Atrophoderma of Pasini and Pierini

Sir,

A 38-year-old man was under treatment for cutaneous leishmaniasis (single lesion) in our department. The skin examination revealed multiple asymptomatic, hyperpigmented patches of various sizes (2 to 6 cm) over back and thighs, which according to the patient were present since childhood and he had not taken any treatment for them. These patches were depressed below the level of skin surface with well-defined border giving an impression of "cliff drop" appearance [Figure 1]. There was no induration, tenderness but slight atrophy was perceived. The lesions were nonprogressive. There was no history of any trauma, infection, or ulceration at the sites. Laboratory examination including total and differential lymphocytic count, erythrocyte sedimentation rate, renal function tests, liver function test, and antinuclear antibody test were within normal limits. Chest X-ray was normal. Histopathological examination revealed normal epidermis, hyalinization of collagen in dermis with minimal scattered inflammatory infiltrate. Dermis was slightly reduced in thickness [Figures 2 and 3] as compared with normal adjacent skin. A diagnosis of atrophoderma of Pasini and Pierini (APP) was made on the basis of clinical and histopathological examination.

In 1923, Pasini[1] described the entity under the name 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, Canizares *et al*.[3] reviewed Pierini's findings and renamed it idiopathic APP. Canizares *et al*. believed that idiopathic APP differed sufficiently from morphea to classify it as a distinct entity. In 2000, Yokoyama *et al*.[4] reported that skin glycosaminoglycans extracted from idiopathic APP lesions are different from those in typical morphea lesions. The cause of APP is not known. The pathophysiologic events that cause the discrete lesions seen clinically, as well as the timing of their appearance, are also unknown. Some authors have suggested a role for infection with *Borrelia burgdorferi.* APP is a benign, asymptomatic disease and is not associated with any significant complications or mortality. It usually begins insidiously in individuals during the 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, with one report of congenital atrophoderma.[5]

The precise classification and exact nosology of APP have always been a matter of debate. Some authors believe it to be a variant of localized scleroderma or morphea on the basis of certain clinical or histopathological grounds. These are sclerosis and induration in some lesions of APP. Morphea and APP were observed in the same patient; in its late atrophic stage, lesions of morphea resemble APP and homogenization of collagen and a perivascular lymphocytic infiltrate are common histological features seen in morphea and APP.[6] 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 center and characteristic peripheral lilac rim. APP lack sclerosis, and lesions commonly coalesce over time, producing a moth-eaten appearance that is not consistent with morphea.[7]

