Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report

DOI: 10.6003/jtad.17113c1

Inflamed Bilateral Linear Atrophoderma of Moulin in Adult Woman: A Case Report

Yesim Akpinar Kara[1], MD, Evren Sarifakioglu[2], MD

Address: [1]Ufuk University, Department of Dermatology, [2]Ankara, Private Dermatology Clinic, Ankara, Türkey
E-mail: yesim_akpinar@yahoo.com
** Corresponding Author:* Dr.Yesim Akpinar, 1428.sk No:16/8, Çukurambar, Ankara, Turkey

Published:
J Turk Acad Dermatol 2017; **11 (3)**: 17113c1
This article is available from: http://www.jtad.org/2017/3/jtad17113c1.pdf
Key Words: Lineer atrophoderma of Moulin, Atrophoderma Pasini and Pierini, inflamation

Abstract

Observation: Linear Atrophoderma of Moulin (LAM) is a rare dermatosis characterized by a hyperpigmented atrophoderma that follows Blaschko's lines with onset usually during childhood and adolescence. LAM is etiologically unknown form of dermal atrophy. Generally they were characterized as ovally or round shaped, atrophic, non-sclerotic, hyperpigmented patches following the lines of Blaschko. These patches are usually located in the trunk in addition to upper and lower extremities. In this study, we discussed about a case who had the nonclassical form of LAM with the initial lesions as papules.

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 unilateral, depressed plaque following the lines of Blaschko.[3] It affects usually children or adolescents [3]. Baumann et al.considered LAM is a variant of progressive idiopathic atrophoderma of Pasini and Pierini. On the basis of the clinical and the histopathologic findings, we diagnosed the case as linear atrophoderma of Moulin.

Case Report

29 years old female patient was admitted to our clinic due to resident brown-gray spots on the buttock and both thighs. It was learned from the story that her complaints started as small, numerous bumps without tips in her buttock about 8 months ago, that these bumps were also occured for the first time on her nipple 2 days ago, then they were healed with hyperpigmentation and she did not receive any medical treatment in the past. There was no history of trauma, infection, or insect bites before.

Complete blood count, erythrocyte sedimentation rate, liver and kidney function tests, ANA and Anti-DNA were within normal limits in laboratory tests. The Borrelia Burgdorfery serology was negative (-)

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

Histopathological examination of atrophic pigmented plaques showed that thickening of the basal

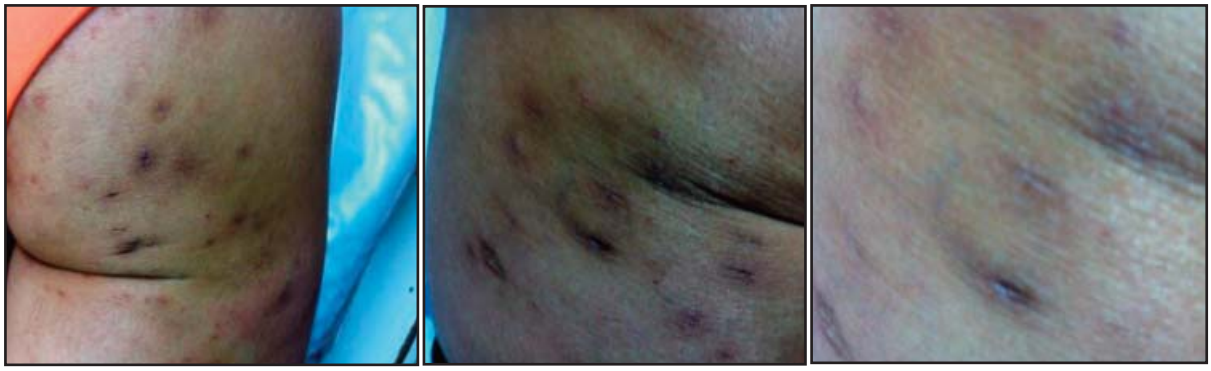

Figure 1. Atrophic plaques in buttock, and upper legs

layer and increased melanin pigmentation, flattening of dermal papillae and mild perivascular lymphocytic infiltration.(**Figure 2**)

The patient was diagnosed with 'Linear Atrophoderma of Moulin' consideration with clinical and pathological findings. We also think that it is a inflammatory type of LAM which initiated with papules. Local corticosteroid cream therapy was initiated for the atrophic plaques on the buttock and thighs with 100 mg doxycycline/daily for the cause of inflammatory papules. There was no regression in the papules at the end of two weeks of antibiotic treatment. Corticosteroid cream th erapy was stopped at the end of two months due to no change.

