A Picture Says a Thousand Words

Multiple depressed plaques over back in a young boy

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CASE DESCRIPTION

A 12-year-old boy presented with a 6-month history of asymptomatic but gradually progressive depressed lesions over the back. There was no history of any preceding local trauma, infection, ulceration, or any inflammatory dermatoses. He was not on any medications and his family members had no history of similar skin lesions. On examination, multiple discrete, sharply demarcated, hyperpigmented, smooth-surfaced, nonindurated, noninflammatory atrophic plaques of varying diameters (2 cm to 7 cm) were noticed dispersed over the back; an abrupt border transitioning into normal skin gave it the typical ‘cliff drop’ appearance (Figure 1). Other mucocutaneous and systemic examination was unremarkable. Routine laboratory investigation was noncontributory and serum antibody for *Borrelia burgdorferi* was negative. Histopathological examination revealed normal epidermis, hyalinization of collagen in dermis with minimal scattered inflammatory infiltrate, and decrease in the thickness of dermis compared to surrounding skin. What is the diagnosis?

Figure 1. Multiple well-demarcated hyperpigmented atrophic plaques over the back with an abrupt border giving an impression of ‘cliff drop’ appearance.

DIAGNOSIS

Based on distinctive clinical and corroborative histopathological features, a diagnosis of idiopathic atrophoderma of Pasini and Pierini (IAPP) was established. He was counselled about the benign nature of the condition; topical tacrolimus (0.1%) ointment was started twice daily with subsequent partial improvement of lesions in 3 months.

DISCUSSION

IAPP is a rare cutaneous disorder thought to affect dermal collagen organization resulting in skin atrophy. Pasini first described this condition in 1923 as 'progressive idiopathic atrophoderma' and subsequently its possible link to morphea was established by Pierini and Vivoli in 1936 (1). Finally, in 1958, Canizares et al. renamed this entity as 'idiopathic atrophoderma of Pasini and Pierini'. However, still there has been an ongoing controversy on whether IAPP represents an atypical, primarily atrophic form of morphea, or 'burnout' morphea or a separate entity (2). Although the exact cause of the disease is not known, *Borrelia burgdorferi* infection has been proposed to play a role in the pathogenesis as supported by findings of high *Borrelia* antibody titres in some patients with IAPP (3). Genetic predisposition, neurogenic factors, and immunological factors have also been considered as potential triggers for development of the disease (4).

IAPP is a benign, asymptomatic disease characterized by single or multiple sharply demarcated, round to oval, hyperpigmented, nonindurated patches of varying sizes ranging from a few millimetres to several centimetres. These patches are marked by a slight depression of the skin with an abrupt edge often exhibiting a 'cliff drop border'. (5). The typical area affected is the back, but other regions can be affected. The disease is typically seen in early adolescence with female preponderance; however, congenital onset, late onset, and manifestation in male patients have also been reported (6).

Histopathological examination revealed a characteristic decrease of dermal thickness with a mild perivascular infiltrate consisting of lymphocytes and histiocytes; collagen bundles show varying degrees of homogenization and clumping. The appendageal structures and subcutaneous tissue are usually intact (7). An enzyme-linked immunosorbent assay should be performed to detect anti-*Borrelia burgdorferi* antibodies.

This condition ought to be differentiated from morphea (indurated plaques with characteristic peripheral lilac rim), linear atrophoderma of Moulin (earlier onset, distributed linearly along Blaschko's line), and anetoderma (herniated sac-like skin showing loss of dermal elastic tissue).

The disease has typically been reported to have no consistently effective treatment. A course of doxycycline has proved to be beneficial in cases with documented *Borrelia burgdorferi* infection (8). Topical treatments with calcineurin inhibitors, corticosteroids, and retinoids have been used with variable success. Systemic steroids, antimalarials, D-penicillamine, and phototherapy have been reported to be effective in some cases. Q-switched alexandrite laser has found to be effective in diminishing the hyperpigmentation (9).

This benign disease has protracted, yet self-limiting course. New lesions tend to appear with existing lesions enlarging variably over 10 to 20 years before the disease become static in nature with stabilization of number and size of lesions. IAPP is usually a skin-limited disease without any significant complications or mortality. However, very rarely it has been found to be complicated with progressive systemic sclerosis and lung fibrosis (10).

The purpose of documenting this case is to create awareness among physicians to promptly diagnose the condition and ensure proper counselling to allay anxiety of the patient.

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