Atrophoderma of Pasini and Pierini in a patient with concomitant psoriasis: Response to methotrexate

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INTRODUCTION

Pasini and Pierini described a form of atrophoderma in 1923 and 1936, respectively.[1] Today, idiopathic atrophoderma of Pasini and Pierini (IAPP) is mostly considered a rare, distinctive form of plaque morphea with approximately 100 reported cases.[2] The trunk is commonly involved with usually symptomless round or ovoid lesions possibly coalescing to large irregularly shaped atrophic patches. Sharply defined depressed patches of different sizes with cliff-drop borders are a diagnostic clue. Most cases of atrophoderma of Pasini and Pierini occur in women with a peak incidence in the second and third decades of life.[1] Methotrexate is a well-established treatment option for plaque morphea. Reports on the successful treatment of IAPP are scant.[2]

CASE REPORT

A 26-year-old woman of European descent with skin type I-II was referred to the dermatologic outpatient clinic for worsening psoriasis started at the age of 5 and had been treated topically since. She further complained about discomfort of both hands with impairment of grip function in addition to pain in the plantar region and lumbar spine. The patient also reported a vague history of an additional symptomless rash on the back being present for at least half a year. Empiric treatment in primary care with a 1-week course of daily ketoconazole 1% shampoo was unsuccessful.

She presented with a Psoriasis Area and Severity Index (PASI) of 7.8, involving psoriatic plaques, mainly on her scalp, and smaller lesions on her trunk and extremities. She had mild psoriatic nail changes. Upon examination, lenticular, depressed, hypopigmented and well-defined patches appeared on the patient's mid-back coalescing into large areas on her upper back and shoulders (Fig 1).

The patient declined an elliptical biopsy but agreed to a 3-mm punch biopsy of the affected skin of the right shoulder that did not show significant histologic changes including hematoxylin-eosin stain, Toluidine blue, Alcian blue, periodic acid—Schiff, and reticulin stain. The clinical findings pointed to a diagnosis of idiopathic atrophoderma of Pierini and Pasini with concomitant psoriasis. Antinuclear antibodies, rheumatoid factor, and anti-CCP antibodies were negative, and C-reactive protein was in the normal range.

After review and examination by the rheumatology department revealing tenderness of 2 proximal interphalangeal joints and bilateral tenderness over the insertion of the plantar fascia, the presence of psoriatic arthritis was confirmed and treatment with methotrexate at a dose of 20 mg/wk was initiated. The patient continued topical treatment with calcipotriol 0.005% plus betamethasone dipropionate 0.05% ointment for psoriatic body areas with 0.1% betamethasone valerate lotion for the scalp psoriasis. No topical treatment was given for the patient's IAPP.

At a 3-month follow-up, the patient's skin psoriasis had almost entirely cleared with just minor improvements in the joint symptoms. Furthermore, major improvement in the patient's atrophic skin changes on her back and shoulders was noted (Fig 2).

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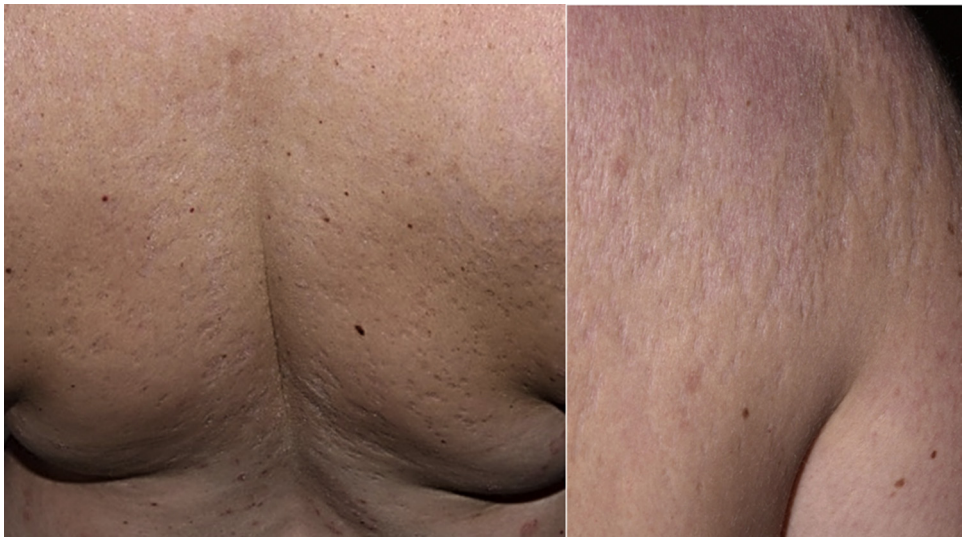

Fig 1. Atrophic patches of variable sizes on the back (left) and right shoulder (right).

