

Inflamed bilateral linear atrophoderma of Moulin in an adult woman: a case report

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Abstract

Linear atrophoderma of Moulin (LAM) is a rare dermatosis characterized by a hyperpigmented atrophoderma that follows Blaschko's lines, with onset usually occurring during childhood and adolescence. LAM is an etiologically unknown form of dermal atrophy. It is generally characterized by oval or round atrophic, nonsclerotic, hyperpigmented patches following Blaschko's lines. These patches are usually located on the trunk and the upper and lower extremities. This case study discusses a patient that had the nonclassical form of LAM with the initial lesions presenting as papules.

Keywords: linear atrophoderma of Moulin, atrophoderma of Pasini and Pierini, inflammation

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Introduction

Atrophoderma of Pasini and Pierini (APP) is a rare dermatosis characterized by well-defined atrophic plaques (1). It is more common in young adults and women (2). Linear atrophoderma of Moulin (LAM) is an acquired condition that presents with unilateral, depressed plaques following Blaschko's lines (3). It usually affects children or adolescents (3). Baumann et al. considered LAM a variant of progressive idiopathic APP (4). On the basis of the clinical and histopathologic findings, this case was diagnosed as LAM.

Case report

A 29-year-old female patient was admitted to the clinic due to brown-gray spots on one buttock and both thighs. The patient stated that her complaints had started as numerous small bumps without tips on her buttock about 8 months earlier, that these bumps had also occurred for the first time on her nipple 2 days

earlier, that they healed with hyperpigmentation, and that she had not received any prior medical treatment for the bumps. There was no history of trauma, infection, or insect bites.

The patient's complete blood count, erythrocyte sedimentation rate, liver and kidney function tests, ANA, and anti-DNA were within normal limits in laboratory tests. The *Borrelia burgdorferi* serology test was negative.

On dermatologic examination, bilateral and erythematous papules on the buttock and thigh region and numerous round, sharp, limited depressed plaques were detected (Fig. 1). The lesions were brown-gray in color and had the central atrophic appearance of the plaques. No induration or sclerosis was detected in the lesions.

A histopathologic examination of the atrophic pigmented plaques showed thickening of the basal layer, increased melanin pigmentation, flattening of the dermal papillae, and mild perivascular lymphocytic infiltration (Fig. 2).

The patient was diagnosed with LAM after considering the clinical and pathologic findings. These findings also indicated that it was an inflammatory type of LAM that began with papules. Local corticosteroid cream therapy was initiated for the atrophic plaques



Figure 1 | Atrophic plaques on buttock and upper legs.

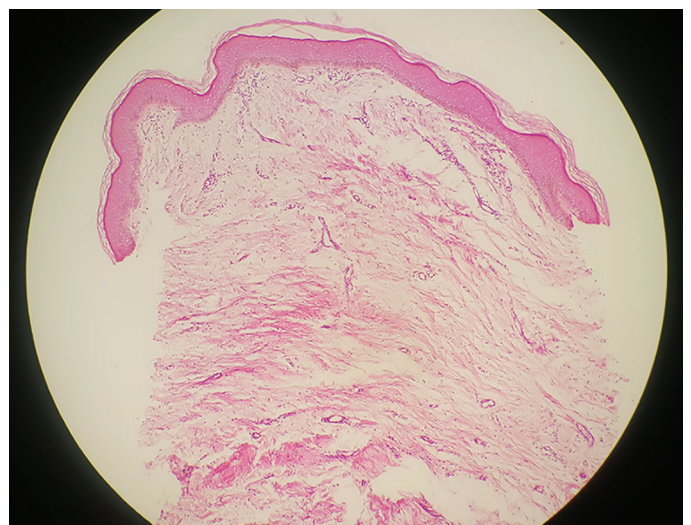


Figure 2 | Thickening in the basal layer and flattening in the dermal papilla (H&E 40x).

on the buttock and thighs, along with 100 mg doxycycline daily for the inflammatory papules. There was no regression in the papules at the end of two weeks of antibiotic treatment. Corticosteroid cream therapy was stopped at the end of two months due to no change.

