Atrophoderma of Pasini and Pierini on the Head and Trunk of a 32 Year-Old Male: A Case Report*

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ABSTRACT

Atrophoderma of Pasini and Pierini (APP) is an idiopathic disorder of dermal atrophy, presenting as round or oval depressed lesions most commonly observed on the trunk and extremities. Herein, we report the first case of APP initially appearing on the head. Histopathologic examination of lesional skin demonstrated a decrease in elastic fibers. Serologic tests for Borrelia and antinuclear antibody titers were negative. There was no noted improvement after 6 weeks tretament with Hydroxycholoroquine 200 mg/tab BID. Definitive disease etiology and treatment of APP remains to be elucidated.

Keywords: Atrophoderma of pasini and pierini, dermal atrophy, elastic fibers

INTRODUCTION

Atrophoderma of Pasini and Pierini (APP), is a rare disease of dermal atrophy with less than 100 cases reported in literature. It is usually seen in the second or third decade of life, with a marked female preponderance. It presents as one or several sharply demarcated depressed patches, usually on the back, chest, abdomen, and extremities. Herein, we present the first reported case of a male patient with APP lesions initially appearing on the head, followed later on by lesions appearing on the trunk.

CASE HISTORY

A 32-year-old man presented to our clinic in 2016 for evaluation of long-standing hyperpigmented and atrophic lesions over the bitemporal areas and trunk which were present for seven years. In 2009, the patient noticed initial involvement of both temporal areas; the findings were described then as hyperpigmented macules along the hairline. The lesions grew in size and coalesced into extensive hyperpigmented patches over the next seven years. New discrete hyperpigmented macules and patches appeared on the pelvic areas of the trunk. The color of the lesions remained hyperpigmented and unchanged throughout the disease course. Six months prior to consult, lesional skin over the temporal areas and some on pelvic areas were noted by the patient to become progressively depressed in comparison to the surrounding normal skin. The lesions on the bitemporal areas particularly caused the patient cosmetic concern, which prompted consult at our institution. The patient reported no obvious accompanying symptoms other than occasional pruritus over the lesions on the temporal area. He denied sustaining trauma prior to the appearance of the lesions, any significant past medical history, use of topical or oral medications; and family history was non-contributory.

Upon physical examination, there were sharply-marginated, brown atrophic patches over the bitemporal areas (Figure 1A B) the anterior borders of which showed distinct "cliff-drop" borders (Figure 1C). Multiple, discrete hyperpigmented macules and atrophic patches were also seen over the pelvic areas (Figure lD, E), extending posteriorly to the lumbar areas (Figure lF). Lesional skin was normal on palpation, with no evidence of sclerosis nor induration. Examination of the hair, nails, and mucous membranes was normal. Review of systems was negative.

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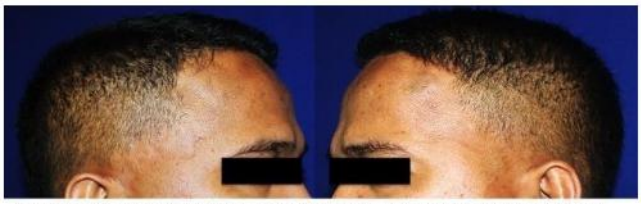

Figure 1A, B. Sharply-marginated, brown atrophic patches over the right and left bitemporal areas, respectively.

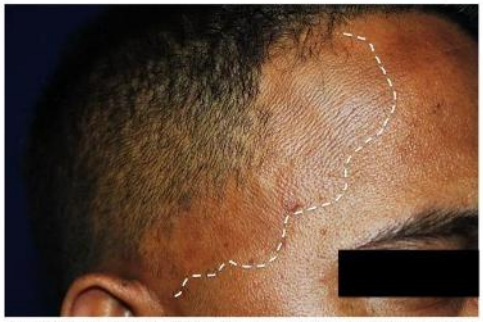

Figure 1C. Characteristic distinct "cliff-drop" border appreciated on the right temporal lesion

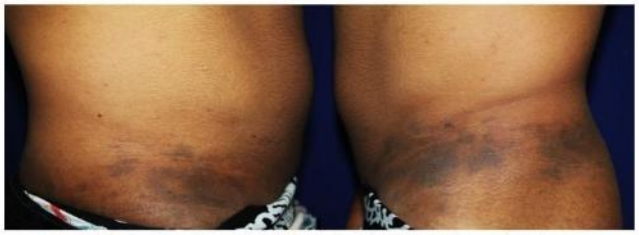

Figure 1D, E. Multiple, discrete hyperpigmented macules and atrophic patches on the pelvic areas.

