Case report

Atrophoderma of Pasini and Pierini simulating extragenital lichen sclerosus et atrophicus: A case report

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Abstract:
We hereby report a 31 year old female who presented with multiple hypopigmented to skin colored atrophic plaques since 7 years. Histopathological examination established a diagnosis of atrophoderma of Pasini and Pierini which is an uncommon form of dermal atrophy.

Key words: atrophy, morphea, Borrelia burgdorferi

Introduction:
The disorder was first described in 1923 by Pasini on trunk of a 21-year-old woman; he referred those lesions as progressive idiopathic atrophoderma [1]. Thirteen years later, Luis Pierini and associates studied 50 Argentinean cases and conclusively defined its clinical and histological features as well as its probable link to morphea [2]. Later in 1958, Canizares et al. introduced the disorder to American literature and also named it after its two pioneers as, “idiopathic atrophoderma of Pasini and Pierini” [3]. Little is known about the exact incidence of this disorder worldwide and it is still debated as to whether atrophoderma represents an atypical, primarily atrophic form of morphea or “burnt-out” morphea or a separate distinct entity.

Case report:
A 33 year old female, working as a tailor, came with multiple brownish dusky colored atrophic lesions present over bilateral upper and lower limbs, since 7 years. It was insidious in onset. These lesions were not associated with any pain, swelling or itching. There was neither any history of trauma nor history of any aggravation on exposure to sun. Patient also failed to recollect any episodes of tick bite prior to appearance of lesions. She also denied applying any topical corticosteroids. Her medical and family history was unremarkable. Her general health was good.

Physical examination revealed multiple well defined, depressed, hypopigmented atrophic plaques with “cliff-drop” border situated over the extensor aspect of extremities. They ranged from few millimeters to several centimeters in size, skin surrounding the lesion appeared normal. There was no erythema or lilac ring. The differential diagnosis that came to our mind were, polymorphous light eruption, extragenital lichen sclerosus et atrophicus. Systemic examination was uneventful. Laboratory work up, including complete blood count, renal and liver function test, thyroid function test, serum electrolyte, urine routine and microscopy were within normal limits. X-ray chest revealed no abnormality. Patient could not afford testing for antibodies against Borrelia burgdorferi.
An incisional skin biopsy taken from an atrophic lesion situated over the right upper arm on the extensor aspect revealed hyperkeratotic as well as atrophic epidermis. Dermis had thick collagen bundles and high lying adnexal structures with atrophy. There was sparse superficial perivascular lymphocytic infiltrate in the dermis. No basal cell vacuolation or papillary edema or dilated blood vessels were seen. Above findings were suggestive of atrophoderma.

We started our patient on tablet doxycycline 100 mg twice a day for 21 days and advised her photoprotection along with daily application of emollient three times a day but found little changes in the lesion. We have currently started her on tablet hydroxychloroquine 200 mg.

**Discussion:**

Atrophoderma of Pasini and Pierini is an asymptomatic, benign condition which is not associated with any significant complications or mortality. Patient may seek medical advice because the lesions can appear cosmetically unacceptable. It usually affects young individuals in second or third decade of life; however, it has been described in individuals as young as 7 years old and as old as 66 years. The disorder is usually sporadic, although a congenital case was recently reported [4].

Etiology of atrophoderma of Pasini and Pierini is poorly understood. Some authors have suggested the possible role of infection with Borreliaburgdorferi [5]. Confusion still prevails regarding its classification, nosology and its relationship to morphea. Although it is worth noting that according to a study by Yokoyama et al in 2000, glycosaminoglycans extracted from idiopathic APP differed from those in typical morphea lesions, probably suggestive of them being different entities [6]. Pathology of morphea appears to be pansclerotic. Morphea lesions usually progress through initial stages of inflammation, where lesions are surrounded by erythema or a violaceous border also known as the lilac ring. These changes are later followed by a sclerotic and eventually an atrophic stage that may be accompanied by hypo or hyperpigmentation.

It is however worth noting that in our case all lesions began with atrophy and ended with atrophy without any intervening stages of inflammation, induration, edema, erythematous or lilac rings. Also, in our case biopsy demonstrated a minimal epidermal atrophy along with hyperkeratosis and thick collagen bundles in the dermis.

Absence of follicular plugging with absent hydropic changes at the dermo-epidermal junction helped us to differentiate this condition from lichen sclerosus et atrophicus.

No effective treatment is available for atrophoderma of Pasini and Pierini. Topical and systemic steroids, D-penicillamine, antibiotics, and phototherapy have been tried with variable efficacy. Carter et al suggested hydroxychloroquine as an option for chronic atrophoderma of Pasini and Pierini refractory to treatment [7]. In one case, Q-switched alexandrite laser (755 nm) was found to be effective in diminishing hyperpigmentation but not atrophy [8].

**References:**

1) Pasini A. Atrophodermaidiopathicaprocessiva. GiorItalDermSif. 1923;58:785.


