CASE REPORT

Idiopathic Atrophoderma of Pasini – Pierini (IAPP): Case report with the review of literature

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ABSTRACT

We describe a 24-year-old Qatari male patient with 2-years history of multiple asymptomatic atrophic sharpy demarcated brown patches affecting shoulders, upper arm, trunk, back and lumbar region consistent with the diagnosis of Idiopathic Atrophoderma of Pasini – Pierini (IAPP) and review the literature.

INTRODUCTION

Atrophoderma of Pasini and Pierini is a rare dermatologic condition that was first described by Pasini and Pierini[1,2] and was introduced to the American Dermatological literature by Canizares who also proposed the term IAPP.[3]

The disorder usually begins during adolescence with slightly erythematous lesions commonly affecting the back or lumbosacral region of the trunk.

The lesions frequently spread to affect chest, arms and abdomen.[4] The disease may begin with single lesion but more often with multiple lesions varying in size from 1–12 cm in diameter that usually appear within 1–2 weeks. These lesions show atrophy and are slate grey or brown in color. The lesions spread slowly over months or years before becoming stable. Discrete new lesions may appear over 10–20 years and old lesions may slowly enlarge giving the skin a moth eaten's appearance. The lesions are usually round or ovoid varying in size from few millimeters to several centimeters and are often along the cleavage lines and coalesce to form large irregular shaped plaques with convex border.[5]

CASE REPORT

24-year-old Qatari male patient was seen in dermatology clinic on 20th April, 2014. He has 2-years history of multiple asymptomatic atrophic sharpy demarcated brown patches affecting shoulders, upper arm, trunk, back and lumbar region (Fig. 1, 2, 3, 4).

The lesions varied in size but the largest area affected was upper back and lumbar region.

There was no history of trauma or insect bites.

His personal and family histories were negative for systemic disorders.

The patient general assessment was normal (normal temperature, pulse, blood pressure, weight 63, height 168 centimeter).

Laboratory findings included CBC, ESR, Blood

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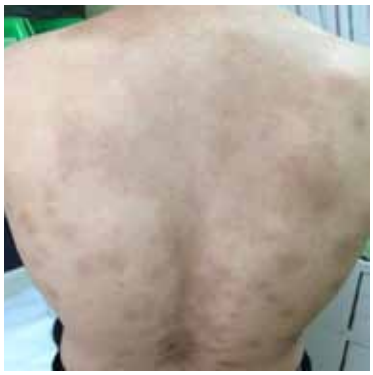

Fig. 1 Affection of back.

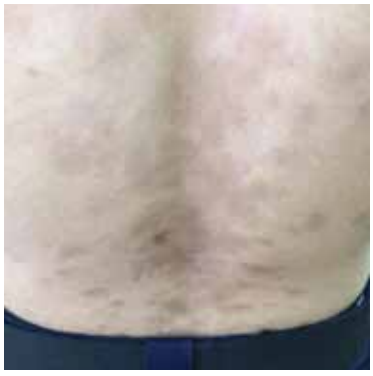

Fig. 2 Affection of lower back.

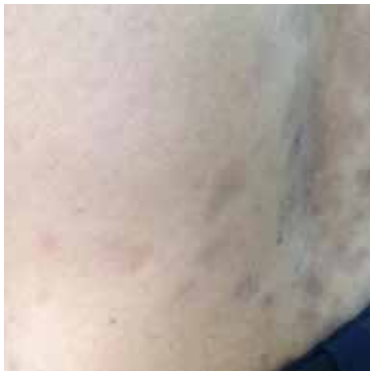

Fig. 3 Affection of lower back and loin.

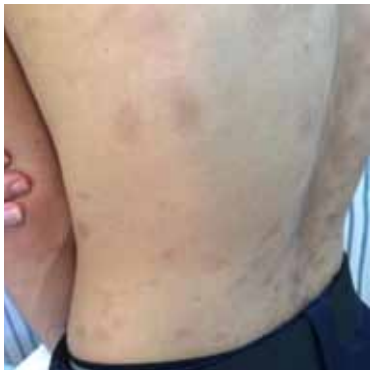

Fig. 4 Affection of side of chest and upper arm.

chemistry, immunology, HIV, Hepatitis screening, Rheumatoid factor, ANA, Anti Ds DNA, Ant: Scl 70 antibodies and serum antibodies for Borrelia Burgodoferi were negative. Results of all previous investigations were normal.

