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Editorial

Linear atrophoderma of Moulin: Do we need to redefine this controversial disease entity?

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Linear atrophoderma of Moulin (LAM) is a rare dermatologic disorder characterized by hyperpigmented depressed plaques following the lines of Blaschko.[1,2] It was first reported by Moulin et al. in 1992. They reported five healthy young patients who were having unilateral hyperpigmented depressed plaques along Blaschko's lines on the trunk and extremities without any evidence of long-term progression. These plaques were not associated with inflammation, induration, or sclerosis.[2,3] Hyperpigmentation of basal layer with unremarkable changes in epidermal and dermal layers including collagen and elastin, was the characteristic histopathological finding reported in LAM.[2-4] The term "blaschkose" was suggested by Moulin et al. in contrast to the term "blaschkitis" implying the non-inflammatory nature of the disease.[2] Bauman et al. in 1994 reported a patient with a similar disease and suggested the term LAM. The original diagnostic criteria for LAM have been mentioned in **Table 1**.[1,5]

Since its first description by Moulin et al., several authors throughout the world have reported cases with variable clinical and histopathological features such as preceding inflammation, telangiectasias, collagen sclerosis and psoriasiform changes not complying with the diagnostic criteria of LAM and yet have been published under the umbrella term LAM. Hence, some authors proposed atypical variants of LAM while others presumed that different disease entities were identified as LAM.[5-7] Whether all those cases represented classical LAM as described by Moulin et al. or they represented a related dermatosis; depends on whether we confine the diagnosis of LAM as described by Moulin et al. or consider it to be a diseases spectrum where lesions may have varied clinical and histopathological findings. Thus, it is clear that the diagnostic criteria of LAM do not stand at present in view of contrasting clinical and histopathological findings labelled as LAM and demand a meticulous update for the same. In the following text the author will explain why the existing criteria of LAM are neither sufficient nor accurate by making assessment of each criterion and comparing it with the clinically published cases of LAM:

1. Onset during childhood or adolescence: Ang et al. have described a congenital case of linear atrophoderma in a 9 month old Hispanic girl that had been present on her right leg since birth.[3] On the contrary Zampetti et al. described a 42-year-old Caucasian woman who presented with a 5-year history of atrophic brown macules on the left arm and trunk.[8] Similarly, Danarti et al. reported 4 cases of LAM in 2003, one of them was aged 38 years.[9] Thus limiting the onset of LAM to childhood or adolescence is not

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Table 1 Diagnostic criteria for LAM.

1	Onset during childhood or adolescence
2	Development of hyperpigmented, slightly atrophic, unilateral lesions following Blaschko's lines on the trunk or limbs
3	Absence of preceding inflammation and absence of subsequent induration or scleroderma
4	Stable, nonprogressive clinical course without a pattern of remission
5	Histologic findings showing hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibres

justified in view of the above mentioned reported cases.

2. Development of hyperpigmented, slightly atrophic, unilateral lesions following Blaschko's lines on the trunk or limbs: There are three parameters in this criterion viz, hyperpigmented lesions, unilateral localisation and presence of lesions on trunk or limbs. Researchers have described case reports where not only hyperpigmented but also hypopigmented plaques were observed in LAM in association with lentiginosis.[10-12] Contrary to Moulin et al. description of unilateral localization of lesions in LAM, the author could find five cases where lesions were present bilaterally.[7,10,11,13,14] Cecchi et al. and Tukenmez et al. have separately reported cases of LAM localised to neck.[6,15] It seems logical that these three restricting parameters of criterion 2 are not holding true for diagnosing LAM and hence should be reassessed.

