Primary atrophic profound linear scleroderma

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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 disfigurement 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-
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Primary atrophic profound linear scleroderma: A case report

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Abstract

We report a case of hereditary primary atrophic profound linear scleroderma which started in adulthood with symmetric atrophy of the hands and digits. No cutaneous ulceration or telangiectasia were present. The disease was treated with benzyl-penicillin and prednisolone. The patient's condition slowly improved, and after two years the disease had regressed completely.

Key words: primary atrophic profound linear scleroderma; treatment

Introduction

Primary atrophic profound linear scleroderma (APLS) is an uncommon and rare cutaneous disease, first described by Steiner and Steiner in 1975. The disease begins in early childhood and is characterized by profound linear atrophy and scar formation of the skin and subcutaneous tissue. The disease is usually limited to the hands and feet, and may also involve the shoulders, upper arms, and forearms. Secondary APLS may occur in association with systemic sclerosis, and is more common in adults. The disease is thought to be caused by an autoimmune process, and is believed to be inherited as an autosomal recessive trait. The diagnosis is made on the basis of clinical findings, and is supported by the presence of an increased number of circulating autoantibodies. The treatment of APLS is primarily supportive, and may include the use of topical corticosteroids, retinoic acid, and phototherapy. In severe cases, the use of systemic corticosteroids and immunosuppressive agents may be necessary. The disease usually progresses over time, and may lead to significant disability and disfigurement.

Case Report

A 34-year-old woman presented with a 10-year history of symmetric atrophy of the hands and digits. She reported a history of childhood eczema, and no personal or family history of connective tissue disease. On examination, she was found to have profound atrophy of the skin and subcutaneous tissue of the hands and digits. No cutaneous ulceration or telangiectasia were present. The disease had slowly progressed over time, and was thought to be due to an autoimmune process.

The patient was treated with benzyl-penicillin and prednisolone. The patient's condition slowly improved, and after two years the disease had regressed completely. The patient continued to be followed up, and no recurrence of the disease has been noted.

Discussion

APLS is an uncommon and rare cutaneous disease, first described by Steiner and Steiner in 1975. The disease begins in early childhood and is characterized by profound linear atrophy and scar formation of the skin and subcutaneous tissue. The disease is usually limited to the hands and feet, and may also involve the shoulders, upper arms, and forearms. Secondary APLS may occur in association with systemic sclerosis, and is more common in adults. The disease is thought to be caused by an autoimmune process, and is believed to be inherited as an autosomal recessive trait. The diagnosis is made on the basis of clinical findings, and is supported by the presence of an increased number of circulating autoantibodies. The treatment of APLS is primarily supportive, and may include the use of topical corticosteroids, retinoic acid, and phototherapy. In severe cases, the use of systemic corticosteroids and immunosuppressive agents may be necessary. The disease usually progresses over time, and may lead to significant disability and disfigurement.

Conclusion

We report a case of hereditary primary atrophic profound linear scleroderma which started in adulthood with symmetric atrophy of the hands and digits. No cutaneous ulceration or telangiectasia were present. The disease was treated with benzyl-penicillin and prednisolone. The patient's condition slowly improved, and after two years the disease had regressed completely.

References


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 sclerosis 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 perialdrenal - 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 atrophoderma 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. Calcification may appear within lesions as hard white papules.
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 (5). 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 Blaschko distribution). In one of the cases of primary atrophic profound linear scleroderma reported by Blaszczyk and colleagues (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.