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IDIOPATHIC ATROPHODERMA OF PASINI AND PIERINI: A CLINICO- PATHOLOGICAL STUDY.

KEYWORDS: atrophoderma of Pasini–Pierini, dermal atrophy, morphea

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ABSTRACT Idiopathic atrophoderma of Pasini–Pierini (IAPP) is an uncommon dermatologic condition of unknown etiology, characterized by asymptomatic,single or multiple sharply demarcated, round to oval, hyperpigmented, nonindurated depressed patches of varying sizes. The lesions may be discrete or confluent and do not show any signs of inflammation. Etiology of atrophoderma of Pasini–Pierini is poorly understood. The diagnosis is typically made by clinical findings; however, biopsy is commonly used to exclude other entities. In this study, we describe the clinical findings of three patients with IAPP. The histopathologic features of two patients were studied. This study also discusses whether the IAPP is an individual entity or associated with morphea.

Introduction

Idiopathic progressive atrophoderma as described by Pasini[1] in 1923 and later by Pierini and Vivoli[2] in 1936 is characterized by single or multiple, sharply but often irregularly demarcated, gray or brown areas that are slightly depressed below the surrounding tissue. The lesions are usually asymptomatic and do not show inflammation. They conclusively defined its clinical and histological features as well as its probable link to morphea. In 1958, Canizares et al[3] proposed the term idiopathic atrophoderma of Pasini and Pierini. He believed that IAPP differed sufficiently from morphea to classify it as a distinct entity. 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 a separate distinct entity. The histological changes are slight and variable and the diagnosis is usually made on the basis of clinical features. We report three cases of this rare dermatoses and reviewed their clinical and histological characteristics and its nosological position.

CASE 1:

A 21-year-old female presented with a 7-year history of multiple asymptomatic depressed areas of skin on her arms, back, abdomen and thighs. It was insidious in onset. She was unable to recall any other preceding event such as a bite or local injection over those sites or a history of panniculitis, fever, systemic symptoms or diabetes prior to the onset of eruption. Her medical and familial histories were unremarkable. The patient's general health was excellent. For almost one year, there has been little change in her disease. Laboratory findings, including a complete blood count, liver function test, urinalysis, electrolytes and antinuclear antibody test were within normal limits. Cutaneous examination revealed multiple well defined depressed hyperpigmented plaques with "cliff-drop" border situated on her back, abdomen, upper limbs and both thighs. They ranged from few millimeters to several centimeters in size, skin surrounding the lesion appeared normal (Figure 1&2). There was no erythema or lilac ring. These lesions looked like "footprints in the snow" or "Swiss cheese." No induration, tenderness or sclerosis was present. Skin biopsy was taken from one of the atrophic pigmented lesion over upper back keeping morphea and atrophoderma of Pasini and Pierini as differentials. The section revealed an epidermis having normal thickness, but flattening of rete pegs and slight increase of melanin pigment in the basal layer. Dermis showed reduced thickness and homogenous, hyalinised collagen bundles with minimal scattered inflammatory infiltrate and unaffected skin adnexal structures (Figure3).

