LICHEN PLANUS PIGMENTOSUS WITH BLASCHKOID DISTRIBUTION- CASE STUDY

MĂDĂLINA MOCANU*, MĂDĂLINA HÂNCU*, BIANCA LOREDANA MARINESCU *, DANIELA ROTARU*, TATIANA ȚĂRANU*

Summary

Introduction

Linear Lichen Planus Pigmentosus (LPP) is a rare form of lichen planus characterized by macular or papulous, hyperpigmented, dark brown lesions, in plaque or cross-linked, asymptomatic with blaschkoid localization.

Case Study

We present the case of a 50-year-old patient, diagnosed with lichen planopilaris, three months ago, who addressed our clinic for the appearance of new, discreetly atrophic maculo-pigmentary lesions with blaschkoid distribution in the hemicranium and left hemifacial area, as well as because of the persistence of scalp lesions. The illness started at the same time with a dental treatment on the upper left dental arcade. The patient followed systemic antihistamine and topical corticosteroid treatment without significant clinical effects. The histopathological examination reveals hyperkeratosis, irregular hypergranulosis acanthosis, hyaline corpusculary diskeratosis, and basal strain remodelling. Histopathological aspects plead for the diagnosis of LPP.

Discussion

LPP is described as an unknown aetiology disorder, different from the classical lichen planus, through lesion morphology and the absence of subjective symptomatology. A particular feature of the disease is the lack of interest in the scalp, nails and mucous membranes. The disease occurs frequently in the middle age, with a peak incidence in decade 3-4 of life. The case presented shows a particular image of LPP associating blaschkoid distribution lesions, rare clinical variant, with unknown prevalence.

Conclusions

The particularity of this case is the blaschkoid distribution of typical lesions of lichen planopilaris and subsequent pigmentary macules in the left hemifacial and left submental area. The traumatic etiopathogenesis of lichen planopilaris is a relatively rare occurrence in current clinical practice.Linear LPP does not have a standardized treatment due to the rarity of cases, but in the case of our patient, the combination of Elidel 1% cream with Regen sil resulted in a mild clinical improvement after 6 weeks of treatment.

Key words: *linear lichen planus, pigmentosus lichen planus, endobucal trauma.*

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Introduction

Lichen planus pigmentosus is a rare lichen planus pattern characterized by dark brown, macular or papillary lesions, hyper-pigmented, spotted or cross-linked, located in the areas exposed to sunlight and flexion folds [1, 2].

The clinical-anatomical form of linear lichen planus pigmentosus is considered a rare entity with unknown prevalence combining the features of the two variants.

The present paper is a case report of a lichen planus with papulo-keratoses lesions of the scalp

* University Clinical Hospital C.F. Iași, Dermatology-venerology Clinic.

