Hong Kong J. Dermatol. Venereol. (2016) **24**, 197-200

Case Report

A rare case of unilateral linear lichen planus pigmentosus

單側線狀色素性扁平苔癬的罕見病例

N Dizen-Namdar, E Kural-Unuvar, MH Metineren, C Kocak

Lichen planus pigmentosus (LPP) is a rare clinical variant of the lichen planus. It presents with pigmented, dark brown macular lesions on the flexural or sun exposed regions. Linear LPP is presented with hyperpigmented lesions along the lines of Blaschko. To the best of our knowledge, there are a few cases of linear LPP which have been reported in the literature. In our case report, we present a female Turkish patient with unilateral linear LPP that manifested as asymptomatic brown macular lesions, distributed along the lines of Blaschko between the middle part of the left gluteal region and the left calf.

色素性扁平苔藓是扁平苔藓的一種罕見臨床變異型，呈現爲在皺摺位或日光暴露位置的色素性（暗棕黃色）斑狀皮損。而線狀色素性扁平苔癬則呈現沿著布拉許口氏線的色素沉著病變。據我們所知，文獻曾報導過少數線狀色素性扁平苔癬的病例。在我們的病例報告中，我們提出一個女性土耳其患者的單側線狀色素性扁平苔癬，表現爲無症狀的棕色斑狀皮損，沿著布拉許口氏線分佈由左邊臀部中間直至左小腿。

Keywords: Blaschko lines, lichen planus, lichen planus pigmentosus, linear pattern

關鍵詞：布拉許口氏線、扁平苔藓、色素性扁平苔藓、線狀形態

Department of Dermatology, School of Medicine, Dumlupinar University, Kutahya,Turkey
N Dizen-Namdar, Assist. Prof.

Dermatology Clinic, Central Hospital, Istanbul, Turkey
E Kural-Unuvar, MD

Department of Pathology, School of Medicine, Dumlupinar University, Kutahya, Turkey
MH Metineren, Assist. Prof.
C Kocak, Assist. Prof.

Correspondence to: Asisst. Prof. N Dizen-Namdar

Dumlupinar University, School of Medicine, Department of Dermatology, Kutahya-Turkey

Introduction

Lichen planus pigmentosus (LPP) is described as a condition of unknown aetiology, characterised by mottled or reticulated pigmented, dark brown macular lesions on the flexural or sun-exposed regions. The characteristic histopathological findings are hyperkeratosis, atrophic epidermis with vacuolar alteration of the basal cell layer, and scarce lymphohistiocytic or lichenoid infiltrates in the dermis with pigmentary incontinence and the presence of melanophages.[1,2] Linear LPP presents with hyperpigmented lesions along the lines of Blaschko. Until now there are only a few

cases of linear LPP which have been reported.[3-5] In this case report, we present a female Turkish patient with unilateral linear LPP.

Case report

A 33-year-old female Turkish patient presented with asymptomatic dark brown macules which appeared on the left gluteal region and spread gradually to the calf for one year. There was no history of drug intake, systemic illness, prolonged sun exposure or trauma at the lesional sites.

Physical examination showed dark brown macules in linear streak-like distribution related to the Blaschko lines, extending from the left gluteal region to the calf (Figure 1). There was no scalp, nail or mucosal involvement. Laboratory investigations revealed normal full blood count and liver function tests. The serology for hepatitis C was negative.

The histological examination showed orthokeratosis, epidermal atrophy, vacuolar alteration of the basal layer, lichenoid lymphocytic infiltration with pigmentary incontinence and melanophages in the superficial dermis (Figure 2a). Immunohistochemical examination revealed that the lymphocytic infiltrate consisted of CD8 (+) T cells (Figure 2b), with an increase in the number of CD1a (+) Langerhans cells (Figure 2c). These findings were consistent with unilateral linear LPP. Topical mometasone furoate cream twice daily was given. However, there was no improvement after one month.