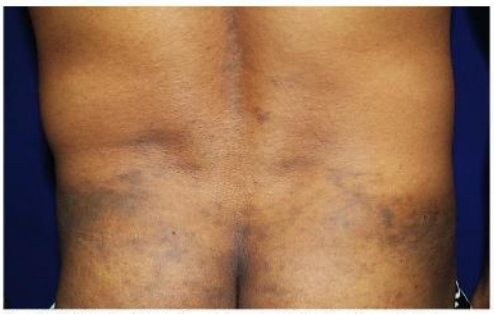

Figure 1F. Multiple, discrete hyperpigmented macules and atrophic patches on the lumbar area.

Histopathologic examination of skin punch biopsies taken from both the bitemporal and pelvic areas revealed no significant alteration in the epidermis (Figure 2A, B). Sections taken from the temporal area revealed a mild superficial perivascular infiltrate in the dermis composed of lymphocytes and melanophages, and thickened collagen bundles with spaces in between (Figure 2C, D). Verhoeff-Van Giesson staining was done, which showed loss of elastic fibers in the papillary and mid-dermis (Figure 2E, F).

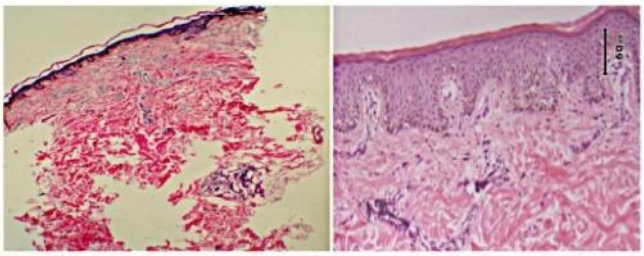

Figure 2A, B. Sections from the temporal and pelvic areas, respectively, showing normal epidermis, H&E, 40x.

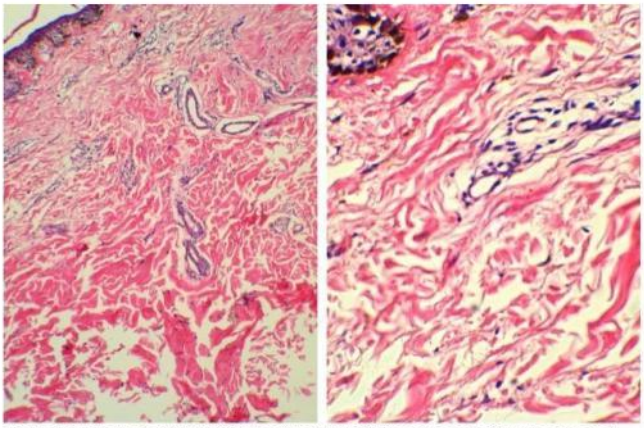

Figure 2C, D. Dermal mild superficial perivascular infiltrate composed of lymphocytes and melanophages, and thickened collagen bundles with spaces in between, H&E, 100x.

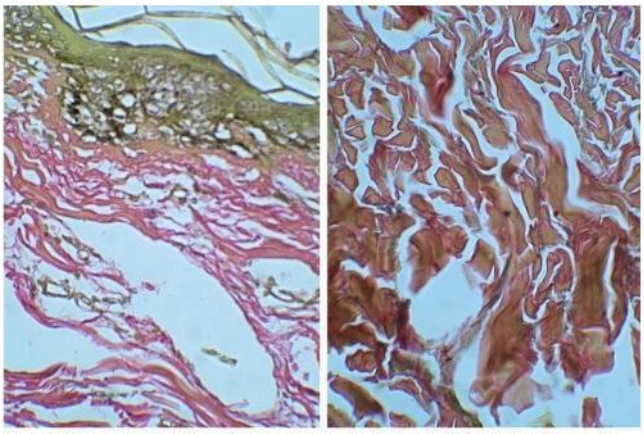

Figure 2E, F. Loss of elastic fibers in the papillary and mid-dermis. Verhoeff-Van Giesson, 40x.

