Biomedical & Pharmacology Journal Vol. **8**(Spl. Edn.), 491-493 (Oct. 2015)

Atrophoderma of Pasini and Pierini - A Case Report

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DOI: http://dx.doi.org/10.13005/bpj/731

(Received: July 25, 2015; accepted: September 10, 2015)

ABSTRACT

Atrophoderma of Pasini and Pierni is a type of dermal atropy of unknown etiology. It presents as one or several, sharply depressed patches, with no surrounding erythema and induration. Commonly occurs in young adults, usually in the trunk. Atrophoderma of Pasini and Pierini is primarily an atrophic variant of morphea or a separate distinct entity is still debated.

Key words: Atrophoderma, young adult and cliff-drop borders.

INTRODUCTION

In 1923, Pasini described an atrophic lesion on the trunk which he reported as progressive idiopathic atrophoderma. In 1958, Canizares et al renamed this condition as 'Idiopathic atrophoderma of Pasini and Pierini'[1]. It is usually a benign disorder which presents as asymptomatic, hyperpigmented, slightly depressed patches with sharp "cliff-drop" borders with symmetrical and bilateral distribution.

Case report

A 29 year old man presented to our skin OPD with a dark colored asymptomatic skin lesion over the back for past 8 months. No history of other skin lesions in the body and pain. Dermatological examination revealed 8cm x 5cm single, well circumscribed, hyperpigmented, atrophic, depressed plaque with characteristic cliff-drop borders. No induration and tenderness present. Systemic examination was normal (Fig. 1-2).

Routine laboratory investigations such as CBC, urine analysis, RFT, LFT and serum electrolytes were normal.

A skin biopsy was taken from the plaque in the upper back. Histopathological examination [Fig-3] showed normal epidermis, collagen clumped and homogenous and high uptake of eccrine glands.

Diagnosis [Atrophoderma of Pasini and Pierini] was made based on clinical and histopathological findings.

DISSCUSION

Synonyms: morphea plana atrophica, Sclerodermie atrophique d'emblee, dyschromic & atrophic variation of scleroderma[2].

In 1923, Pasini described an atrophic cutaneous lesion on the trunk which he reported as progressive idiopathic atrophoderma. Pierini studied and defined the condition and its possible link to morphea in 1936. In 1958 Canizares et al. renamed this condition as 'Idiopathic atrophoderma of Pasini and Pierini'[1]. Exact etiology is not known. Whether atrophoderma is primarily an atrophic variant of morphea or a separate distinct entity is still debated [3] Role of Borrelia burgdorferi has also

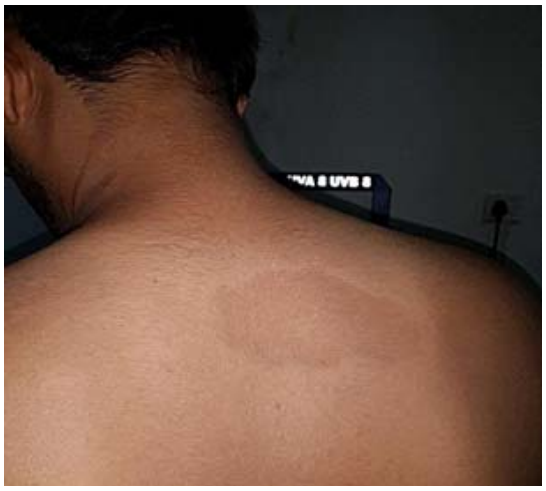

Fig. 1:

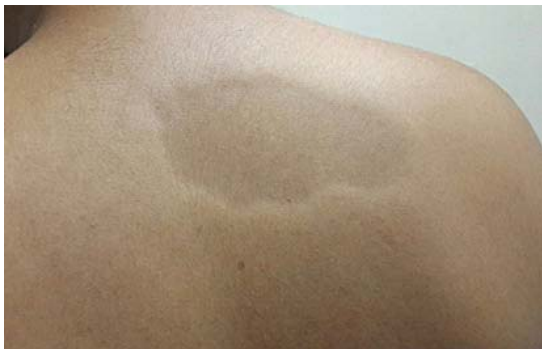

Fig. 2:

Fig. 1-2: Shows clinical picture of single, well circumscribed, hyperpigmented, atrophic, depressed plaque, with characteristic cliff-drop borders

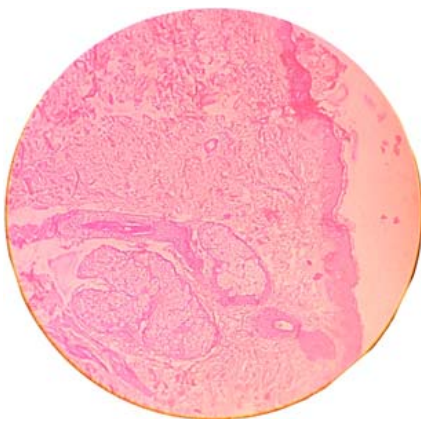

Fig. 3: Histology shows normal epidermis, collagen clumped and homogenous and high uptake of eccrine glands

been suggested [4]. This disorder is more frequently encountered in women than in men [5], with a ratio of 6:1 in adults. It usually starts insidiously during the second or third decade of life, but it is also been reported in children younger than 13 years of age. It presents as one or several, asymptomatic, round or oval shaped, sharply depressed, hyperpigmented, patches about 2cm to several centimeters in diameter with no induration and surrounding erythema. The borders are sharply defined, and they are described as, "cliff drop" borders with a depth of 1 to 8 mm. These borders can also have a gradual slant. These depressed patches are characteristic and give the impression of inverted plateaus, if multiple lesions are present they can have Swiss Cheese appearance [3]. Commonly seen in trunk ie upper back and lumbosacral region, but also seen in chest, arms and abdomen. The distribution is often symmetrical and bilateral. It has a benign course, but development of sclerodermatous changes has also been observed.

Histopathological examination in early lesion shows perivascular inflammatory infiltrate consisting of marcophages and T lymphocytes in dermis with odematous collagen bundle and scanty clumped elastic tissue. Older lesion shows normal or atrophic epidermis. Increased pigmentation of basal layer. Thickness of the dermis is reduced when compared to surrounding skin. Clumped and homogenous collagen bundles seen in the reticular dermis. Preserved appendages are seen.

Immunofluorescence studies may show IgM and C3 in the dermal blood vessels[6].

Striking clinical and histological features like that of Morphea intervenes the relationship between the atrophoderma and same . Earlier onset and protracted course of 10 to 20 years and absent violaceous ring surrounding the lesions may suggest atrophoderma as separate disease entity[6].

No treatment has been proved effective, but PUVA, hydroxychloroquine and Q-switched Alexandrite laser has been tried [7]. In view of the possibility of an underlying *Borrelia* infection, penicillin and tetracycline has been used for treatment.

CONCLUSION

Atrophoderma of Pacini and Pierini is a type dermal atropy with unknown etiology, whether it's a separate disease entity or primary variant of morphea has to be further studied.

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