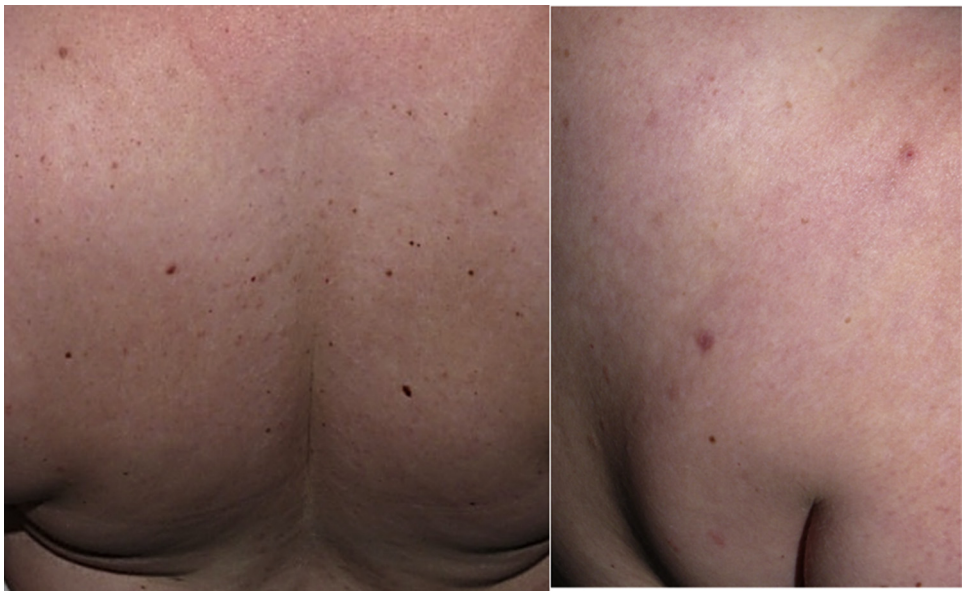

Fig 2. Improvement after 3 months of treatment with methotrexate.

DISCUSSION

As its name indicates, the etiopathogenesis of IAPP is unknown. *Borrelia burgdorferi* has occasionally been found as a potential underlying trigger.[3] Serologic tests or polymerase chain reaction of *Borrelia burgdorferi* in the patient's tissue were not performed, as the patient did not live in and had never travelled to an endemic area of Lyme disease. Whether atrophoderma should be classified as a variant of plaque morphea rather than a distinct entity remains a topic of debate. Arguments for the former mainly consist of findings that both atrophoderma and plaque morphea can occur in the same patient. Arguments for the latter include the absence of the lilac ring in IAPP and findings that IAPP normally persists longer than morphea.[1,4]

The clinical manifestations of IAPP are classically described as footprints in the snow or Swiss cheese–like and consist of soft, round-to-oval, hyper- or hypopigmented, depressed patches with characteristic cliff-drop borders. The sizes of the lesions can vary considerably and range from few millimeters to several centimeters.[5,6] This patient showed classical Swiss cheese–like lesions on middle of her back that seemed to have coalesced into a large lesion on her shoulders.

IAPP seems to be histologically indistinguishable from normal skin in most cases including elastic

stains. Saleh et al[4] found no abnormalities in 12 of 17 specimens stained with hematoxylin-eosin. Possible histologic changes include thickening, minimal homogenization and hyalinization of the collagen bundles, and fragmentation in the elastic fibers. A punch biopsy performed in the lesional skin did not find significant changes. An elliptical biopsy including the involved and normal area perpendicular to the edge of the lesion or 2 simultaneous biopsies from the affected and adjacent nonaffected skin could have been beneficial for finding subtle changes.[4]

Another rarely described differential diagnosis of IAPP is self-involuting atrophoderma of the lateral-upper arm.[7] This diagnosis seemed unlikely in this patient, as her back and shoulders were affected, and IAPP is not self-involuting.

Various forms of morphea associated with psoriasis have been published in case reports.[8] Both conditions are immune mediated, and early lesions of localized morphea and psoriasis are considered helper T cell (Th) 1/Th17 mediated, whereas atrophy is deemed Th2 driven.[9] This case demonstrates IAPP with concomitant psoriasis. The author considers IAPP as a coincidental finding to the patient's long-standing psoriasis.

There is no established treatment for IAPP. Carter et al[10] demonstrated a response to hydroxychloroquine in a patient with presumed lupus-associated IAPP.[10] Antibiotics may be helpful in cases associated with Lyme disease.[3] The assumed relationship with morphea suggests the effectivity of psoralen with ultraviolet A and methotrexate. As the current patient also had moderate-to-severe plaque psoriasis, including nail psoriasis and psoriatic arthritis, methotrexate treatment was initiated. A marked improvement of the atrophic skin changes 3 months after the initiation of methotrexate was revealed. It remains to be seen if methotrexate will become an established treatment for IAPP.

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