Primary atrophic profound linear scleroderma

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SUMMARY

We describe an 18-year-old male with primary atrophic profound linear scleroderma. It was not preceded by an inflammatory reaction or sclerosis, but it involved the subcutaneous and deeper tissues of the fingers. It did not involve the dermis, or show either discoloration or changes in texture. Involvement of deeper tissue and the progression of the disease suggest atypical primary atrophic profound linear scleroderma. Our case of primary atrophic profound linear scleroderma appears to be a unique variant of localized scleroderma.

KEY WORDS

scleroderma, circumscribed, profound, linear, atrophic, male patient

Introduction

In 2000 Blaszczyk and associates (1) delineated primary atrophic profound linear scleroderma as a nosologic entity. It is characterized by areas of deep atrophy involving the subcutis and deeper tissues, which are not preceded by inflammatory changes. In contrast to typical profound scleroderma, the variant is not associated with generalized disfiguration and crippling deformities. Cases have been described with areas of involvement including the arm, the leg, the scapular region and the buttocks (1). We describe an 18-year-old male patient with atropic profound linear scleroderma of the fingers.

Case report

History

The patient's parents first noticed skin indurations involving the middle and fourth fingers of the left hand following the development of a small crack in the skin of the middle finger resulting from injury caused by a wood chip. Over the following three months, a triangular patch of indurated skin developed on the dorsum of the left hand as well. A biopsy of the skin lesion including the dorsal aspect of the left hand failed to confirm the diagnosis of scleroderma; the direct immunofluorescence test (DIF) was negative. The patient was pla-

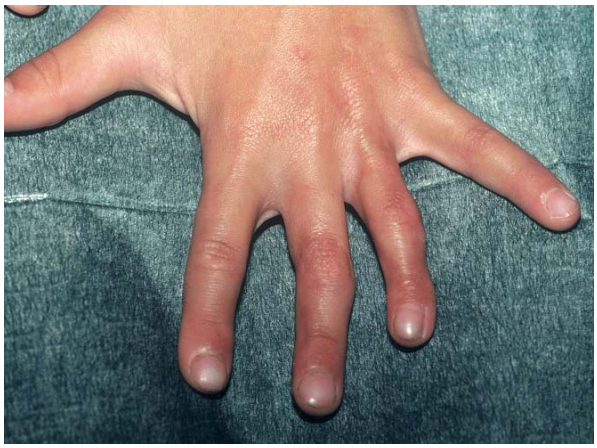

Fig. 1 Left hand: radial deviation of the third and fourth finger, atrophy of the skin on the dorsal aspect of the hand and on the mentioned fingers.

ced on a 10-day course of chloramphenicol due to infection with R conori, that had been identified on serological examination. Serology for Borrelia burgdorferi (B. burgdorferi) was negative.

About seven months later the patient developed new sclerodermiform foci on the upper sternum and the left axilla, lesions on the inner aspect of the left wrist, contracture of the left index finger, and later on, contracture of the small finger. Eosinophilic fasciitis was suspected, yet the disease had begun about a year before, and there was no eosinophilia. The diagnosis was re-evaluated by means of a biopsy of the skin, subcutaneous tissue and fascia obtained from the focus on the sternum. The histopathologic results were compatible with scleroderma (Biopsy 329/90). He was treated with benzyl-penicillin 800,000 I.U. for 21 days, and prednisolone 5 mg/day and Ospen q.i.d. for one month. The lesions regressed slowly during the following two years: finger contractures were ameliorated following physiotherapy, while the sclerotic lesions involving the dorsum of the left hand, sternum and axilla regressed completely. There remained however a persisting atrophy of the tissue on the interdigital areas of the let hand, and the growth of the small finger on the left hand was delayed.