Other laboratory testing included complete blood count, urinalysis, and tests for both liver and kidney functions, which were all within normal range. To rule out the possibility of Borrelia burgdorferi as an infectious etiology as has been

stated in previous case reports, enzyme-linked immunosorbent assay for borrelia IgG and IgM was done, which were both non-reactive. Antinuclear antibody titers were likewise negative. Clinicopathologic diagnosis was Atrophoderma of Pasini and Pierini. Treatment of APP remains anecdotal, and the negative borrelia serologies pointed towards the use of antimalarials instead of antibiotics and thus the patient was started on Hydroxychloroquine 200 mg/tab BID. Ophthalmologic clearance was obtained, and baseline laboratory determinations were done prior to starting the medication. Lesion appearance remained unchanged after 6 weeks of treatment.

DISCUSSION

Atrophoderma of Pasini and Pierini is a cutaneous condition characetrized by varying degrees of dermal atrophy. First described by Pasini in 1923, and by Pierini and V ivoli in 1936, the disease went by many names, until the official diagnosis of Atrophoderma of Pasini and Pierini was suggested in 1958 [10]. The exact world-wide incidence of APP is unknown. Much of the scarcity of data regarding this condition can be attributed to it being underreported since it is largely asymptomatic. This disorder is more frequently encountered in women than in men, with a ratio of 6:1 [9]. It starts insidiously during the second or third decades of life [1], although a congenital case was recently reported [4].

The lesions are often distributed bilaterally and in a symmetrical fashion [1], although a zosteriform distribution has also been observed [S]. Lesions have been reported most commonly to occur on the trunk, especially on the back and lumbosacral region, followed in frequency by the chest, arms, and abdomen. Characteristic lesions initially appear as single or multiple round or oval patches, 2 to 3 cm in diameter, which persist and may progress into atrophic patches over many years. The affected skin is described as having an inverted plaque appearance, which Stoner and Dixon described as resembling "footprints in the snow" *6+. Atrophic patches may grow and coalesce to encompass large areas of the trunk and proximal extremities, resulting in sharply demarcated edges referred to as "cliff drop" borders. The disease tends to be progressive over a course of 10-20 years. Eventually, no further progression occurs, yet existing lesions characteristically do not involute. Diagnosis is based on clinical findings, with histology playing a role in excluding other diagnoses.

The histopathologic changes in APP are often minimal and non-diagnostic, consist of a decrease in the size of the dermal papillae, with flattening of the rete ridges. 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 subsequent reduced dermal thickness.

The etiology and exact nosology of APP have always been a matter of debate. Some authors have linked it to infection with Borrelia burgdorferi, such as Buechner and Rufli [11] who studied the sera of 26 patients with typical APP lesions. Ten of the 26 patients had significantly elevated titers of IgG anti-Borrelia burgdorferi antibodies. However, none of the patients had an elevated IgM titer. The role of B. burgdorferi in APP is unclear, but some physicians have utilized positive test results for B. burgdorferi as support for treating APP with antibiotics. Some authors believe it as well to be a variant of localized scleroderma or morphea. Buechner and Rufli describe APP as an "abortive primarily atrophic variant of morphea" *11+ and Amano et al believe that morphea and APP are a "similar nosologic entity" *12+. Although similarly atrophic in nature the conditions can be distinguished by their contrasting age of predilection, duration of disease course, and by the striking lack of inflammation in lesions of APP. Lesions of morphea are ivory with a lilac border and tend to appear more inflamed, exhibiting induration, edema, and erythema [6]. In contrast, atrophic lesions of APP are violaceous or brown, with sharply demarcated borders that coalesce over time, producing a moth-eaten appearance that is not consistent with morphea. Whether atrophoderma is a nonsclerotic, primarily atrophic variant of morphea or a separate distinct entity is still debated [3].

