

Linear Scleroderma Associated with Hypertrichosis in the Absence of Melorheostosis

Sir,

Hypertrichosis, an increase in non-androgen-modulated hair on the body, may be congenital or acquired, localized or generalized. It may be limited to cosmetic significance or may represent a cutaneous sign of underlying systemic disease (1). There are a few reports of hypertrichosis on the indurated skin in melorheostotic scleroderma (2–4). We describe here a 12-year-old Korean boy with linear scleroderma associated with hypertrichosis in the absence of melorheostosis (thickening of cortex of long bones).

CASE REPORT

A 12-year-old Korean boy presented with an asymptomatic hard lesion and hypertrichosis of the skin on the left buttock and lower extremity. The lesion had developed slowly 5 years earlier and 2 years later hypertrichosis developed within this lesion. He had no apparent pain or disability and no history of trauma, contact with any irritants, or application of topical steroid ointment to the area.

Examination of the skin lesion revealed hypertrichosis on the linear firm ivory-coloured lesion, 30–70 mm in width, extending posterolaterally from the left buttock to the calf (Fig. 1). There was no inflammatory halo and hyperpigmentation. The involved skin felt taut and unyielding. No limitation of motion of the left lower extremity was found.

The findings of laboratory evaluation, including complete blood



Fig. 1. Lesions on the left lower extremity, showing the asymptomatic hardening ivory-coloured patch (arrows) and hypertrichosis.

count, liver function test, urinalysis, RA factor, antinuclear antibody, anti-ss-DNA antibody, anti-scl-70 antibody, anticentromere antibody and chest radiography, were within normal limits or negative.

Roentgenographic findings of both lower extremities showed no bony abnormalities or leg length discrepancy.

Histopathological examination of the hypertrichotic region on the left thigh revealed irregular thick collagen bundles and downward proliferation of collagen into subcutaneous fat. The collagen fibres were swollen and eosinophilic. Verhoeff-van Gieson stain showed normal elastic fibres. Based on these findings, a diagnosis of linear scleroderma associated with hypertrichosis was made.

The patient was treated with penicillamine for 6 months, but did not show remarkable improvement.

DISCUSSION

Contact or nummular eczema, thrombophlebitis or venous insufficiency, pretibial myxedema, arthritis or osteomyelitis, occupational or self-induced (e.g. biting) trauma, and occlusion from a plaster cast have been associated with hypertrichosis. It may be related to increased cutaneous blood flow or deposition of exogenous material in the dermis (1).

Until now, only a few cases of melorheostotic scleroderma associated with hypertrichosis have been reported (2–4). Melorheostosis is a rare disorder of bone characterized roentgenographically by linear hyperostosis that appear to flow down the cortex of the bone (5).

Regarding the pathogenesis of linear melorheostotic scleroderma, Wagers et al. (4) believe that skin changes are representative of the same proliferative disorder which produce the bony hyperostosis. Inflammatory (5) and vascular (6) theories have been proposed. Muller & Henderson (7) postulate that linear sclerodermatous changes are a primary mesenchymal defect that occasionally spills over into the skeletal tissues. However, many others favour a hypothesis of developmental error (8). If skin involvement occurs from a localized proliferative disorder, such as mesodermal dysplasia or an osteocutaneous syndrome, then excessive hair growth in the affected area probably has a similar origin and is not secondary to the bone disorder. Embryological research has shown that hair germ cells originate in epidermal basal cells, which are an ectodermic component (9). If the hypertrichosis has the same origin as the bony abnormality and associated scleroderma, it may be speculated that the melorheostosis probably represents a congenital disorder with both ectodermic and mesodermic components (2). The patient described here had not been receiving any of the medications known to induce hypertrichosis and he did not have any infectious, rheumatological, or vascular conditions for which hypertrichosis may be an associated adverse sequelae. The hypertrichosis was limited to the scleroderma lesion in the absence of melorheostotic change. This finding suggests that there is a direct relationship between linear scleroderma and hypertrichosis.

