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A Rare Case of Idiopathic Atrophoderma of Pasini and Pierini

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Abstract

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is a rare pigmented atrophic cutaneous disease. Here, we report a case of a 27-year-old woman who presented with asymptomatic brown patches on her upper back for one year. There was no prior history of swelling or induration of the lesions. Physical examination showed atrophic patches with clear boundaries, consistent with snow footprints or cliff-drop-like changes on edge. Dermoscopy revealed numerous bare linear, branched blood vessels and perifollicular brownish areas, which differs from only one previously reported IAPP. Biopsy showed perivascular mononuclear cells infiltrating the dermis and reduced elastic fibers in the upper and middle dermis. Typical clinical findings of “footprints in the snow” appearance and dermoscopy could combine for a definite diagnosis aiding in differentiating from morphea presenting with whitish structureless and hairless areas. This case report might stimulate clinicians to diagnose such a rare disease in patients with similar lesions to avoid unnecessary treatment.

Key words: Atrophoderma of Pasini and Pierini; Dermoscopy; Idiopathic; Rare disease

Dear Editor,

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is a rare cutaneous pigmented atrophic disease predominantly affecting young women with a peak incidence in the second and third decades. It is preferably located on the trunk and limbs, rarely involving the face. Here we report a rare case of IAPP highlighting its clinical and dermoscopic features.

A 27-year-old Chinese female presented with asymptomatic brownish patches on her upper back for one year. In the beginning, a palm-sized pigmented patch appeared on her lower side of upper back which gradually extended, and a similar mirror lesion arose on upper side of upper back. Growth of classic lesion was consistent with “footprints in the snow” like appearance. There was no swelling or induration in the lesions. Her family and medical history was unremarkable. Physical examination revealed two uniform brownish atrophic patches, smooth surfaces, and sharply irregular borders on the upper back with no sclerosis or infiltration on palpation (Figure 1A). Dermoscopy showed an irregularly distributed large number of bare linear and arborizing blood vessels. In addition, regular perifollicular whitish areas with hair and brownish pigmented areas were observed (Figure 1B).

Biopsy from the lesion revealed perivascular mononuclear cellular infiltration in upper dermis (Figure 1C) and reduced elastic fibers in the upper and middle dermis (Figure 1D), confirming IAPP. She was treated with 0.1% tacrolimus ointment for a month, showing slight improvement.

IAPP is an unusual disorder of dermal atrophy. The etiology remains uncertain, and the onset is insidious, without any symptoms of redness or swelling before and during the disease. IAPP has only approximately 100 reported cases.1 Only 20 cases of IAPP have been reported from China by the CNKI review. Lesions are mostly asymmetric palm-sized, with faintly visible blood vessels. Classic lesions are "footprints in the snow" or "Swiss cheese-like", with the “cliff-drop” borders and clear hyperpigmented depressions. Our

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case showed classical “footprints in the snow” like lesions on her back. Histopathologic features tend to be non-specific and are easily ignored. The epidermis varies from normal to mild atrophy and is often hyperpigmented in the basal layer. There may be mild perivascular mononuclear cell infiltration in the upper dermis and edema, homogenization, and clumping of collagen in the deeper dermis. Some studies have found changes in elastic fibers on Verhoeff-van Gieson stains elusive within a spectrum ranging from normal to severe diminution.2

Non-invasive examination for IAPP is very limited in the literature. Ultrasound examination revealed a slight decrease in dermal thickness3 and increased vascularity.4 Only one case reported an irregularly distributed, prominent pigment network, with some areas showing a wavy/storiform pattern on dermoscopy.5 The regular perifollicular whitish areas with hair were consistent with the dermoscopic findings by Kaliyadan F et al.5 Unlike the features of morphea in dermoscopy, showing whitish structureless areas with the absence of hair resulting from reticular dermal fibrosis.5 Non-invasive procedures are not helpful but might assist in differentiating from morphea.

IAPP may demonstrate controversial findings about a distinct entity or a form of localized scleroderma. However, its classical clinical findings of snow footprints or cliff-drop-like changes on the edge of the lesion may help differentiate from indurated, whitish structureless and hairless morphea.