3. Absence of preceding inflammation and absence of subsequent induration or scleroderma: Moulin et al. had suggested that there should be absence of preceding inflammation for diagnosing LAM.[2] However, Brown and Fisher have reported a case with prior inflammation. They suggested that LAM has two variants viz., inflammatory and non-inflammatory types. They proposed that the initial inflammatory phase ultimately might lead to hyperpigmentation with atrophy.[14]

4. Histological findings showing hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibres: In the original case series by Moulin et al., they reported only basal layer hyperpigmentation; however the rest of the epidermis and dermis was unremarkable.[2] Subsequently, authors have reported new histopathological findings in LAM that include epidermal atrophy, perivascular lymphocytic infiltrates, altered dermal collagen, acanthosis, decreased elastic tissue, dilated dermal blood vessels and plasma cell infiltrate.[1,13] In fact, a perivascular lymphocytic inflammatory infiltrate in the superficial dermis combined with abnormal collagen fibres have been the most common histopathological findings reported in LAM.[5] Since these cases with new histopathological findings presented sufficient clinical evidence to support a diagnosis of LAM, these authors suggested that LAM was not as limited in scope as has been described originally by Moulin et al. but probably comprised a wider set of clinical and histopathological features that fit rather within a spectrum of related disorders.[13]

From the aforementioned facts, it is evident that four out of five criteria of LAM are not working currently for diagnosing LAM. This implies that either these cases which are not complying with the original diagnostic criteria of LAM are referring to a different dermatosis; or they represent atypical or novel varieties of LAM; or we need to redefine this controversial entity and reframe its diagnostic criteria with a wider clinical and histopathological spectrum to account for all these clinical and histopathological differences in cases of LAM. The author is of the opinion that LAM is not limited in scope in its clinical or

histopathological findings as was proposed earlier but rather encompasses a disease spectrum where there are varied clinical and histopathological features. Hence there exists a need to redefine LAM and its diagnostic criteria should be updated in order to prevent any confusion in diagnosing and treating this rare disease entity.

References

1. Arif T, Adil M, Sami M, Saeed N. Hyperpigmented patches following Blaschko's lines: Two probable cases of linear atrophoderma of Moulin with subtle atrophy and a critical review of the subject. Indian J Paediatr Dermatol 2019 [Ahead of print].
2. Moulin G, Hill MP, Guillaud V et al. Acquired atrophic pigmented band-like lesions following Blaschko's lines. Ann Dermatol Venereol 1992; 119: 729–36.
3. Ang G, Hyde PM, Lee JB. Unilateral congenital linear atrophoderma of the leg. Pediatr Dermatol 2005; 22: 350–4.
4. Ang GC, Lee JB. Linear atrophoderma of Moulin: is it a single disease? J Am Acad Dermatol 2005; 52: 923–924; author reply 924-5.
5. Lopez N, Gallardo MA, Mendiola M, Bosch R, Herrera E. A case of linear atrophoderma of Moulin. Actas Dermosifiliogr 2008; 99:165-7.
6. Cecchi R, Bartoli L, Brunetti L et al. Linear atrophoderma of Moulin localized to the neck. Dermatol Online J 2008;14:12.
7. Utikal J, Keil D, Klemke CD, et al: Predominant telangiectatic erythema in linear atrophoderma of Moulin: novel variant or separate entity? Dermatology 2003; 207: 310–15.
8. Zampetti A, Antuzzi D, Caldarola G, et al: Linear atrophoderma of Moulin. Eur J Dermatol 2008; 18: 79–80.
9. Danarti R, Bittar M, Happle R, et al: Linear atrophoderma of Moulin: postulation of mosaicism for a predisposing gene. J Am Acad Dermatol 2003; 49: 492–8.
10. 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: 1097–9.
11. Ozkaya E, Yazganoglu KD: Lentiginosis within plaques of linear atrophoderma of Moulin: a twin-spotting phenomenon? Br J Dermatol 2010; 163: 1138–40.
12. Yücel S, Özcan D and Seçkin D. Lentiginosis within plaques of hypopigmented linear atrophoderma of Moulin: another example of twin spotting? Int J Dermatol 2013; 52: 1427-9.
13. Golian ED, Echols K, Pearl H, and Davis L. Linear Atrophoderma of Moulin: A Distinct Entity? Pediatric Dermatol 2014; 31: 373–7.
14. Browne C, Fisher BK: Atrophoderma of Moulin with preceding inflammation. Int J Dermatol 2000; 39: 850–2.
15. Tukenmez Demirci G, Altunay IK, MertogluE et al: Linear atrophoderma of Moulin on the neck. J Dermatol Case Rep 2011; 5: 47–9.