APP is a form of dermal atrophy that manifests as single or

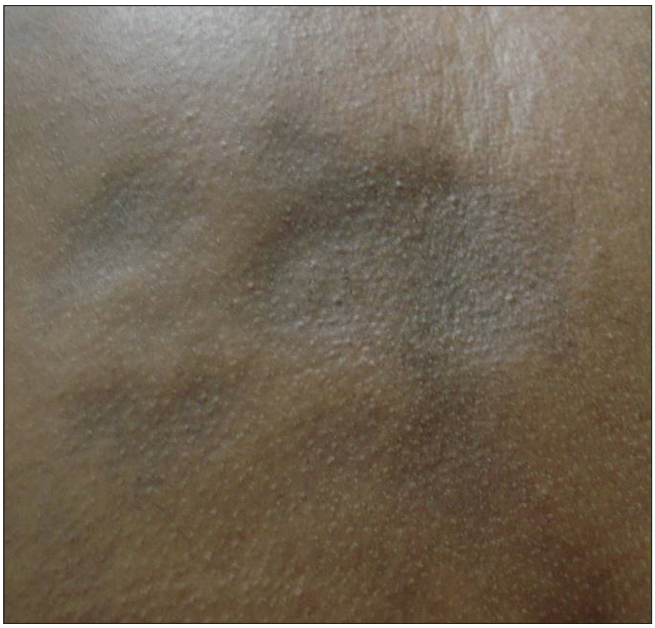

Figure 1: Multiple well-defined depressed plaque over back

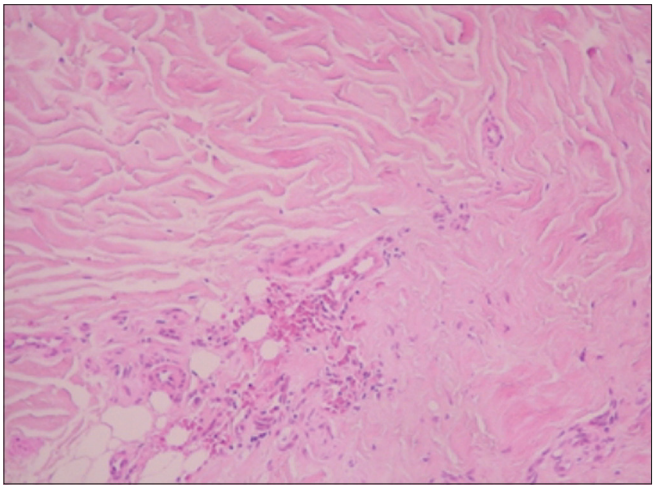

Figure 3: Hyalinization of collagen with minimal inflammatory infiltrate in dermis (H and E, ×100)

multiple sharply demarcated, round to oval, hyperpigmented, nonindurated patches of varying sizes ranging from few millimeters to several centimeters. The lesions may be discrete or confluent and are usually asymptomatic and do not show any signs of inflammation. These patches are marked by a slight depression of the skin with an abrupt edge often exhibiting a "cliff-drop border," usually on the backs of adolescents or young adults. Another description commonly used is "footprints in the snow," characterizing the common oval shape of the depressed lesions. Histopathologic changes are often minimal and are notdiagnostic. The epidermis is usually normal or slightly atrophic. Interstitial edema and a mild perivascular infiltrate consisting of lymphocytes and histiocytes may be present. Collagen bundles show varying degrees of homogenization and clumping. When compared with adjacent normal skin, dermal

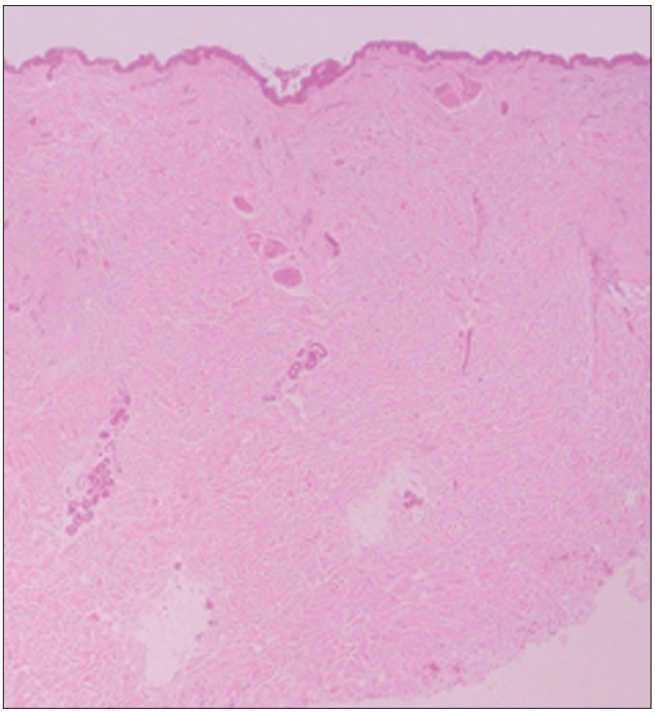

Figure 2: Normal epidermis, dense collagen bundles in dermis, and minimal inflammatory infiltrate (H and E, ×10)

thickness is reduced. The sweat glands and the pilosebaceous units are not affected. The appendages are preserved. Elastic fibers appear normal after elastic tissue staining and with electron microscopic studies.[6] No effective treatment is available for APP. Topical and systemic steroids, antimalarials, D-penicillamine, antibiotics, and phototherapy have been used with variable efficacy. Q-switched alexandrite laser (755 nm) was found to be effective in diminishing the hyperpigmentation by 50% after three treatments in one case.[8]

APP should always we differentiated from morphea so as to avoid unnecessary treatment to the patient.

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Access this article online

Quick Response Code:

Website: www.idoj.in

DOI: 10.4103/2229-5178.86012

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