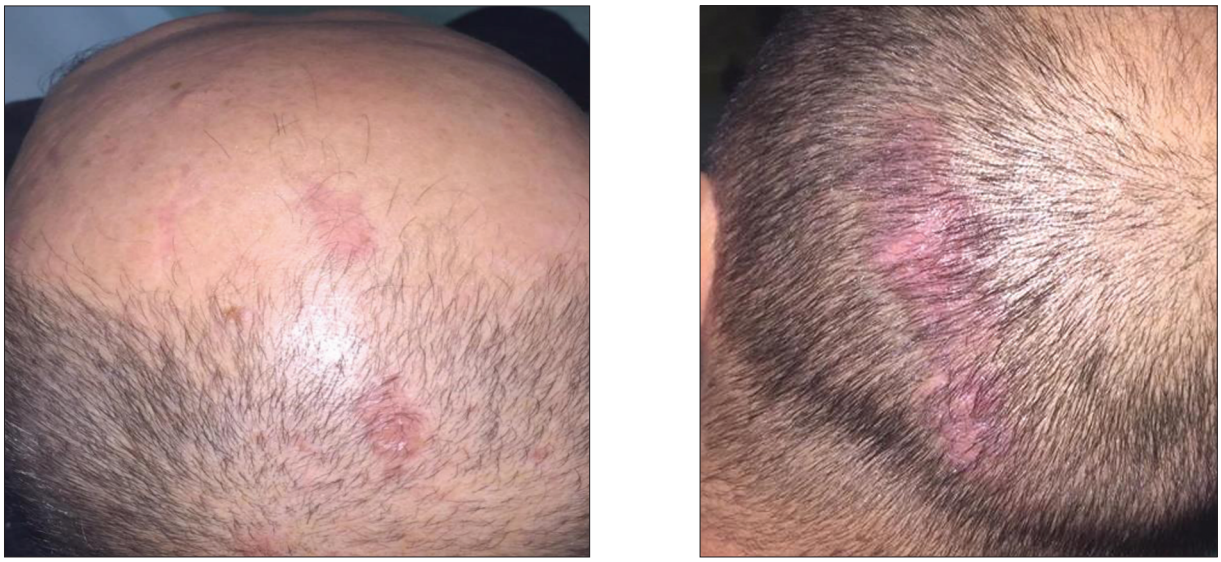

Fig. 1. Papulo-keratosis and atrophic-scar tissue lesions in the scalp in the left parietal-occipital region

characteristic to the lichen planopilaris and macular pigmentary lesions, having a blaschkoid distribution in the hemicranium and left hemifacies. The particularity of this case is the onset on scalp coincidental with left endobuccal maxillary trauma and subsequent appearance of pigment macules in the left hemifacies.

Clinical case

A 50-year-old patient from the urban environment initially diagnosed with lichen planus of the scalp 3 months ago, is addressing to our clinic for the appearance of new macular-pigmentary, discreetly atrophic lesions on the left cheek, chin, under the chin and submandibular left, as well as because of the persistence of scalp lesions.

The history did not releave any significant pathological or heredo-collateral personal history and the general physical examination releaved data within physiological limits. Biochemical, immunological and virological explorations were within the normal range.

The patient reports on the onset of skin scalp condition about 8 months ago, which coincided with a dental treatment on the upper left arch. We mention an incident during the dental manevers, which resulted in the breaking of the canal needle in the context of pulpectomy. The needle was later extirpated by means of a surgical procedure.

The hyperkeratotic and pruritic skin lesions originally occurred at the level of the scalp, in the left parietal-occipital region, being observed after about 4 weeks of dental surgery (Fig. 1). At the indication of a dermatologist, he followed a local treatment with Asorian solution for external use, Clobesol cream and oral antihistamine therapy. He cannot appreciate the benefit of treatment and finds the occurrence of new lesions, pigmentary, asymptomatic, homolateral, on the left cheek, chin and under the chin (Fig. 2).

Later, due to the lack of response to the treatment, a scalp biopsy was performed that concluded on the diagnosis of lichen planus. Local treatment with Cicaderm and Protopic 0.1% was initiated for one month, resulting in a slight improvement of the lesions without complete resolution, which is why he addresses our clinic for a new evaluation. The general physical examination revealed a patient in apparent health status, skin type III, and paraclinical investigations (haematological, biochemical, immunological tests for viral hepatitis) resulted in normal results.

Repetition of a biopsy of a tissue fragment from a left macular facial pigmentary lesion was performed (Fig. 3). The histopathological examination revealed hyperkeratosis, unregulated

