Case Report

Atrophoderma of Pasini and Pierini - A case report

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ABSTRACT

Atrophoderma of Pasini and Pierini (APP) is a form of dermal atrophy that manifests as either single or multiple, sharply demarcated, hypo pigmented and non-indurated patches. These patches are marked by a slight depression of the skin, with an abrupt edge (i.e., the "cliff-drop" borders), mainly located on the backs of adolescents or young adults. Here by we report a case of 22 year-old female, who presented with asymptomatic, hypo pigmented, depressed patches over body especially both arms, forearms, thighs and legs since five years. Laboratory evaluations were normal. Biopsy showed changes suggestive of Atrophoderma of Pasini and Pierini.

KEYWORDS: *Atrophoderma of Pasini and Pierini, "cliff-drop" borders.*

INTRODUCTION

In 1923, Pasini[1] described a case of pigmentary atrophoderma that was both clinically and histologically unique from any other known atrophy, including localized scleroderma, under the name of progressive idiopathic Atrophoderma. In 1936, in Argentina, Pierini and Vivoli[2] extensively studied and defined the condition and its possible link to morphea. In 1958, the disorder was first introduced into the American dermatologic literature by Canizares et al,[3] who reviewed Pierini's findings and renamed the disorder as idiopathic Atrophoderma of Pasini and Pierini. They believed that the idiopathic Atrophoderma of Pasini and Pierini (APP) differed sufficiently from morphea to classify it as a distinct entity.

APP is a benign, asymptomatic disease and is not associated with any significant complications or mortality. It is insidious in onset, twice as common in females as in males. The most common age group affected is 20-30 years. However, it has been described in individuals as young as 7 years old and as old as 66 years, with one report of congenital atrophoderma. The clinical appearance of Atrophoderma of Pasini and Pierini has been likened to "footprints in snow" or depressions with "cliff drop" border. [2] The trunk is the most commonly involved site, especially the back and the abdomen.[3] No effective treatment is available for APP. Here by we report a case of APP in 22yr old female mainly involving extremities which are unusual sites of involvement and its partial response to doxycycline.

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CASE REPORT

A 22year-old female presented with asymptomatic lesions all over body since five years. No history of trauma or infections present at that site. Cutaneous examination revealed numerous oval to round hypo pigmented atrophic macules of 1-4 mm in size without perilesional erythema over both extensor part of thighs and legs (Fig. 1a) and flexor aspect of arms and forearms (Fig. 1b). Lesions were also present over upper part of chest and neck. Their distinct margins and depressions gave them typical "cliff-drop" appearance with no induration, tenderness or sclerosis. No nail or hair changes were present. Laboratory investigations, including Complete blood count, blood sugar, urinalysis, liver function test, renal function tests, ESR, Serum electrolyte were within normal limits. Chest radiograph was normal. Skin biopsy was taken from one of the atrophic lesions over left arm keeping localised morphea and APP as differentials. The section

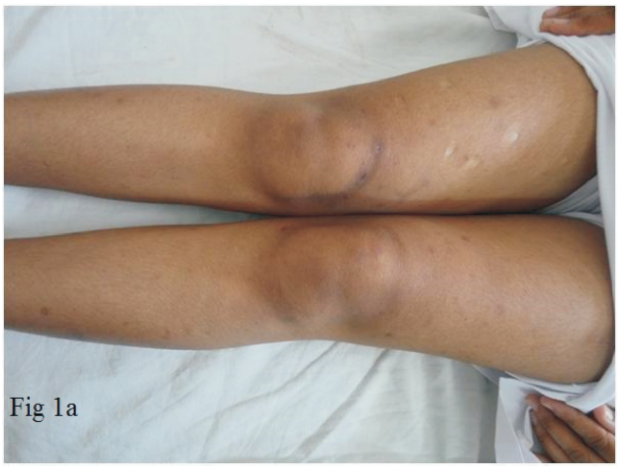

Fig1a. Multiple hypopigmented atrophic macules over both extensors of thighs and legs without perilesional erythema

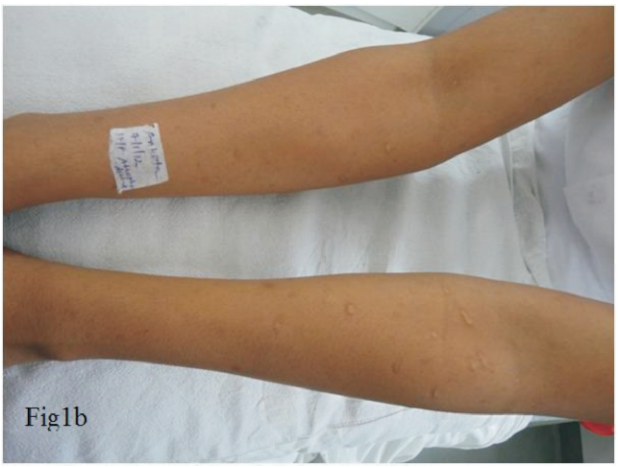

Fig1b. Multiple discrete hypopigmented atrophic macules over both flexor aspect of arms and forearms having typical "cliff-drop" appearance