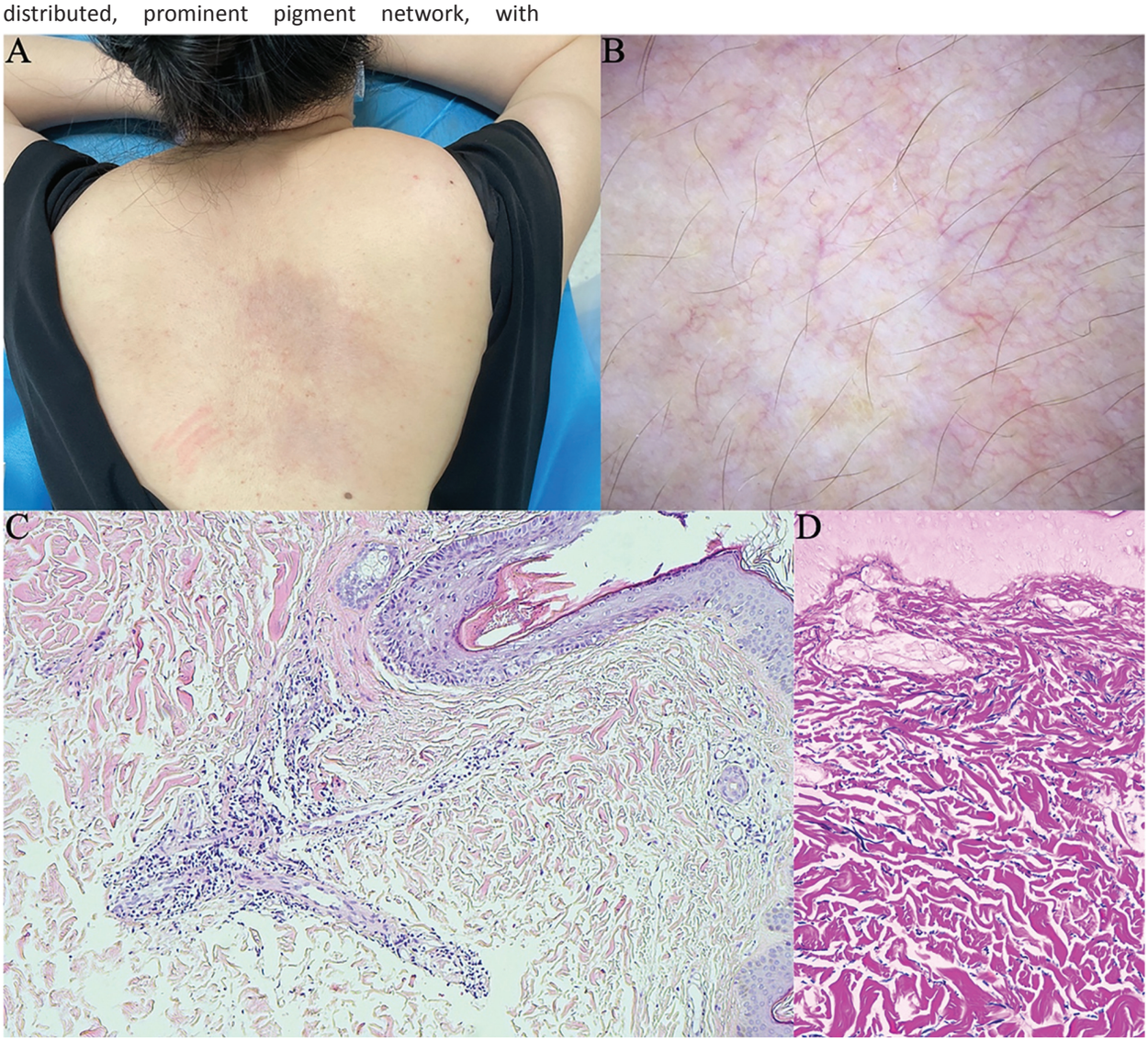

Figure 1: (A) A 27-year-old female showed two pigmented brownish atrophic patches with sharp and irregular borders on the upper back. (B) Polarized dermoscopy of the patch showed an irregularly distributed large number of bare linear and arborizing blood vessels. In addition, regular perifollicular whitish areas with hair and brownish pigmented areas were also observed (original magnification ×10). (C) Histopathology of the patch revealedperivascular mononuclear cells infiltrating upper dermis (hematoxylin and eosin [HE], original magnification ×100). (D) The elastic fibers in the upper and middle dermis were reduced (Verhoeff-Van Gieson, original magnification ×200).

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