Skin biopsy taken from lesions showed local epidermal atrophy with decrease in the size of the dermal papillae, homogenous collagen stroma with capillaries and pilosebaceous unit that are morphologically normal (Fig. 5, 6).

DISCUSSION

IAPP is a benign condition characterized by solitary or multiple slowly progressive patches that remain stable for years. The lesions have symmetrical distribution and mainly affect the trunk especially the back and to a lesser degree on extremities. Few lesions may be unilateral or zosteriform.[5,6,7] The affected skin shows

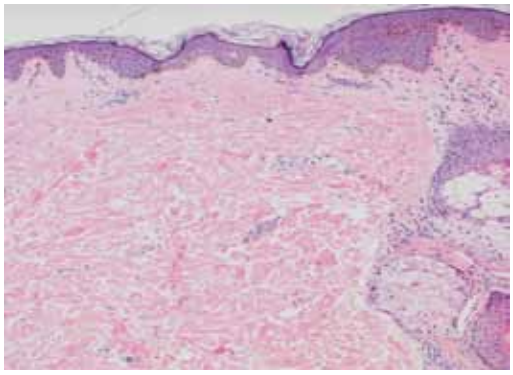

Fig. 5 Histopathology with a degree of epidermal atrophy and decreased dermal papillae and homogenous collagen.

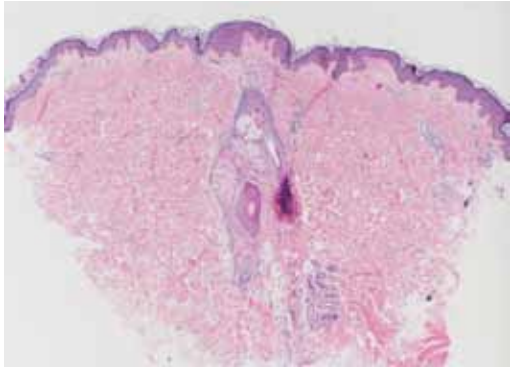

Fig. 6 Normal pilosebaceous unit.

asymptomatic multiple round or oval depressed lesions 2–3 cm in diameter. The lesions can be bluish-violet, slate grey or brown in color with irregular sharply demarcated borders.[8] Usually the lesions start small and can grow and coalesce to form large areas. Extremities were reported to be predominantly affected most commonly with hypopigmented patches.[9]

The IAPP is mainly due to atrophy and significant decrease in thickness of dermis and subcutes.[10,11]

The role of Borrelia burgdorferi in APP remains controversial[12] and some reported positive serology.[13,14]

Some consider atrophic morphoea and superficial morphea synonymus to APP[15,16] and APP could be an abortive morphoea.[17,18] They can be differentiated clinically as morphea has a later age of onset and have a shorter course than APP as it clears with 3–5 years while APP may have a course for 10–20 years and morphea appear more inflamed with induration and erythema.[8] While atrophic lesions of AAP are brown with well demarcated border and no signs of inflammation.

Study of the relation between Borrelia burgdorferin and APP revealed that 28.5% to 53% of APP have positive serology for Borrelia burgodorferi compared with control subjects.[12,17] Localized scleroderma associated with Borrelia burgdorferi infection was reported.[19]

Histopathologically APP shows normal or slightly atrophic epidermis with flat rete ridges. The mid or deep dermis shows interstitial edema and mild perivascular infiltrate consisting of lymphocytes and histiocytes may be present. Collagen bundles show varying degree of homogenization and clumping. Dermal thickness is reduced when compared to adjacent normal skin. The eccrine sweat glands, sebaceous glands and hair follicles are normal and elastic fibers appear normal.[20,9,21]

APP should always be differentiated from morphea.[21]

MANAGEMENT

APP is a benign condition but is cosmetically distressing. It is mostly a diagnostic challenge and need detailed history, careful physical examination, many laboratory investigations and dermatopathology.

No definite treatment for APP exists. Some reports have described beneficial treatment with the use of hydroxychloroquine 400 mg daily with marked improvements after 5-months and complete clearance of facial and trunk lesions after one year.[22] Cases with Borrelia burgdorferi infection can be treated by oral penicillin 2 million IU (international unit) per day or oral tetracycline 500 mg three times daily for 2–3 weeks. Show clinical improvement.[13]

Associated pigmentation could be treated successfully with Q-switched Alexandrite laser.[23]

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