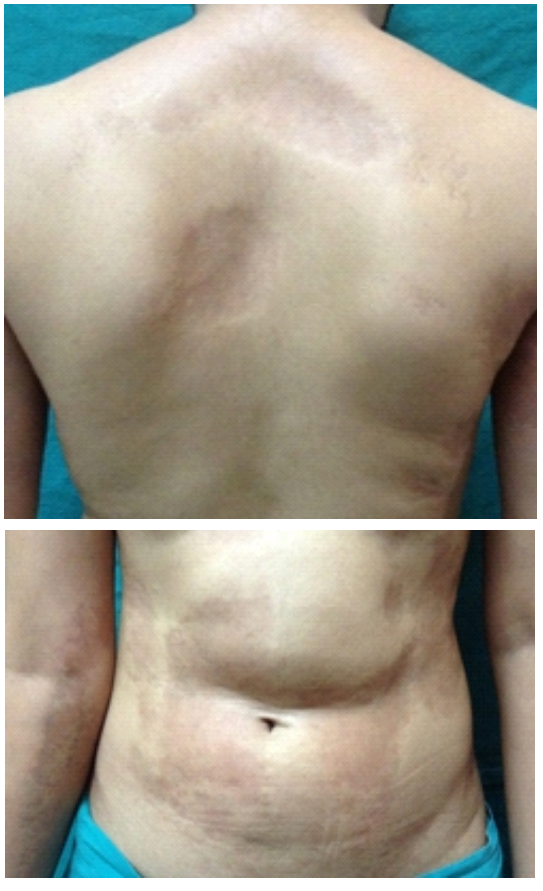

Figure1&2: variably sized depressed patches over back, abdomen and upper limbs

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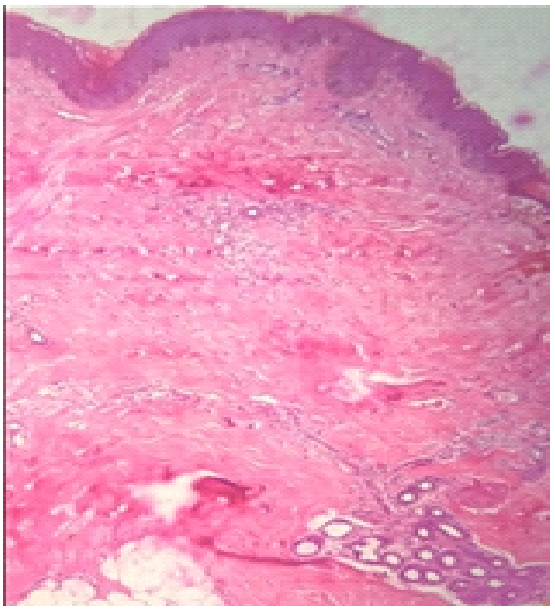

Figure 3: H&E(10x)Epidermis shows flattening of rete pegs, increased melanin in the basal cell layer, dermal atrophy with thickening and focal homogenization of collagen; with normal skin appendages

Subcutaneous fat was unremarkable. Verhoeff-van Gieson stain for elastic tissue showed clumping and fragmentation of elastic fibres. The final diagnosis of atrophoderma of Pasini-Pierini was made on the basis of clinical and histopathological examination and the patient was comforted by the fact that this disease usually follows a protracted course and will eventually stop progressing.

CASE 2:

A 61-year-old woman presented to our department with a 5-year history of asymptomatic hyperpigmented and depressed patch on her lower back. The lesions were non progressive. She had no history of trauma, infection and injection. There were no burning, itching, pain and any other symptom. Laboratory examination including total and differential leucocyte count, erythrocyte sedimentation rate, renal function tests, liver function test, and antinuclear antibody test were within normal limits. Systemic examination was uneventful. On examination, a single, slightly depressed and hyperpigmented patch with normal texture, approximately of size 17 × 8 cm, was present on the lower back and lumbosacral region. The patch was irregularly shaped with ill-defined borders. Close examination of the skin demonstrated the unique cliff-drop border, giving the impression of an inverted plateau (Figure4). There was no induration

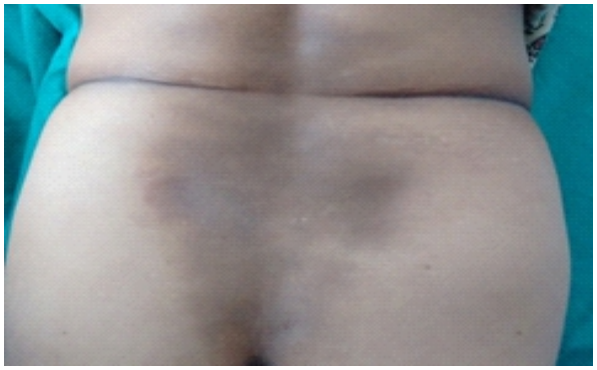

Figure 4: A single slightly depressed hyperpigmented patch with normal texture was present over lower back involving lumbosacral region