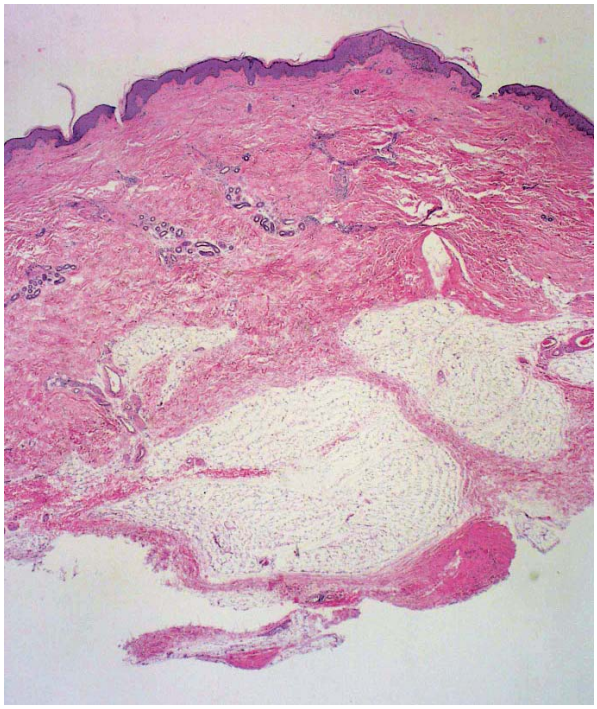

Fig 2. Biopsy 329/90. In the lower layers of the somewhat thickened epidermis there is a slight increase of brownish pigment. The dermis is thickened. Homogenized and swollen collagen bundles in certain areas are extending into the subcutis. Pilosebaceus units are absent, sweat glands are surrounded by collagen bundles (x 100).

About one year later a hyperpigmented spot, 2x1 cm in size, developed on the right forearm. The patient had previously reported a pain in the same area. Eosinophilia was established at that point. The hyperpigmented patch on the right forearm soon spread towards the elbow pit, and the veins were visible through the skin. Since the serological test for B. burgdorferi was borderline positive, the patient was given two courses of ceftriaxone (Lendacin) over the next five years. The PCR test of a skin sample was positive. Skin lesions during this period remained unchanged showing neither improvement nor deterioration.

At the end of the five-year period, deformities involving the middle phalanges of the right hand developed, identical to those on the left hand. The hyperpigmented spot increased in size. Prominent veins were visible through the transparent, soft and non-sclerosed skin. The histological findings at the time suggested acrodermatitis chronica atrophicans (Biopsy 97/98). Also at this time, small ulcerations developed on the fingertips. An extensive battery of diagnostic tests, including cryoglobulin determination and capillaroscopy, disclosed nothing of note. Neurological tests, including EEG and EMG, carried out the following year, were within normal limits. A magnetic resonance imaging (MR) scan of the left hand showed signs of inflammation.

During the past year and a half, the skin lesions have spread: the area that presents translucent veins has increased in size, and extended from the forearm to the mid-upper arm. New hyperpigmented patches have appeared on the forearms; and there has been incipient induration of the skin on the dorsum of the right hand and interphalangeal deformities involving the fingers of the right hand.

Physical examination

Examination disclosed radial deviation of the third and fourth fingers, atrophy of fingers involving the

middle interphalangeal joint and a teleangiectasis under the nail plate. Koilonychia was present on the middle finger. There was also atrophy of the skin on the dorsal aspect of the left hand with hyperpigmented patches extending from the upper extremity to the axilla and right mammilla. The back of the left hand was atrophic with hyperpigmented areas spreading to the flexor aspect of the forearm and arm. Three hypertrophic scars were noted, i.e. the largest pre-sternal scar secondary to diagnostic excision, and two smaller ones on the right forearm and the dorsum of the right hand.

Histopathology

Biopsy 329/90 pectoral skin

In the lower layers of the somewhat thickened epidermis there is a slight increase of brownish pigment. The dermis is also thickened. The collagen bundles are slightly thickened, and in certain areas they extend into the subcutis. In some places of the lower dermis there are prominent homogenized and swollen collagen bundles. Orcein staining revealed that elastic fibers are present throughout the dermis and the subcutaneous septa.

Pilosebaceous units are absent, save some remains of arrector pili muscles. Collagen has replaced the fat around the sweat glands which are situated at a relatively high level, but otherwise appear normal.

The walls of some blood vessels appear slightly thickened, however vascular changes are not prominent.

A sparse inflammatory cell infiltrate is distributed around the blood vessels, in some parts more diffusely through the dermis. It is composed mostly of lymphocytes with some macrophages and rare plasma cells and eosinophils, and is more pronounced in the lower part of the dermis and subcutis.

The histologic pattern suggests a diagnosis of scleroderma.

Biopsy 97 /98 left forearm

Orthokeratosis. A moderate atrophy of the epidermis, more pronounced in one area, where the epidermal border is flattened. The sweat glands are situated high in the dermis, where only a single follicle is present. A perivascular and partially interstitial infiltrate is also to be observed. In some areas the collagen fibers have thickened and there are signs of sclerosation, the fibroblasts are sparse or missing, and here has been an occasional fragmentation of elastic fibers. The subcutis is missing.

It is difficult to diagnose whether this is acrodermatitis atrophicans chronica or scleroderma, but the incipient sclerosation of the dermis favors a diagnosis of scleroderma (morphea).

