effects of corticosteroid therapy in the form of skin atrophy with striae were present on the lower trunk and calf area. The patient, however, did not have "moon-faces" or "buffalo-hump". There was no clinical evidence of superior venacaval obstruction, respiratory complaints or peripheral neuropathy.

Erythematous scaly papules and plaques were present on the face and extensors of the forearms when the patient was first seen in the outpatient department. These subsided within 2 weeks of hospitalisation with photoprotection and topical application of betamethasone valerate (0.1%) ointment. Oral corticosteroids were gradually tapered and stopped over a period of 4 weeks.

On examination, the patient was found to be suffering from diabetes mellitus. Skin biopsy from one of the growths revealed a non-encapsulated lipoma. Electrophysiological study of the nerves was suggestive of sensorimotor neuropathy involving both upper and lower limbs. Routine haemogram, liver and kidney function tests were within normal limits and so were his serum cortisol and uric acid levels. A skiagram of the chest and ultrasonographic study of the abdomen revealed no abnormality.

DISCUSSION

Two patterns of distribution of lipomas have been described in MSL (2). In the type I variant lipomatous deposits are found primarily in the nape of the neck, supravacular and deltoid region, while in the type II variant lipomas are not localised to the neck but extend down over the trunk and proximal part of extremities, whereas the lower part of the trunk and lower extremities are conspicuously spared (2). Mediastinal extension of lipomas with obstructive symptoms due to pressure on trachea and superior vena cava have been described in the type I pattern (3). The disorder is frequently associated with alcoholic habit, peripheral neuropathy and deranged liver functions (1). Rarely association with gout, hypertension, obesity, diabetes and various neurological symptoms has been reported (4, 5). Certain metabolic abnormalities in the form of hyperuricemia, hypertriglyceridemia and hyperinsulinism may be found (2, 4). However, there is little convincing evidence that these associations are other than due to chance.

Our patient had a clinical presentation consistent with type II MSL. On examination, the patient was found to have diabetes mellitus and peripheral neuropathy, commonly associated with MSL. However, alcoholic habit frequently reported

Fig. 2. Lipomatous deposits on the trunk.

Fig. 1. Patient having lipomatous deposits involving shoulders, arms and trunk.