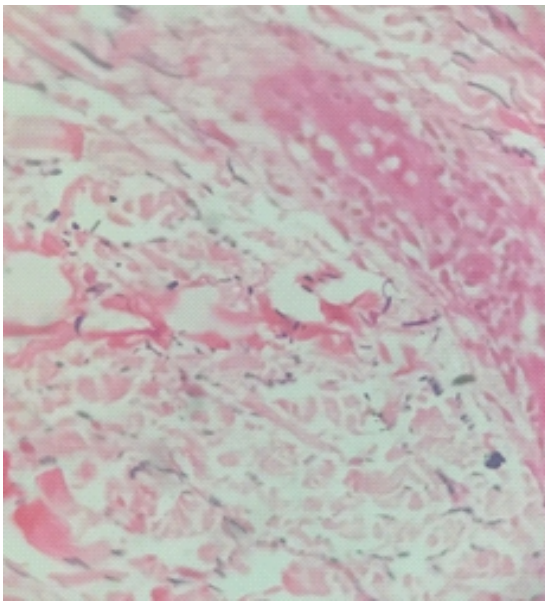

Figure 5: Verhoeff-van Gieson section (40x) Rarefaction and fragmentation of the elastic fibres

or tenderness. Histopathological examination revealed normal epidermis, perivascular and periappendigeal lymphohistiocytic aggregate in the upper dermis and thickening of the collagen fibres with marked hyalinization of collagen in reticular dermis. Melanin was increased in the basal layer with pigment incontinence. The elastic tissue stain showed rarification and fragmentation of elastic fibers (Figure5). Both the clinical and histopathological features were consistent with atrophoderma of Pasini and Pierini.

CASE 3:

A 30- year- old presented with a 6-year history of asymptomatic, depressed brownish lesion on his back. The lesion was not associated with any pain, swelling or itching. He had no history of trauma, infection or injection. His medical and family history was unremarkable. His general health was also good.

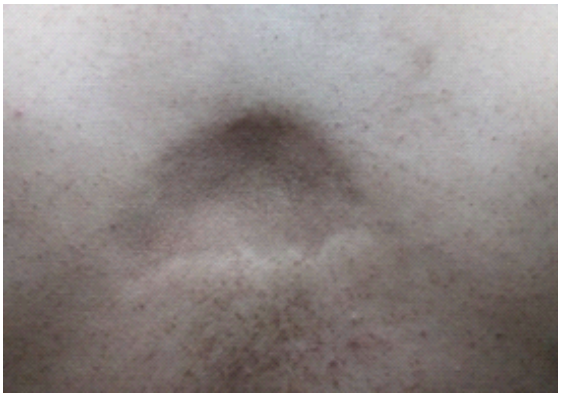

Figure 6: Well defined depressed patch over interscapular region with characteristic "cliff-drop" border

Laboratory investigations revealed normal full blood count, erythrocyte sedimentation rate, renal function tests and liver function tests. Physical examination revealed a single depressed hyperpigmented patch of size 8x4cm in the interscapular area. The lesion had a well demarcated, depressed, non-inflamed border just like a "cliff-drop" (Figure6). There was no induration, sclerosis or inflammation. Although our request for biopsy was refused, the diagnosis of atrophoderma of Pasini and Pierini was made in consideration of the clinical features.

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DISCUSSION

Idiopathic atrophoderma of Pasini and Pierini is a benign, asymptomatic disease and is not associated with any significant complications or mortality. Patient may seek medical advice because the lesions may appear cosmetically unacceptable. It usually begins insidiously in 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 with few reports of congenital atrophoderma[4,5]. It manifests as single or multiple sharply demarcated, hyperpigmented, 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 trunk, particularly on the back and abdomen and to a lesser extent on the extremities. The lesions may be discrete or confluent, and the affected skin appears discolored[6]. Distribution is often symmetric and bilateral but unilateral cases have also been reported[7,8]. The consistency and feel of the affected skin remains normal and there is no edema, thickening, sclerosis or leathery feeling. When several depressed patches are present, they would give a "Swiss-cheese like" or "footprints in snow" appearance. There is no induration, scaling, "lilac ring" or other changes on the skin surrounding the lesions[9]. IAPP usually follows a benign and protracted course over 10-20 years. Although several therapeutic modalities have been tried including antibiotics (penicillin and tetracycline) and antimalarials, no treatment is consistently effective and some investigators suggest that no aggressive therapy is required[6,10,11].

