

Atrophoderma of Pasini and Pierini in a young adult: a case report

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Abstract

Atrophoderma of Pasini and Pierini is a skin atrophy presenting as single or multiple sharply demarcated, hyperpigmented, non-inflamed patches, with a slight depression of the skin, that can converge and form a confluent area with atrophy as a consequence. The condition was first described by Pasini in 1923 and subsequently by Pierini in 1936. They distinguished this form of atrophy from other diseases and conditions in which the atrophy is morphologically and clinically different. The disease was initially associated with *Borrelia burgdorferi* infection; however, at present, various theories have emerged for the appearance of the disease, linked to genetic, neurogenetic, and immunological factors. Here we present a patient that was admitted to the hospital due to disseminated lesions on the skin of the lower limbs, with slightly pigmented and atrophic skin along with irregular borders varying in size, from several mm to a few cm, clearly demarcated from the healthy skin, with no history of a tick bite or a family history of similar skin disorders.

Keywords: atrophoderma of Pasini and Pierini, atrophy, hyperpigmentation

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Introduction

Atrophoderma of Pasini and Pierini is a skin disease manifested with depressed skin in areas that have a histopathology confirmed as atrophy with hyperpigmentation. The disease was first described by Pasini and later by Pierini and Vivoli (1). At that time the disease was linked to localized scleroderma, and in 1958 it was classified as idiopathic atrophoderma (2). Youkoyama et al. discovered that the glycosaminoglycans found in atrophoderma of Pasini and Pierini were different from the ones observed in morphea (3). The exact cause of atrophoderma as described by Pasini and Pierini remains unknown (1, 4–9). It is a disease that is more common in adolescent and middle-aged females, although there have been cases described in children and in elderly patients. Furthermore, it has been reported that this disease can be contracted at birth.

Case report

Here we report a case study of a 20-year-old man attending our outpatient clinic with a 2-year history of slightly depressed hyperpigmented patches of the skin of the lower limbs. The patient had noticed that the changes were more visible during autumn and winter, whereas in summer the skin patches become less intense in color. The patient was initially treated as an outpatient with local corticosteroids, and nourishing and neutral creams. The patient was subsequently admitted, presenting with disseminated lesions on the skin of the lower limbs (Fig. 1), with slightly pigmented and atrophic skin along with irregular borders. The skin lesions varied in size from a few mm to several cm and were clearly demarcated from the surrounding healthy skin. After admission to the hospital, we carried out the following analyses: sedimentation, full blood count, urea, creatinine, hepatogram, transaminases, anti-DNA, antinuclear antibody (ANA), LE cell, Scl-70, CRP, and serological test for *Borrelia*. All parameters were within reference ranges. The only collateral finding that we noticed was subclinical Hashimoto's thyroiditis with a normal level of thyroid

hormones and very high levels of anti-TPO (1,200 in a reference range of < 30 IU/ml). The previously unidentified thyroiditis was detected by a dermatologist during the hospitalization at our clinic because the patient did not have any noticeable symptoms before admission. Moreover, a detailed skin biopsy report, obtained via standard pathology diagnostics services, revealed flattening of the dermal papillae rete ridges, perivascular, perifollicular lymphocytic infiltrate, and clumping of collagen fibers.



Figure 1 | Atrophoderma of Pasini and Pierini: disseminated lesions on the skin of the lower limbs.



Figure 2 | Atrophoderma of Pasini and Pierini: pigmented and atrophic skin characterized by irregular borders.

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Case analysis including histological findings suggested that the patient was suffering from atrophoderma of Pasini and Pierini. The patient was discharged from the hospital with advice to apply cream containing topical steroids, having rejected our suggestions for therapy with hydroxychloroquine or retinoids. During routine follow-up and regular controls we did not notice any improvement of the patient's condition.

Discussion

Atrophoderma of Pasini and Pierini is a disease with an unknown cause (2, 4), even though in some cases *Borrelia burgdorferi* was the primary factor as a cause due to findings of high *Borrelia* antibody titers in some patients with atrophoderma of Pasini and Pierini (10). In this particular case, serological tests for *Borrelia* were negative. Nevertheless, the role of *Borrelia burgdorferi* infection in the pathogenesis of atrophoderma of Pasini and Pierini remains disputable. Some studies suggest that a neurogenic cause, immunological factors, and genetic predisposition may play significant roles in the appearance of the disease (11, 12). A connection with morphea was also determined due to the similarity of our findings in the skin (13) because we found no evidence of an inflammatory process. The classification of atrophoderma is a challenge in medical literature, and differential diagnostics should include other disorders identified by skin atrophy. Some authors have classified atrophoderma as a disease with a long course and a unique appearance without signs that resemble morphea, including characteristic lilac rings (13), which were ob-

served in the case presented. The disease is characterized by a single or sometimes multiple confluent depressed hyperpigmented atrophic scars appearing in patches, with a distinctive border known as the "cliff-drop" border. The typical area affected is the back, but other regions can be affected and there is significant symmetry. In our case, symmetry was evident but the location of the disease was unusual because the changes were concentrated on the lower extremities and numbered more than 50. The disease mostly appears in early adolescence and it affects females more than males (5–9). These cases describe the disease in children and elderly female patients, whereas here we present a case of atrophoderma of Pasini and Pierini in an adult male. Laboratory evaluations were within normal range and histological examination revealed a characteristic decrease of dermal thickness. The course of the disease is usually benign, but the poor response to conventional therapy is considered a problem for both the patient and the physician (10, 14). Skin changes that persist and remain the same size over time nonetheless sometimes develop further and increase in both number and size for 10 to 20 years without a significant improvement despite treatment.

The diagnosis is based on clinical findings and skin biopsy (13). However, treatment does not show significant results. Some positive results have been achieved with retinoids and topical steroids. In some cases there have also been good reports on administration of hydroxychloroquine (14). In cases in which *Borrelia burgdorferi* infection is documented, a course of antibiotics should also be prescribed (10).

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