No definitive treatment for APP exists. 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 of APP lesions after three treatments in one case [7]. Two case reports exist in which oral therapy was instituted for APP: one employed 200 mg/day of Doxycycline for a patient that had positive Borrelia antibodies, in which improvement was seen after six weeks [14], while in 2006, Carter et al [8] described a 38-year-old female patient who responded dramatically to a 400-mg daily dose of hydroxychloroquine. The patient had positive antinuclear antibody serology for which underlying lupus was suspected. She also had low levels of rheumatoid factor, and no lyme disease antibodies detectable on serology and experienced "notable improvement" in all lesions after 5 months and complete clearing of facial and trunk lesions after 1 year. Our patient received Hydroxychloroquine 200 mg/tab BID for 6 weeks which did not improve the patient's lesions. We believe that Hydroxychloroquine may still prove to be a viable treatment option in patients with APP, more so in those with positive atinuclear antibodies, and given over a more prolonged treatment period.

CONCLUSION

This is the first reported case of APP to occur initially on the head of a male patient, with subsequent involvement of the trunk. Though the disease remains stable and with a favorable prognosis, it can have cosmetic implications. There remains a need to determine a definitive, albeit effective treatment for APP.

REFERENCES

1. Goldsmith L., Katz S., Barbara A., et al. Fitzpatrick's Dermatology in General Medicine 8th edition, United States, 2012.
2. Pramod Kumar. Atrophoderma of Pasini and Pierini. Indian Dermatology Online Journal- December 2011, Vol.2, Issue 2, 126-128. [DOI: 10.4103/2229-5178.86012]
3. Kencka D Blaszczyk M Jabłooska S Atrophoderma Pasini-Pierini Is a Primary Atrophic Abortive Morphea. Dermatology 1995;190:203-206.DOI:10. 1159/000246685]
4. Kim SK, Rhee SH, Kim YC, Lee ES, Kang HY. Congenital atrophoderma of Pasini and Pierini. *J Korean Med Sci* 2006;21(1):169-71.[PMID: 16479086]
5. Wakelin SH, James MP. Zosteriform atrophoderma of Pasini and Pierini. Clin Exp Dermatol1995;20:244-6. [PMID: 7671424]
6. Stoner MF, Dixon SL. Purple depressions on the trunk and extremities. Atrophoderma of Pasini and Pierini. *Arch Dermatol* 1990;126(12): 1641,1644 [PMID: 2256692]
7. Arpey CJ, Patel DS, Stone MS, Qiang-Shao J, Moore KC. Treatment of atrophoderma of Pasini and Pierini-associated hyperpigmentation with the Q- switched alexandrite laser: A clinical, histologic, and ultrastructural appraisal. Lasers Surg Med 2000;27: 206-12. [PMID: 11013382]
8. Carter JD, Valeriano J, Vasey FB.Hydroxychloroquine as a treatment for atropho-derma of Pasini and Pierini. *Int J Dermatol* 2006;45(10):1255-6. [PMID: 17040461]
9. Pullara TJ, Lober CW, Fenske NA. Idiopathic atrophoderma of Pasini and Pierini. *Int J Dermatol* 1984;23(10):643-5. [PMID: 6396245]
10. Canizares 0, Sachs P, Jaimovich L, Torres VM. Idiopathic atrophoderma of Pasini and Pierini. *AMA Arch Derm* 1958;77(1):42-60.[DOI:10.1001/ archderm.1958.01560010044007]
11. Buechner SA, Rufli T. Atrophoderma of Pasini and Pierini. Clinical and histopathologic findings and antibodies to Borrelia burgdorferi in thirty-four patients. J Am Acad Dermatol 1994;30(3):441-6. PMID: 8113457]
12. Amano H, Nagai Y, Ishikawa 0. Multiple morphea coexistent with atrophoderma of Pasini-Pierini (APP): APP could be abortive morphea. J Eur Acad Dermatol Venereol 2007:21(9):1254-6. PMID: 17894716]
13. Wojas-Pelc A, Wielowieyska-Szybinska D, Kieltyka A. Presence of the antinuclear antibodies and antibodies to Borrelia burgdorferi among patients with morphea en plaque, deep linear scleroderma and atrophoderma Pasini-Pierini [in Polish]. Przeglad Lekarski 2002;59(11):898-902. [PMID: 12715718]
14. Yoonhee L, Yoonseok 0, Seok-yong A, et.al. A case of atrophoderma of Pasini and Pierini associated with borrelia burgdorferi infection successfully treated with oral doxycycline. Ann Dermatol Vol. 23, No. 3, 2011. [PMID: 21909207]