SKIN

BRIEF ARTICLE

Zosteriform Atrophoderma of Pasini and Pierini: A Case Report

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ABSTRACT

Atrophoderma of Pasini and Pierini (APP) is a rare cutaneous entity of unknown etiology. It usually presents with sharply demarcated dermal atrophy, giving a “cliff-drop” appearance. Very few cases occurring in a zosteriform distribution have been reported. In this article, the author reports a rare case of APP in a 42-year-old women where the lesions presented in a zosteriform distribution on the trunk. Diagnosis is made with high clinical suspicion and confirming histopathology.

CASE REPORT

A 42-year-old woman presented with a 6-year history of an asymptomatic, stable rash on her abdomen. The rash had been previously treated by her primary care physician, as intertrigo, with topical antifungals. The patient denied any history of trauma to the site or preceding rashes, including herpes zoster. No personal or family history of autoimmune conditions. Past medical history was noncontributory.

Physical examination demonstrated a Fitzpatrick skin type V with a dark brown, sharply demarcated, atrophic, linear patch on her right flank extending to her right lower abdomen in a zosteriform distribution **(Figure 1)**. No induration or sclerosis was observed. The remainder of the physical examination was unrevealing, except for striae and crusted erosions from prior biopsy. The clinical differential diagnosis included atrophoderma of Pasini and Pierini (APP), morphea, post-inflammatory hyperpigmentation with atrophic scarring from herpes zoster.

A punch biopsy revealed subtle dermal atrophy, mild edema in the mid and deep dermis, a mild superficial lymphohistiocytic perivascular infiltrate and sparse melanophages in the papillary dermis **(Figure 2A)**. Verhoeff stain showed decreased and fragmented elastic fibers **(Figure 2B)**. The findings compatible with atrophoderma.

APP is a rare, cutaneous condition that causes dermal atrophy of uncertain etiology. There is controversy regarding whether it falls within the spectrum of morphea.[1] The role of *Borrelia burgdorferi* infection as an inciting factor is also controversial.[2] This condition commonly appears in the second or third decade of life and has a strong female predilection (5:1). [3] It is more frequently reported in Whites. It usually presents with bilateral and symmetric round macules or patches that are sharply demarcated and depressed, giving a “cliff-drop” border

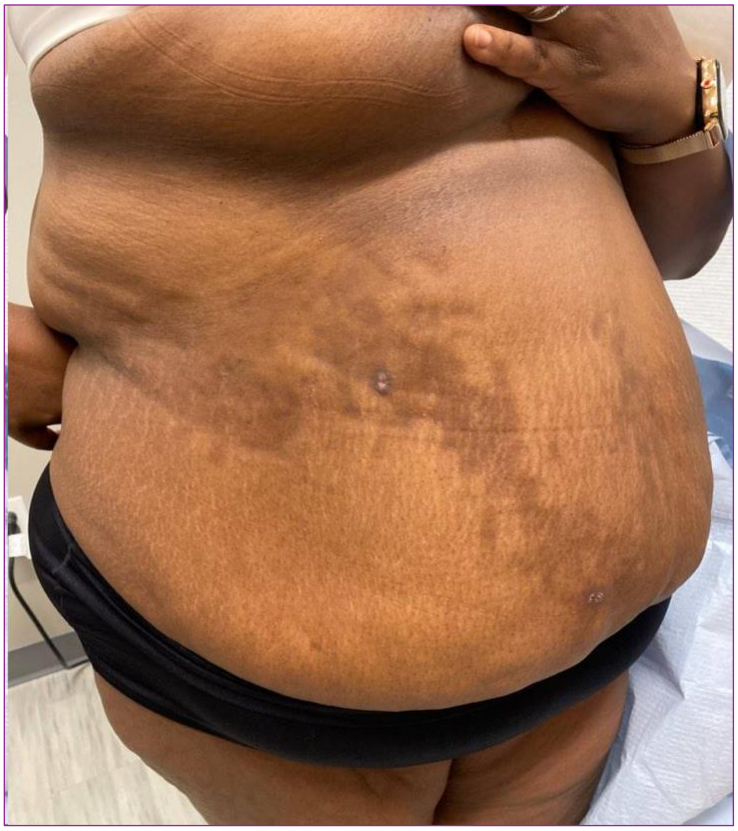

Figure 1. Patient with sharply demarcated, atrophic patch in a zosteriform distribution on the right flank extending to the right lower abdomen.