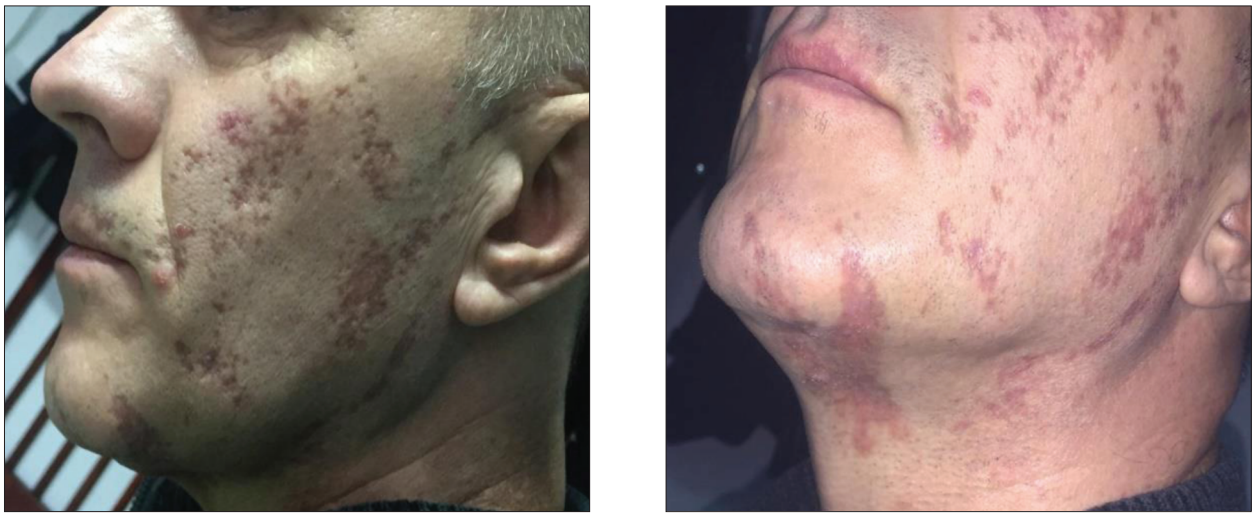

Fig. 2. Discrete atrophic pigmentary lesions with blaschkoid distribution in the left hemifacies and left under chin

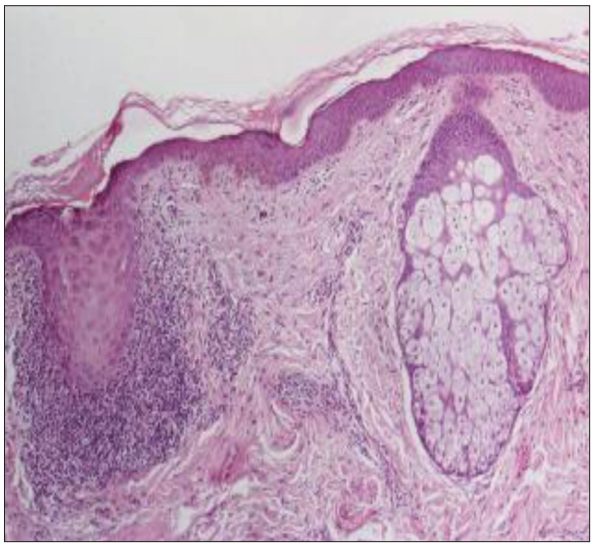

Fig. 3. Overview. H.E. x 40

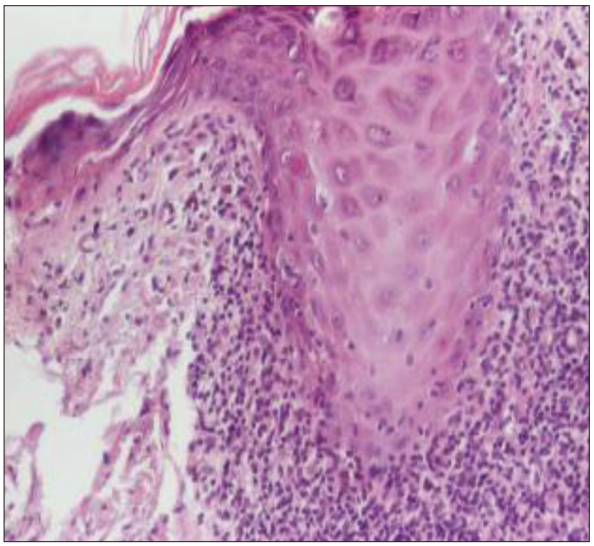

Fig. 4. Hyperkeratosis, irregular acanthosis, basal layer remodelling. H.E. x 100

acanthosis with hypergranulosis, and hyaline corpusculum-forming diskeratosis accompanied by rare melanophages (Fig. 4). In the superficial dermis, the presence of a dense lymphocytic inflammatory infiltrate disposed parallel to the epidermis, with remodelling of the basal layer can be found (Fig. 5). Histopathological aspects plead for the diagnosis of linear lichen planus pigmentosus.

It was decided to treat him with Elidel 1% cream (2 applications/day) alternately with Regen-sil (2 applications/day). After one month of therapy, there was an insignificant improvement in the cutaneous lesions.