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Eosinophil Activation in Wells' Syndrome Demonstrated Immunohistochemically with Antibodies Against Eosinophilic Cationic Protein

Sir,

The important role of the eosinophilic granulocyte has been recognized in a number of skin disorders. The mediators released are especially important in defence against parasites and bacteria, but the toxic proteins may also cause damage to keratinocytes (1). The number of eosinophilic granulocytes in peripheral blood and tissue has been reported to correlate well with clinical severity in disorders such as allergic bronchial asthma, allergic rhinitis and atopic eczema (2). The clinical effects are thought to be mediated partly through cationic proteins released from the eosinophils (3). The granules in the eosinophilic granulocytes have been shown to contain 4 predominant cationic molecules with cytotoxic properties, including eosinophilic peroxidase, major basic protein, eosinophilic cationic protein and eosinophil-derived neurotoxin (4).

Recurrent granulomatous dermatitis with eosinophilia was first described by Wells in 1971 (5). Typically, single or multiple lesions erupt on a limb, the face or the trunk, after a short prodromal itching or burning, and blistering may occur. The skin is usually restored after a few weeks, although recurrences over many years are common (6).

Histopathologically, different stages have been described; in the acute stage the dermis is infiltrated with leukocytes, especially eosinophils, and shows oedema. In the subacute phase histiocytes together with eosinophils adhering to collagen bundles ("flame figures") dominate the picture, and in the later stage microgranulomas with histiocytes are present (7, 8).

In order to investigate the role of eosinophilic proteins in Wells' syndrome, biopsies from 2 patients with this rare skin disorder were reacted immunohistochemically with antibodies against the eosinophilic proteins eosinophil-derived neurotoxin, eosinophilic peroxidase and eosinophilic cationic protein.

PATIENTS AND METHODS

Patient 1

A 12-year-old boy presented at the department with bluish-erythematous infiltrations on his trunk and extremities of 1–5 cm in diameter. There were no general symptoms, no fever, but marked

peripheral blood eosinophilia ($550 \times 10^9/l$). Histopathological investigation of a 3-mm punch biopsy revealed infiltration of eosinophilic granulocytes around vessels, but also between collagen fibres and extending down to the subcutaneous tissue. Together with the clinical picture, an early stage of eosinophilic cellulitis was diagnosed. As he did not suffer from general symptoms, he was observed, and on the clinical follow-up after 3 weeks presented only post-inflammatory hyperpigmentations.

Patient 2

A 43-year-old woman had experienced erythematous papules and dermal nodules on the face for about 12 months, associated with fever, malaise and peripheral blood eosinophilia ($460 \times 10^9/l$). The histopathological investigation of a 4-mm punch biopsy specimen showed a diffuse and perivascular infiltration of lymphocytes, histiocytes, and numerous eosinophils. In one section neutrophilic and eosinophilic granulocytes centred around a granular eosinophilic degenerated material, consistent with flame figures were seen. In all, and considering the clinical course, a subacute stage of eosinophilic cellulitis was diagnosed. She received treatment with oral steroids starting with 30 mg daily for 4 weeks, and after 1 month showed only scattered erythematous macules on the face, but no dermal nodules or infiltration.

Immunohistochemical procedure

Immunohistochemistry was performed on sections from the formalin-fixed, paraffin-embedded biopsies from these 2 patients. Serial sections 6 μ m thick were de-waxed, trypsinated and incubated for 12 h at room temperature with antibodies against eosinophilic cationic protein (ECP; EG-2, dilution 1/320; Pharmacia, Sweden), and with antibodies against eosinophilic peroxidase and eosinophil-derived neurotoxin (both kindly supplied by Pharmacia, Sweden). After washing, antigen localization was accomplished by the alkaline phosphatase-anti-alkaline phosphatase (APAAP) method (9), using the APAAP-complex (Dakopatts, Denmark) in dilution 1/40. The sections were counterstained with haematoxylin and mounted in Apathy's mounting medium (RA Lamb Lab., London, UK).

RESULTS AND DISCUSSION

In the conventional histopathology, eosinophils were seen extending into the subcutaneous tissue in patient 1, and in the dermis and around vessels in patient 2. Immunohistologically,