The lesions in case 1 were involved in extensive regions, including the back, abdomen, shoulders, and upper and lower extremeties. The characteristic "cliff-drop" borders, "Swiss-cheese appearance", absence of induration or sclerosis and lack of "lilac ring" appearance leads to a diagnosis of IAPP. Case 2 and 3 had characteristic hyperpigmented, depressed patches with a unique cliff-drop border, giving the impression of an inverted plateau present over lower back with involvement of lumbosacral region (case 2) and upper back (case 3) respectively. There was no induration, sclerosis or inflammation. Buechner and Rufli[6] had reported upper back and lumbosacral region as the commonest site for localized variety in their study of thirty-four patients.

The histopathologic changes, usually subtle 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, and interstitial edema 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. The dermal thickness is reduced and the sweat glands, pilosebaceous units and appendages are not affected[6,12]. Most of the earlier studies have shown no abnormalities in elastic fibers with either elastic tissue staining [6] or electron microscopy[13]. However, Saleh et al[14] described a spectrum of histopathological findings ranging from normal to severe diminution and fragmentation of elastic fibers, with 35.3% of cases showing moderate-to-severe reduction and fragmentation. In our patients histopathology revealed normal epidermis. Melanin was increased in the basal layer. Perivascular and scattered lymphohistiocytic infiltrate was present in the upper dermis. Collagen bundles showed varying degrees of homogenization and clumping in the mid and reticular dermis. Histological features of morphea such as dermal sclerosis, perivascular and interstitial lymphoplasmacytic infiltrate and eccrine gland entrapment were absent in all the biopsy specimens. The Verhoeff-van Gieson for elastic tissue showed rarification and fragmentation of elastic fibers in both the specimens. Our light microscopic findings were consistent with the diagnosis of atrophoderma of Pasini and Pierini.

The cause and etiopathogenesis of IAPP remains unknown. The pathophysiologic events that cause the discrete lesions seen clinically, as well as the timing of their appearance, are also unknown. IAPP is classically thought of as an idiopathic atrophy of the dermis. Franck et al[15] had observed that the skin depression in IAPP lesions is solely because of dermal atrophy. Genetic factors, neurogenic factors, immunological factors, and abnormal metabolism of dermatan sulfate have all been implicated in the pathogenesis of IAPP5,[12,16,17]. The role of Borrelia burgdorferi remains controversial[6].

The precise classification, nosology and its relationship to morphea have always been a matter of debate despite differences in the origin, development, and outcome of the lesions. Canizares et al[3] concluded that IAPP lesions are different from true morphea and distinctive enough to justify their classification as a separate entity. Whether IAPP is a distinct entity or a variant of localized scleroderma has been disputed since its original description. There have been several attempts at characterizing the clinical and histopathological features of IAPP and explicating its etiology. Some authors think that IAPP is a variant of morphea due to likelihood of developing systemic sclerosis in some patients. Co-occurence of IAPP and morphea has been reported. IAPP is considered by many to be an abortive morphea in which indurations failed to develop. However, several features distinguish IAPP from morphea. IAPP presents a decade earlier than morphea and usually has a longer course. Second, IAPP lesions have a peculiar cliff-drop border that is not seen in morphea. Third, inflammation and loss of appendegeal structures seen in morphea is not a feature of IAPP. Fourth, sclerosis which is prominent in morphea, is minimal or absent in IAPP. Fifth, elastic tissue may show clumping or fragmentation in IAPP but are always normal in morphea[6,14,18,19]. Yokoyama et al[20] reported that glycosaminoglycans extracted from IAPP lesions differed from those in typical morphea lesions, probably suggestive of them being different entities.

In our series, in all patients lesions began with atrophy without any intervening stages of inflammation, induration, or a peripheral lilac ring. None of the patients had personal or family history or evidence on physical examination of systemic diseases particularly scleroderma. None of the patients reported preceding redness, induration or a history of insect bite. Of note, testing for antibodies against *Borrelia burgdorferi* was not taken because of the low prevalence of Lyme disease in our area. Moreover, the histological features including changes of elastic fibers on Verhoeff-van Gieson stain support the concept that IAPP is a distinct entity. Also the differentiation of IAPP from morphea is of practical importance because of different management and prognosis.

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