Discussion

The Atrophoderma of Pasini and Pierini (APP) was first described by Pasini in 1923 [**4**]. Pierini and Vivoli suggested that in 1936 it may be associated with morphea, Yokoyama and colleagues suggested that the atrophy is different from the classical morphea of skin glycosaminoglycans [**5**]. Sharply and well shaped (cliff-drop), brown-gray depressed plaques are typical for atrophoderma [**6**]. Lesions are symmetrically located in the body, usually in the back and lumbosacral region [**1**]. Hand, foot and upper extremity involvement is very rare [**1**]. In some patients, zosteriform distribution was reported in parallel to the skin folds. It is thought that the general course of the disease lasts for as long as 10-20 years and there is no involution [**1**].

The etiopathogenesis is not known [**5**]. Previous studies have reported that genetic, 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 accumulation of IgM and C3 in small blood vessels and the detection of T lymphocytes and macrophages between the perivascular region and collagen fibers in the electron microscope [**7**].

It is thought that the disease does not have a current effective treatment. Antibiotic therapy has been recommended in cases of Borrelia antibody titer positive [**8,9**]. Treatment with systemic steroids, antimalarials, calcitriol and phototherapy have been tried in sclerotic cases [**10**]. In current treatment approaches, Q-switched lasers for hyperpigmentation treatment are used in patients with cosmetic concern [**2,3**].

Histopathology is usually not diagnostic [**5**]. Early lesions have moderate chronic inflammatory cells. This finding is not seen in the late period. Pigment enhancement can be seen in the basal layer [**5**]. Skin attachments are usually protected.

Linear atrophoderma is a rare dermatosis first reported by Moulin in 1992 in 5 patients [**3**]. It is an acquired unilateral hyperpigmented atrophic patches following the

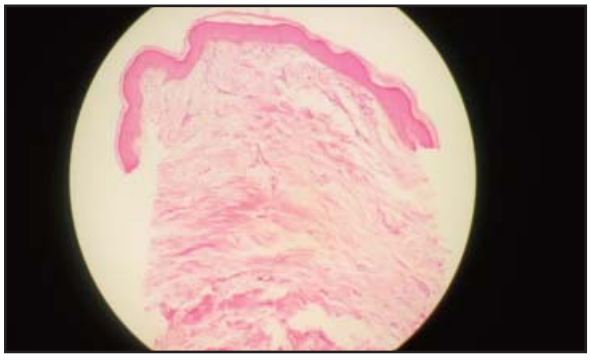

Figure 2. Thickening in the basal layer and flattening in the dermal papilla (HE.x40)

lines of Blaschko. The disease affects children or adolescents.

There are many clinical and histologic similarities between LAM and APP. APP may also resemble LAM but it does not follow Blashko's lines. LAM was suggested a var iant of APP by some authors. [**3**] They co nsidered LAM as a variant of progressive idiopathic atrophoderma of Pasini and Pierini.

Based on clinical and histopathological findings in the present case, it is thought to be compatible with Linear atrophoderma of Moulin. The initial complaints of our case are inflammatory features such as papules which are not compatible with the classical at LAM clinic. For this reason, inflammatory lesions such as dermatitis cruris pustulosa et artophicans and keratosis folicularis decalvans, and other atrophic diseases are considered to be distinctive [**11,12**]. However, the lesions in dermatitis cruris pustulosa are usually found in the form of pustules and in the lower extremity region, in the keratosis folicularis decalvans, as clinical findings that start with more erythematous papules and result in hairy deep cicatricial alopecia [**11,12**]. The other two diseases were excluded with clinical and histopathological findings.

In the differential diagnosis, two Moulin's atrophodema (LAM) cases were encountered, starting with inflammatory papules defined by Browne and Fisher [**10**]. The authors suggested that this disease has inflammatory and non-inflammatory variants. Pasini and Pierini stated that there may be variants of the same disease due to the clinical and histopathological similarity of LAM with atrophoderma [**9**]. In another case report of Moulin's lineer atrophy, pigmentation and atrophy with 20 mg / week methotrexate were reported to decrease successfully for 6 months [**9**]. Because of the efficacy of methotrexate, LAM, APP, and linear scleroderma are thought to be other variants of the same disease [**9**]. There are hyperpigmented atrophic plaques in the LAM, just as in APP, which are usually located on the trunk and extremities. But these plaques are parallel to the Blaschko lines in LAM. The age of onset is between 5 and 20 [**9**]. As in the case, which started in the literature as inflammatory papules, but developed by atrophic plaques following Blaschko lines, we think of LAM in our patient which first started with inflammation. Some may think that this is atypical variant of LAM, but Browne et al reported a case with a ntecedent inflammation and they suggested that there are 2 variants of LAM, an inflammatory and non-inflammatory, or that an antecedent inflammatory phase ultimately may evolve into hyperpigmentation with atrophy [**13,14**]. Later, Utikal et al. referred 2 patients with prominent teleangiectatic erythema within the lesions of linear atrophoderma and argued that these cases may represent a novel variety of LAM or a separate entity [**13,15**]. While LAM usually starts in childhood or adolescence, in 2 cases developed in subjects over 30 years [**16,17**]. Even though Moulin et al. presented unilateral localizations, in the literature there are 5 reports of bilateral LAM [**14, 15, 18,19, 20, 21**]. Furthermore, in most of the patients LAM occurred in the trunk and limbs .