Biopsy 566 /2002 right forearm

Mild hyperplasia of the epidermis with pronounced pigmentation of the lower layers and a mild vacuolar degeneration in some areas. There is a discrete and mostly perivascular - but partly also periadnexal - mononuclear infiltrate consisting of lymphocytes and cells with hyperchromatic nuclei of irregular shape. Several dendritic and stellate cells (probably fibroblasts) are observable in the interstitium. In the central part of the dermis there are thickened and hyalinized collagen bundles with narrow inter-fibrilar spaces. Elastic fibers are present in orcein-stained sections.

The section comprises the epidermis with the corresponding dermis and is not suitable for a complete analysis. The above-mentioned area of thickened collagen fibers is suggestive and compatible with scleroderma, but it also corresponds to the diagnosis of atrophodermia as described by Pierini and Pasini. The histological findings, however, are not specific.

Reevaluation and synopsis of histopathological findings

The 329/90 is the only representative biopsy to comprise the subcutis. After analysis, the biopsy favors a diagnosis of *scleroderma*. It is inappropriate to make a diagnosis of *sclerodermia profunda* on account of the missing fascia, even though the thickened collagen fibers in the lower parts of the section are typical of that condition.

Discussion

Localized scleroderma is a relatively uncommon disorder, and includes multiple variations. Circumscribed, sclerotic, indurated plaques with white centers, which are not fixed to underlying structures, are typical of localized morphea. Morphea profunda (subcutaneous morphea) is a poorly defined form of localized scleroderma characterized by deep, bound down plaques (2). Its onset has been described in one case as a progression from deep nodular vasculitis, the regression of whose vascular lesions is associated with the onset of sclerotic changes in the cutis (3). Profound morphea has been associated with high titers of antifibronectin antibodies in cases where nodular lesions occur within plaques (4). Morphea may occur without underlying sclerosis: superficial atrophic hyperpigmented plaques are characteristic of the atrophoderma of Pierini and Pasini, which has a chronic benign course.

Linear scleroderma occurs most often during childhood. It is characterized by a linear band, affecting both the superficial as well as the deeper layers of the skin, often with a fixation to the underlying structures. Calcinosis may appear within lesions as hard white papules

(5). This form of scleroderma is usually unilateral, and is most often localized at the extremities and the face. When it occurs at the extremities, the band-like lesions may cause a contracture of the fingers that limits their range of motion (4). The occurrence of linear depression on the forehead has been termed *coup de sabre*; it appears as a furrow in the skin and is the result of a localized atrophy. When the atrophy is extensive, the facial hemiatrophy can result in a significant deformity. The facial hemiatrophy may be deeper than that seen in *coup de sabre*; the facial dermis as well as subcutaneous fat, muscle and even bone are atrophic in Parry - Romberg syndrome (6). Its relationship with linear scleroderma is debated (4).

The linear atrophoderma of Moulin consists of superficial linear atrophies, with minimal depression and no associated inflammation. These atrophic streaks are usually unilateral and follow Blaschko's lines (7). Similar to the atrophoderma of Pierini and Pasini, it has a stable, benign course; indeed the atrophoderma of Moulin is considered by some to be a linear variant of the atrophoderma of Pierini and Pasini (8,9).

While the atrophy is profound involving the subcutis and deeper tissues, the dermis is not affected, with no discoloration or change in texture. Profound linear atrophies are thought to have a greater relationship to scleroderma than to the atrophoderma of Moulin. In addition to its obvious contrast to the characteristically superficial atrophoderma of Moulin, linear scleroderma has a zonal distribution and is not believed to follow Blaschko's lines (the atrophoderma of Moulin most typically follows a Blaschkoid distribution). In one of the cases of primary atrophic profound linear scleroderma reported by Blaszcyk and collegues (1), it occurred alongside a partial facial hemiatrophy, which is considered a variant of linear scleroderma. In conclusion, our case of deep atrophy of the fingers with no dermal involvement is a unique atrophic variant of linear scleroderma which we have referred to as primary atrophic profound linear scleroderma.

Conclusion

Our case of primary atrophic profound linear scleroderma appears to be a unique variant of localized scleroderma. In contrast to typical profound scleroderma, neither inflammation nor sclerosis followed the onset of atrophy. A possible diagnosis is an atypical variant of the Parry Romberg beginning on the extremities. The fact that the subcutis was present only in the first biopsy may be due to the progression of the disease. It is reasonable to expect that the follow-up will enable us to establish the final diagnosis.

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