Discussion

APP was first described by Pasini in 1923 (5). Pierini and Vivoli suggested in 1936 that APP may be associated with morphea. Yokoyama et al. suggested that the atrophy associated with APP is different from the classical morphea of skin glycosaminoglycans (6). Sharply demarcated and well-shaped (cliff-drop), brown-gray depressed plaques are typical of atrophoderma (7). Lesions are located symmetrically on the body, usually in the back and lumbosacral regions. Hand, foot, and upper extremity involvement is very rare. In some patients, zosteriform distribution was reported in parallel with skin folds. It is thought that the general course of the disease lasts for as long as 10 to 20 years and that there is no involution (1).

The etiopathogenesis of APP is not known (6). Previous studies have reported that genetic and neurogenic factors, as well as defects in dermatan sulfate metabolism, may cause this disease (2). Direct immunofluorescence studies have focused on immunologic factors due to the accumulation of IgM and C3 in small blood vessels and the detection with an electron microscope of T lymphocytes and macrophages between the perivascular region and collagen fibers (8).

It is thought that the disease does not currently have an effective treatment. Antibiotic therapy has been recommended in cases of a positive *Borrelia* antibody titer (9, 10). Treatment with systemic steroids, antimalarials, calcitriol, and phototherapy has been tried in sclerotic cases (11). In current treatment approaches, Q-switched lasers are used to treat hyperpigmentation in patients with cosmetic concerns (2, 3).

The histopathology of APP is usually not diagnostic. Early lesions have moderate chronic inflammatory cells, but this is not seen in the late period. Pigment enhancement can be seen in the basal layer (6). Skin attachments are usually protected.

Linear atrophoderma is a rare dermatosis first reported in five patients by Moulin et al. in 1992 (3). It is an acquired condition that presents with unilateral hyperpigmented atrophic patches following Blaschko's lines. The disease affects children and adolescents.

There are many clinical and histologic similarities between LAM and APP. APP may resemble LAM, but it does not follow Blaschko's lines. Some authors consider LAM a variant of progres-

sive idiopathic APP (3).

Based on the clinical and histopathologic findings, this case is thought to be compatible with LAM. The patient's initial complaints were inflammatory features such as papules, which are not compatible with the classical LAM clinical appearance. Inflammatory lesions, such as dermatitis cruris pustulosa et atrophicans, keratosis follicularis spinulosa decalvans, and other atrophic diseases, are considered to be distinctive. However, the lesions from dermatitis cruris pustulosa are usually found in the form of pustules and in the lower extremities, whereas keratosis follicularis spinulosa decalvans starts with more erythematous papules and results in hairy deep cicatricial alopecia (12, 13). The other two diseases were excluded based on the clinical and histopathologic findings.

In the differential diagnosis two LAM cases were encountered, each starting with inflammatory papules as defined by Browne and Fisher, who suggested that this disease has inflammatory and non-inflammatory variants (11). Pasini and Pierini stated that there may be variants due to the clinical and histopathologic similarity of LAM and atrophoderma (10). In another case report of LAM, pigmentation and atrophy treated with 20 mg per week of methotrexate were reported to decrease for 6 months (10). Because of the efficacy of methotrexate, LAM, APP, and linear scleroderma are thought to be variants of the same disease (10). There are hyperpigmented atrophic plaques in LAM, just as in APP, usually located on the trunk and extremities. However, these plaques are parallel to Blaschko's lines in LAM but not in APP. Like the case reported by Tan and Tay (10), which started as inflammatory papules but developed into atrophic plaques following Blaschko's lines, the case presented here is one of LAM that started with inflammation. Although this could be considered an atypical variant of LAM, Browne et al. reported a case with antecedent inflammation and suggested that there are two variants of LAM—inflammatory and non-inflammatory—and that an antecedent inflammatory phase ultimately may evolve into hyperpigmentation with atrophy (14, 15). Later, Utikal et al. described two patients with prominent telangiectatic erythema within the lesions of linear atrophoderma and argued that these cases may represent a novel variety of LAM or a separate entity (14, 16). Although LAM usually starts in childhood or adolescence (between 5 and 20 years of age) (10), cases have developed in two subjects over 30 years old (17, 18). Even though Moulin et al. presented unilateral localizations, there are five reports in the literature of bilateral LAM (15, 16, 19–21). Furthermore, in most of the patients LAM occurred on the trunk and limbs. The case presented here is noteworthy because there are few cases of inflammatory LAM reported in the literature.

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