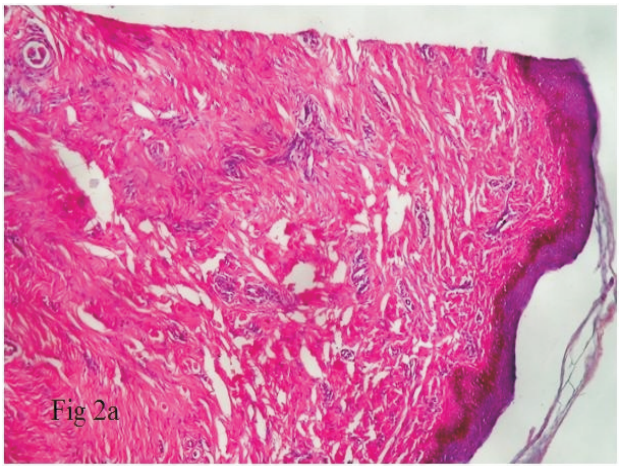

Fig 2a. Atrophy of epidermis with marked thickening of the collagen bundles in dermis H&E stain 10x

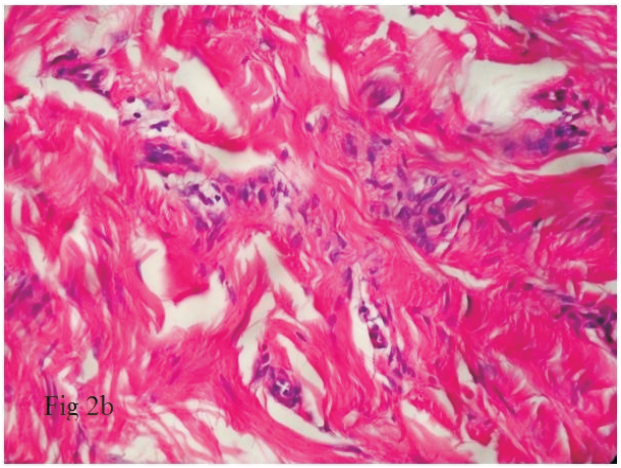

Fig 2b. Atrophy of epidermis with marked thickening of the collagen bundles in dermis H&E Stain 40x

showed atrophy of epidermis with marked thickening of the collagen bundles with few tightly packed bundles. Mild perivascular inflammatory infiltrate compromised by lymphocytes and plasma cells with few congested blood vessels present in dermis (Fig 2a & Fig 2b). Features were suggestive of atrophoderma of Pasini and Pierini.

She was given oral doxycycline 200 mg per day for three weeks, and the depressed depths of the lesions improved. She took oral doxycycline 200 mg per day for an additional three weeks, but there was no further improvement.

DISCUSSION

Atrophoderma of Pasini and Pierini is a form of dermal atrophy that manifests as either single or multiple, sharply demarcated, hypopigmented and non-indurated patches. These patches are marked by a slight depression of the skin with an abrupt edge (i.e., the "cliff-drop" borders), usually located on the backs of adolescents or young adults. The lesions may be discrete or confluent, and the affected skin appears thinned and discoloured.[4] Distribution is often symmetrical and bilateral; however, reports have described solely unilateral cases also.[5,6] The exact cause of Atrophoderma of Pasini and Pierini is not known yet. Genetic factors, neurogenic factors, abnormal metabolism of dermatan sulphate and immunological factors have all been implicated in the pathogenesis of APP.[7] In 2000, Yokoyama *et al*.[8] reported that skin glycosaminoglycans extracted from idiopathic APP lesions are different from those in typical morphea lesions. The pathophysiologic events that cause the discrete lesions seen clinically, as well as the timing of their appearance, are also unknown.

The histopathologic changes, often minimal and non-diagnostic, consist of a decrease in the size of the dermal papillae, with flattening of the rete pegs. The epidermis is usually normal or slightly atrophic. Melanin is increased in the basal layer, interstitial oedema and a mild perivascular infiltrate consisting of lymphocytes and histiocytes may be present. The collagen bundles show varying degrees of homogenization and clumping in the mid and reticular dermis, with a normal papillary dermis. When compared with adjacent normal skin, the dermal thickness is reduced. The sweat glands and the pilosebaceous units are not affected.[4]

Sclerosis and induration are seen in some lesions of APP. Lesions of morphea resemble APP with homogenization of collagen and a perivascular lymphocytic infiltrate. However, there are certain features which suggest that APP and morphea are different entities. Morphea characteristically begins as a discrete circumscribed, erythematous-to-sclerotic plaque, often with a white centre and characteristic peripheral lilac rim. It involves all layers of epidermis, while APP affects only collagen in dermis. APP lack sclerosis and lesions commonly coalesce over time, producing a moth-eaten appearance that is not consistent with morphea. [9] APP should always be differentiated from morphea so as to avoid unnecessary treatment to the patient. Topical and systemic steroids, antimalarials, D-penicillamine, antibiotics, phototherapy and Q-switched alexandrite laser (755 nm) are found to be effective with variable results. A case of Atrophoderma of Pasini and Pierini associated

with *Borrelia burgdorferi* infection was successfully treated with oral doxycycline by Yoonhee Lee et al.[10] A retrospective evaluation of 25 patients treated with either oral penicillin (2 million IU per day) or oral tetracycline (500 mg three times per day) for 2-3 weeks showed clinical improvement, with no new active lesions in 20 patients (80%).

We present the case to highlight the uncommon site of involvement over extremity, and its partial response to doxycycline.

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Source of Support: Nil Conflict of Interest: Nil