Discussion

Lichen planus pigmentosus was first described in 1974 by Bhutani et al.[1] It is clinically different from classical lichen planus by exhibiting the dark brown macules and/or papules and longer clinical course without pruritus or scalp, nail or mucosal

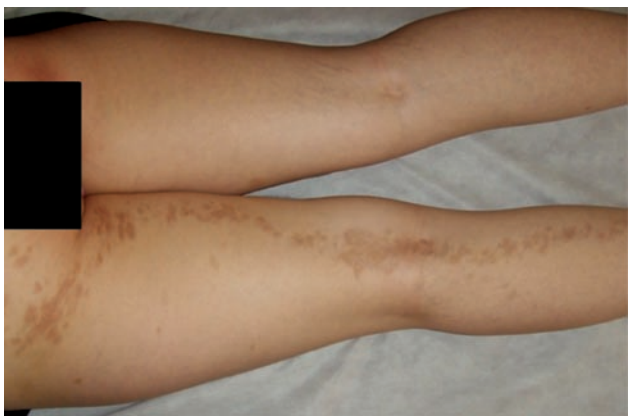

Figure 1. Linear streak of the dark brown macules from the left gluteal region to the calf related to the Blaschko lines.

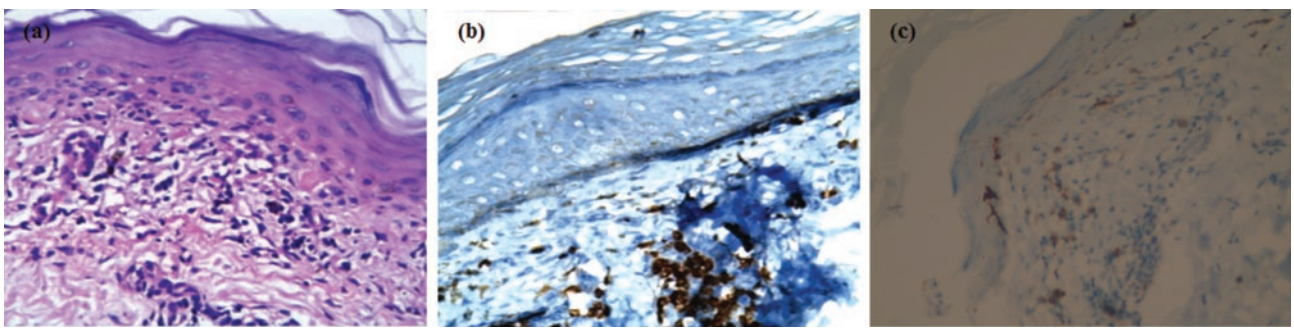

Figure 2. Light and immunohistochemical microphotographs. (a) Epidermal atrophy, vacuolar alteration of the basal cell layer and lichenoid lymphocytic infiltration with pigmentary incontinence, melanophages in the superficial dermis. (b) Lymphocytic infiltrate is composed of CD8 (+) T cells in the superficial dermis. (c) CD1a (+) Langerhans cells in the superficial dermis (original magnifications: a-c, x 100).

involvement. LPP is most commonly found on the sun-exposed areas such as the face, neck, and flexural folds including the axilla, inguinal and submammary regions.[2] Some authors have described LPP-inversus which mostly affects the intertriginous regions.[6,7] Less common presentations including a zosteriform pattern on the trunk and involvement of the non-sun-exposed areas such as the thigh have been reported.[8,9] There are a few linear LPP cases that have been reported in the literature.[3-5] LPP presenting with a linear pattern was first reported by Hong et al.[3] Our patient had lesions in a linear pattern on the left lower extremity. The linearity of the lesions is probably related to the Blaschko's lines, which suggests that the predisposition to develop LPP might be determined during embryogenesis.[3] The histological findings of LPP show atrophic epidermis with vacuolar alteration of the basal cell layer, and sparse lymphohistiocytic or lichenoid infiltrates in the dermis with pigmentary incontinence and the presence of melanophages.[1,2] It is thought that the cytotoxic action of the T lymphocytes against the keratinocytes of basal layer plays a role in the molecular pathogenesis of lichen planus.[7] In the immunohistochemical study, destruction of keratinocytes and CD8 (+) T cell infiltration were observed.