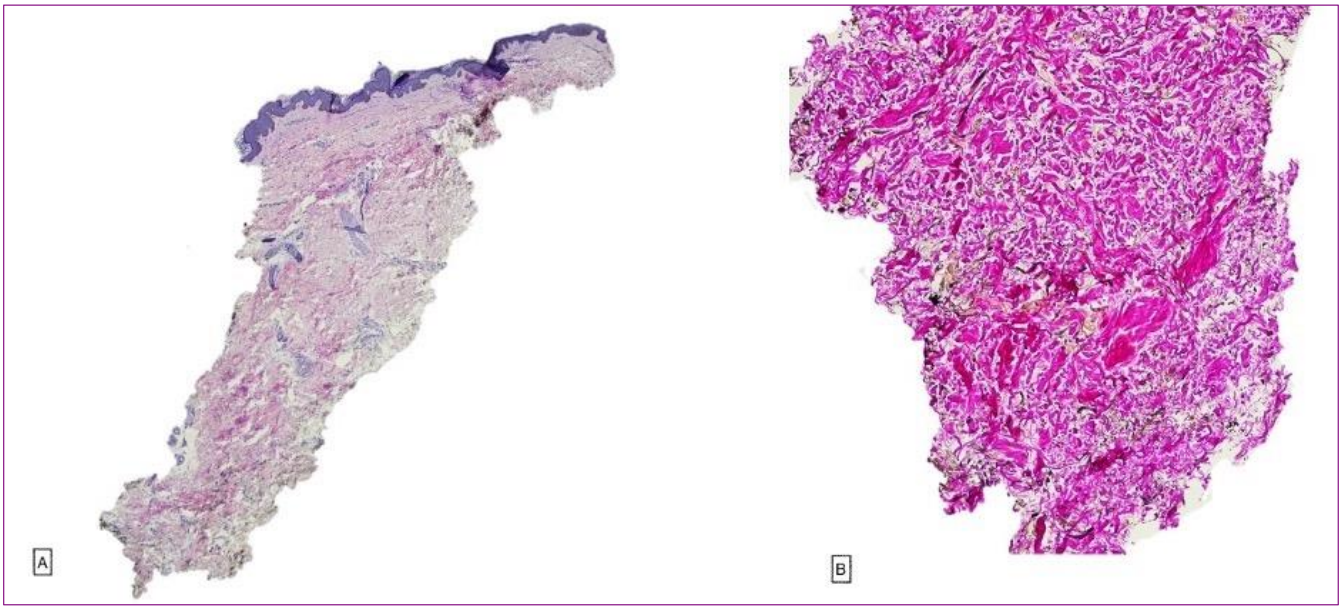

Figure 2. A, Subtle dermal atrophy with associated mild edema in the mid and deep dermis. Mild superficial perivascular infiltrate of lymphocytes and histiocytes with few melanophages in the papillary dermis (hematoxylin-eosin [H&E], magnification 4x). B, Fragmented elastic fibers with decreased density (Verhoeff stain, magnification 100x).

Table 1. Summary of the reported cases in the English literature.

Case	Gender/ Age (yr)	Site	Duration	History of Herpes Zoster	Antibodies	Immunofluorescent Antibodies
Wakelin S.H.	M / 53	Left Trunk	12 years	Unknown	No	Negative IgA, IgG, IgM, and Complement C3
Zheng Y et al.	F /24	Right Chest/ Arm	2 years	No	No	Negative ANA
Arif T.	M / 18	Left Chest	1.5 years	Unknown	No	Negative ANA
This Case	F / 42	Right Flank/ Abdomen	5 years	No	No	Unknown

appearance. The lesions range greatly in size from a few millimeters to greater than 20 cm. APP typically appear on the trunk, especially the back, and show no obvious signs of inflammation or sclerosis.

Diagnosis can be challenging requiring high index of clinical suspicion. APP often presents with subtle histological features making it more difficult to identify. Dermal atrophy may not be appreciated unless normal skin is available for comparison. The epidermis is expected normal and is often hyperpigmented. Few melanophages may be seen in the papillary dermis. Subtle perivascular inflammatory infiltrate, consisting of lymphocytes and histiocytes, is seen in the superficial dermis. The elastic fibers are typically preserved, but fragmented and decreased elastic fibers have been reported, which was also seen in this case.[3] APP tends to variably progress, with new lesions developing for one to two decades, and then remains stable. Currently, there is no effective treatment for APP. However, treatments have included psoralen plus ultraviolet A, potassium benzoic acid, and oral antibiotics in patient with elevated *B. burgdorferi* antibodies (IgM).[1]

Zosteriform distribution of several dermatological conditions, including morphea, has been associated with “Wolf’s isotopic response”. This term describes the occurrence of a new skin disorder at the site of another unrelated and already healed skin disease.[4] Zosteriform APP, which appears in a dermatomal distribution, is a rare entity that has been reported. To our knowledge, there are three other cases previously reported in the literature **(Table 1)**.[1,2,5] Interestingly, none of the reported zosteriform APP cases had preceding herpes zoster. We report here one additional case of this unique presentation of APP to help increase recognition of this rare entity. Lastly, this case serves as a reminder that the relationship between the dermatologist and the dermatopathologist remains a fundamental steppingstone for making an accurate diagnosis.

Conflict of Interest Disclosures: None

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