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Case Report

Atrophoderma of Pasini & Pierini- is it an autoimmune disease?

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Abstract Atrophoderma of Pasini and Pierini is distinct from morphoea in its clinical appearance, course and the treatment options. There is no known etiology and effective treatment of the disease. The presence of antinuclear antibodies in this disease is a clue to its autoimmune basis. This discovery will open new avenues in further research of the disease.

Key words
Atrophoderma of Pasini and Pierini, antinuclear antibodies.

Introduction

The status of atrophoderma of Pasini & Pierini as a distinct entity is not established. Most clinicians still consider it as an atrophic variant of morphoea.[1] The clinical picture and the course of the disease are however, distinct from morphoea but the histopathological appearance is the same.

Anti-nuclear antibodies (ANA) are the autoantibodies directed against cellular protein or nucleic acid. Their presence is a proof of autoimmune basis of the disease. Ours was the first documented case of atrophoderma of Pasini and Pierini in Pakistan. It was significant in the way that ANA was positive in this patient.

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Case report

A 25-years-old male reported in the skin OPD of Combined Military Hospital, Peshawar with complaints of multiple pigmented spots over the chest, abdomen, back and both arms since the last 4 years. There was no itching or pain in the lesions. The lesions were persistent and unchanged since the start of the disease. Past history was not contributory. There was no history of intake of any drug. He was second of four brothers and sisters. There was no family history of any skin or systemic disease.

On examination, the patient was a young man with average build. He was well oriented in time and space. His vital signs were stable. Systemic examination was unremarkable. Dermatological examination showed multiple, round or oval, pigmented patches with vague borders over the chest, abdomen, buttocks and upper arms (**Figure 1**). The patches were slightly depressed and

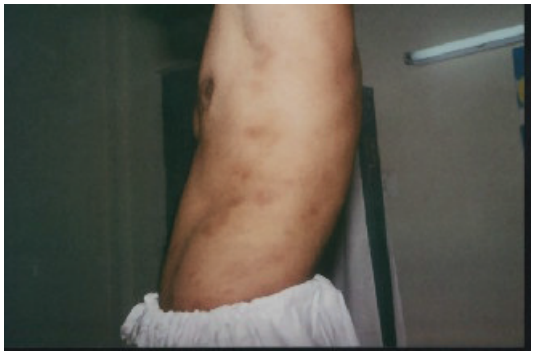

Figure 1 Multiple, round or oval, pigmented patches with vague borders are seen over the chest and abdomen.

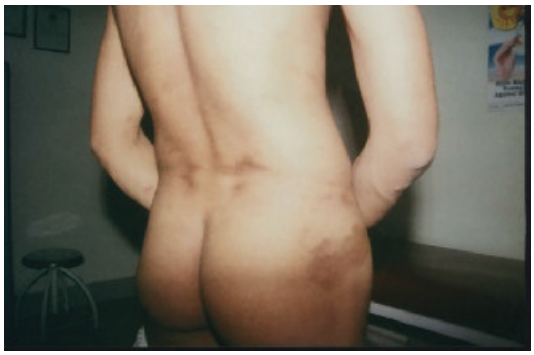

Figure 2 The patches were slightly depressed and atrophic.

atrophic (**Figure 2**). There was no induration in the lesions and the surrounding skin was completely normal. The blood and urine routine examination did not reveal any abnormality. The anti-nuclear antibody test was positive. The histopathological examination of the skin biopsy showed reduction in dermal thickness, and the collagen bundles appeared homogenous and clumped together in reticular dermis. The epidermis was completely normal.

Discussion

Atrophoderma of Pasini and Pierini is characterized by multiple round or oval, slate-coloured or violet-brown, atrophic patches over the trunk and proximal extremities. The patches are different from morphoea in that there is no induration or oedema and the lilac-coloured border is also absent. Its differentiation from morphoea is of practical importance because the course and treatment of the two diseases are very different from each other.

There is no known cause of atrophoderma of Pasini & Pierini. No genetic factor has been reliably incriminated, although familial cases have been reported.[2] Recently anti-nuclear antibodies have been detected in a few patients suggesting immunological pathogenesis.[3] Another study found *Borrelia burgdorferi* antibodies in a few patients of atrophoderma of Pasini and Pierini, suggesting an infective origin.[4] It is proposed that atrophoderma of Pasini and Pierini is primary atrophic abortive morphoea, in which induration fails to develop.[5]

Histopathological appearance is that of increased pigmentation of the basal layer of the epidermis. In early stages, collagen bundles in the lower dermis are edematous. Later on they become clumped and homogenous, hence, reducing the total thickness of the dermis. Elastic tissues are clumped and scanty. Later on there may also be some epidermal atrophy.[6] Electron microscopy demonstrates macrophages and lymphocytes around blood vessels and between the collagen fibers in the dermis. Monoclonal antibody studies of these cells demonstrate cells reacting with anti-Leu-1, anti-Leu-3a and OKM-1 antibodies. Direct immunofluorescence examination showed IgM and C3 staining in small dermal blood vessels.[7]

The course of the disease is prolonged and the lesions persist unchanged. There is no effective treatment, but psoralen and UVA (PUVA) has helped some patients.[8]

Conclusion

It may be concluded that recognition of atrophoderma of Pasini & Pierini, as a distinct entity from morphoea is important as the patient may be briefed about its prolonged course and unavailability of any treatment options. The detection of anti-nuclear antibodies in the disease has opened new frontiers in further research on the disease.

References

1. Kee CE, Brothers WS, New W. Idiopathic atrophoderma of Pasini and Pierini with co-existing morphoea. A case report. *Arch Dermatol* 1960; **82**: 154-7.
2. Barsky S, Ke M. Congenital atrophoderma of newborn. *Arch Dermatol* 1970; **101**: 374-5.
3. Wokalek H, Schmidt HG, Niedner R. Idiopathic progressive atrophoderma of Pasini and Pierini with the presence of antinuclear antibodies. *Hautarzt* 1985; **36**: 154-60.
4. Wojas-Peic A, Wielowieyska-Szybinska D, Kieltyka A. Presence of antinuclear antibodies and antibodies to Borrelia burgdorferi among patients with morphoea en plaque, deep linear scleroderma and atrophoderma of Pasini and Pierini. *Przegl Lek* 2002; **59**: 898-902.
5. Kencha D, Blaszczyk M, Jablonska S. Atrophoderma Pasini-Pierini is a primary atrophic abortive morphoea. *Dermatology* 1995; **190**: 203-6.
6. Beuchner SA, Rufli T. Atrophoderma of Pasini and Pierini. *J Am Acad Dermatol* 1994; **30**: 441-6.
7. Berman A, Berman GD, Winkelmann RK. Atrophoderma (Pasini-Pierini). Findings on direct immunofluorescence, monoclonal antibody and ultra structural studies. *Int J Dermatol* 1988; **27**: 487-90.
8. Burton JL, Lovell CR. Disorders of connective tissue. In: Champion RN, Burton JL, Burns DA, Breathnach SM eds. *Textbook of Dermatology, 6th edn.* Oxford: Blackwell Science; 1998. p. 2003-71.