We report this case of LAM because of few cases of inflammatory type in the literature

References

1. Arif T, Adil M, Amin SS, Ahmed M. Atrophoderma of Pasini and Pierini in a blaschkoid pattern. J Dtsch Dermatol Ges 2017; 15: 663-664. PMID: 28346752

2. Zhang RZ, Zhu WY. Two uncommon cases of idiopathic atrophoderma of pasini and pierini: multiple and giant. Indian J Dermatol Venereol Leprol 2011; 77: 402. PMID: 21508593

3. Tukenmez Demirci G, Altunay IK, Mertoglu E, Kucukunal A, Sakız D. Linear atrophoderma of Moulin on the neck. J Dermatol Case Rep 2011; 5: 47-49. PMID: 22187579

4. Avancini J, Valente NY, Romiti R. Generalized lenticuler atrophoderma of Pasini and Pierini. Pediatr Dermatol 2015; 32: 389-391. PMID: 25234089

5. Garg A, Kumar P. Atrophoderma of Pasini and Pierini. Indian Dermatol Online J 2011; 2: 126-128. PMID: 23130246

6. Ravic-Nicolic A, Djurdjevic P, Mitrovic S, Milicic V, Petrovic D. Atrophoderma of Pasini and Pierini associated with extramedullary plasmacytoma. Clin Exp Dermatol 2016; 41: 837-839. PMID: 27443586

7. Yan W, Wang S, Liu HJ, et al. Linear atrophoderma of Moulin:a disease related to immunity or a kind of connective tissue disease? Australas J Dermatol 2017; 58: e126-e128. PMID: 27283080

8. Buechner SA, Rufli T. Atrophoderma of Pasini and Pierini. Clinical and histopathologic findings and antibodies to Borrelia burgdorferi in thirty-four patients. J Am Acad Dermatol 1994; 30: 441-446. PMID: 8113457

9. Tan SK, Tay YK. Linear atrophoderma of Moulin. JAAD Case Rep 2016; 2: 10-12. PMID: 27051814

10. Zahedi Niaki O, Sissons W, Nguyen VH, Zargham R, Jafarian F. Linear atrophoderma of Moulin:underrecognized entity. Pediatr Rheumatol Online J 2015; 13: 39. PMID: 26438123

11. Bens G, Franck F, Diatto G, Preney L, Darie H, Geniaux M. Dermatitis cruris pustulosa et atrophicans a frequent but poorly understood tropical skin condition- case report from Burkina Faso. Int J Dermatol 2008; 47 :473-475. PMID: 18412864

12. Verma R, Bhatnagar A, Vasudevan B, Kumar S. Keratosis follicularis spinulosa decalvans. Indian J Dermatol Venereol Leprol 2016; 82: 214-216. PMID: 26765129

13. Cecchi R, Bartoli L, Brunetti L,Pavesi M. Linear atrophoderma of Moulin localized to the neck. Dermatol Online J 2008; 14: 12. PMID: 18713593

14. Browne C, Fisher BK. Atrophoderma of moulin with preceding inflammation. Int J Dermatol 2000; 39: 850-852. PMID: 11123448

15. Utikal J, Keil D, Klemke CD, Bayerl C, Goerdt S. Predominant telangiectatic erythema in linear atrophoderma of Moulin: novel variant or seperate entity? Dermatology 2003; 207: 310-315. PMID: 14571076

16. Danarti R, Bittar M, Happle R, König A. Linear atrophoderma of Moulin: postulation of mosaicism for a predisposing gene. J Am Acad Dermatol 2003; 49: 492-498. PMID: 12963915

17. Zampetti A, Antuzzi D, Caldarola G. et al. Linear atrophoderma of Moulin. Eur J Dermatol 2008; 18: 79-80. PMID: 18086596

18. De Golian E, Echols K, Pearl H, Davis L. Linear Atrophoderma of Moulin: A distinct entity? Pediatr Dermatol 2014; 313: 373-377. PMID: 23046463

19. Oiso N, Kimura M, Itoh T, Kawada A. Variant of linear atrophoderma of Moulin: Hyper- and hypopigmented linear atrophoderma with aberrant area cutanea and lentiginosis following the lines of Blaschko. J Dermatol 2012; 39: 1097-1099. PMID: 22803683

20. Özkaya E, Yazganoğlu KD. Lentiginosis within plaques of linear atrophoderma of Moulin: a twin-spotting phenomenon? Br J Dermatol 2010; 163: 1138-1140. PMID: 20649796

21. Baumann L, Happle R, Plewing G,Schirren CG. Atrophodermia linearis Moulin.A new disease picture, following the Blasschko lines. Hautarzt 1994; 45: 231-236. PMID: 8014049