The number of Langerhans cells in lichen planus varies and may play a role in the pathogenesis of the process.[10] An increase in CD 1a (+) Langerhans cells in the superficial dermis was revealed in both our case and in the literature.[6,7]

According to these findings, Langerhans cell-mediated immune responses along with cytotoxic cell injury a significant role in LPP pathogenesis. The differential diagnoses of our case include lichen striatus, linear and whorled naevoid melanosis, linear ashy dermatosis and inflammatory linear verrucous epidermal naevus (ILVEN). Lichen striatus is charactarised by preceding inflammatory papules or a scaly eruption that last for four months to four years and a perivascular and periadnexal inflammatory cell infiltration.[4] Linear and whorled naevoid hypermelanosis which is related to congenital anomalies generally appears within a few weeks after birth. Basal pigmentation without pigment incontinence is found in the histology.[4] Vega et al. reported clinical differences in the colour of the lesions of ashy dermatosis and LPP, the presence of an active red border, and the presence or absence of pruritus.[2]

ILVEN which begins during early childhood is persistent with pruritus as a foremost symptom and generally presents as a pruritic, erythematous scaly lesion following Blaschko's lines.[11,12] Histologically, there are columns of hypergranulosis with orthokeratotic hyperkeratosis and equally well-defined columns of agranulosis and parakeratotic hyperkeratosis.[11,12]

Based on the history, the onset age, the appearance and pathological features of the lesions, the patient was diagnosed with linear LPP. The clinical course of LPP is variable: in some cases the lesions disappear spontaneously, although some cases persist for years. There is no evidence-based treatment of LPP. Topical calcineurin inhibitors and topical steroids are commonly used as a treatment but clinical efficacy may be suboptimal. The treatment efficacy is better for localised cases.[5,7]

In conclusion, LPP can present with lesions in a linear pattern. Therefore it should be considered in the differential diagnosis of linear hyperpigmented skin lesions.

References

1. Bhutani LK, Bedi TR, Pandhi RK, Nayak NC. Lichen planus pigmentosus. Dermatologica 1974;149:43-50.
2. Vega ME, Waxtein L, Arenas R, Hojyo T, Dominguez-Soto L. Ashy dermatosis and lichen planus pigmentosus: a clinicopathologic study of 31 cases. Int J Dermatol 1992;31:90-4.
3. Hong S, Shin JH, Kang HY. Two cases of lichen planus

pigmentosus presenting with a linear pattern. J Korean Med Sci 2004;19:152-4.
4. Seo JK, Lee HJ, Lee D, Choi JH, Sung HS. A case of linear lichen planus pigmentosus. Ann Dermatol 2010;22:323-5.
5. Akarsu S, Ilknur T, Ozer E, Fetil E. Lichen planus pigmentosus distributed along the lines of Blaschko. Int J Dermatol 2013;52:253-4.
6. Kashima A, Tajiri A, Yamashita A, Asada Y, Setoyama M. Two Japanese cases of lichen planus pigmentosus-inversus. Int J Dermatol 2007;46: 740-2.
7. Uyar B, Sivrikoz O. A case of lichen planus pigmentosus-inversus. Turkderm - Arch Turk Dermatol Venereol 2012;46:160-2.
8. Cho S, Whang KK. Lichen planus pigmentosus presenting in zosteriform pattern. J Dermatol 1997; 24:193-7.
9. Kim KJ, Bae GY, Choi JH, Sung KJ, Moon KC, Koh JK. A case of localized lichen planus pigmentosus on the thigh. J Dermatol 2002;29:242-3.
10. Gueiros LA, Gondak R, Jorge Junior J, Coletta RD, Carvalho Ade A, Leao JA, et al. Increased number of Langerhans cells in oral lichen planus and oral lichenoid lesions. Oral Surg Oral Med Oral Pathol Oral Radiol 2012;113:661-6.
11. Zhang RZ, Zhu WY. One case of unilateral linear lichen planus pigmentosus. Open Dermatol J 2012; 6:25-8.
12. Gon Ados S, Minelli L, Franzon PG. Case for diagnosis. Inflammatory linear verrucous epidermal nevus. An Bras Dermatol 2010;85:729-31.