A Case of Unilateral Idiopathic Atrophoderma of Pasini and Pierini

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Idiopathic atrophoderma of Pasini and Pierini (IAPP) is an uncommon condition of unknown etiology and typically affects persons in the second and third decades of life. IAPP consists of asymptomatic grey to brown depressed lesions with “cliff drop” borders, which are usually bilateral and located on the trunk.

A 40-year-old woman visited our department because of unilateral atrophic lesions on the abdomen, right arm, and posterior aspect of right thigh for 20 years. Laboratory evaluations were normal and histologic examination revealed significantly decreased thickness in dermis.

We report an unusual case of idiopathic atrophoderma of Pasini and Pierini presenting unilateral distribution. (Ann Dermatol 14(1) 35-37, 2002).

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CASE REPORT

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A 40-year-old woman had asymptomatic atrophic lesions on the right abdomen, the proximal portion of the right arm and the posterior aspect of the right leg for 20 years. There were two, 12 × 20 cm sized and 5 × 8 cm sized, slate-grey or slightly brownish colored, depressed patches in the right abdomen those were not indurated (Fig. 1). In the proximal portion of right arm and the posterior aspect of the right thigh, the lesions showed slate-grey or slightly brownish colored, soft, atrophic macules and coalescing patches that had “cliff-drop” borders and had gradually spread to the adjacent areas (Fig. 2).

Laboratory examination including complete blood cell counts, routine urinalysis, liver function test, a test for antinuclear antibody and VDRL were negative or within normal limits.

Microscopically, the lesions showed an increase in basal pigmentation, markedly thinned dermis with mild perivascular lymphohistiocytic infiltrates and no atrophy of appendageal structure (Fig. 3). An elastic tissue stain showed a slight increase in elastic fibers, which were probably relative to the diminished dermis, but did not show the clumping of elastic fiber (Fig. 4).
DISCUSSION

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is an uncommon dermatologic condition of unknown etiology. Some studies suggested that IAPP may be associated with Borrelia infection. Opinions differ as to whether IAPP is a variant of morphea or a separate entity distinct from morphea. Some authors regard atrophoderma as a disorder consisting of two clinical types: The first, a true idiopathic atrophy that remains unchanged for many years, and the second, a type closely related to morphea either clinically, histologically, or both. In this latter variety, sclerotic changes may appear for several years following the onset of atrophic lesions. There are arguments on IAPP being a variant of morphea.

Arguments for IAPP being a variant of mor-

Fig. 1. Two, unilateral, slate-grey or brownish atrophic patches on the right side of the abdomen.

Fig. 2. Slightly brownish atrophic macules and patches that have “cliff-drop” borders on the posterior aspect of the right thigh.

Fig. 3. Histologic findings of the lesion in the abdomen shows an increase in basal pigmentation and markedly thinned dermis with mild perivascular lymphohistiocytic infiltrates but there are no atrophic change of the appendageal structure. (H&E stain: ×40)

Fig. 4. An elastic tissue stain shows a relative increase in elastic fibers but does not show the clumping of elastic fiber. (Verhoeff-van Gieson stain: ×100)
phea have been based on the following observations: 1) Lesions of “burnt-out” morphea may be atrophic and histologically similar to lesions of IAPP. 2) Some histologic similarity such as mild sclerosis and collagen homogenization exists in a few cases. 3) A case report described a patient with typical IAPP who developed progressive systemic sclerosis.

Arguments against IAPP being a variant of morphea include: Absence of the typical lilac ring in morphea, progressive usually for a much longer duration than those of localized scleroderma, preceding the sclerotic lesion of IAPP contrast to preceding the atrophic lesions of morphea, and histologically several distinctions.

Generally the disorder has a subtle onset and most often affects women during adolescence and young adulthood. The majority of patients are asymptomatic but a few patients have noted warmth or tingling in affected areas. The affected skin is characteristically bluish-violet to slate-grey or brownish colored, depressed, giving the impression of an “inverted plateau” and sharply defined. Abrupt borders form a “cliff-drop” ranging from 2 mm to 8 mm. In our case, brownish atrophic macules and patches having “cliff-drop” borders were also seen on the right arm and the right thigh. The most commonly involved site is the trunk, especially the back and abdomen. Only a few cases showed unilateral distribution. In Korean dermatologic literature, Kim et al. in 1985 firstly reported a case of progressive idiopathic atrophoderma and then three cases were reported. Most of them were distributed bilaterally and only one case showed zosteriform pattern.

Histopathologic findings in IAPP are not striking. The epidermis appears normal except for rare reports of atrophy. There is a quantitative decrease in the thickness of the dermis. Collagen bundles in the mid and deep dermis may show varying degrees of intracollagenous edema, but actual degeneration of collagen is not a feature. Clumping of elastic fibers in the lower dermis occurs concomitantly with dermal edema in some cases. Our case showed a significant decrease in dermal thickness but there was no clumping of elastic fibers.

We experienced an uncommon dermatologic condition of idiopathic atrophoderma of Pasini and Pierini which showed an unusual feature with unilateral distribution.

REFERENCES