Discussions and conclusions

Lichen planus pigmentosus is a rare variant of lichen planus, of unknown etiology, first described by Bhutani in 1974. It is characterized clinically by dark brown macules and / or papules, located in the sun exposed areas (face, neck) or skin

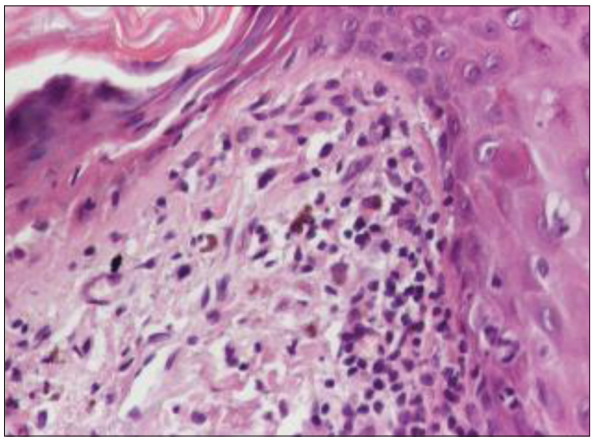

Fig. 5. Melanophoges in the inflammatory lymphocyte infiltrate. H.E. x 200

folds (inguinal, submammary or axillary area) [3]. Clinical manifestations have the particularity of being asymptomatic usually affecting the scalp, nails or mucous membranes [4].

Disease occurs more frequently in females, with a peak incidence in the 3-4 decade of life. Preferential distribution on sun-exposed areas correlates with IV and V skin types explained and with the fact that most studies were performed on subjects in India, the Middle East and South America. The lichen planus pigmentosus of the folds is rare and mainly found in Caucasians [5, 6].

The etiopathogenicity of the lichen planus pigmentosus is incompletely elucidated, but the implication of immunological mechanism similar to those from the classic lichen planus is certain. Lichen planus pigmentosus is considered a type IV hypersensitivity reaction to unknown antigenic stimuli, with dermal inflammation leading to melanocytic and pigmentary incontinence of superficial dermis [7].

The lichen planus is immunologically mediated by activated T lymphocytes, which play a role in the damage to epithelial cells. Epithelial scarring occurs in three stages: interaction of T lymphocytes with antigen, activation of T lymphocytes with lymphokine secretion and basal keratinocyte apoptosis. The inflammatory infiltrate located in the superficial dermis is predominantly formed from CD4 + and CD8 + lymphocytes, characteristic of the lichenoid tissue reaction. Progression of the disease is associated with the predominant accumulation of CD8 + in the inflammatory infiltrate. T-helper activated lymphocytes produce the epidermal activator of thymocytes (ETAF), interleukin 1 (IL-1), IL-2 and α-interferon (INFα), substances that induce keratinocyte activation with cytokine release and proliferation of CD4 + lymphocytes and of T cytotoxic lymphocytes. Lymphotoxicity directed against epidermal basal cells will lead to keratinocyte apoptosis and basal layer liquefaction degeneration [8, 9].

From a clinical point of view, the exposed case associates papulo-keratosis lesions of lichen planopilaris with scarring alopecia at the level of the scalp, with discretely atrophic pigmentary macules with linear distribution. The absence of pruritus has led to the delay in diagnosis and treatment.

Linear lichen planus pigmentosus is rarely reported in the research literature, the blaschikoid distribution suggesting the existence of a genetic predisposition to develop lichen planus [1]. Numerous studies have described a significant association between the class of HLA histocompatibility antigens and the lichen planus pathogenesis. The high frequency of HLA-B27, HLA-B51 and HLA-Bw57 was observed in patients with oral lichen planus, HLA-DR1, HLA-DR9 or HLA-DR6 antigens being observed both in cases of lichen planus with oral mucosal lesions, and in those with only skin damage [8].

The case presented offers a particular image of the lichen planus pigmentosus with linear distribution. The unilateral, linear location of the pigmentary papules on a sun-exposed area (left hemifacies) along with the planopilaris lesions

outlines the picture of a rare association between the lichen planus pigmentosus and the linear one.

We also highlight the onset of scalp lesions that coincided with left endobuccal maxillary trauma and the subsequent appearance of pigment macules in the left hemifacies. Although materials used in dental reconstruction and local trauma have been cited as possible etiologic agents of oral lichen planus, in this case they can be considered trigger factors of cutaneous manifestations, a rare occurrence in current clinical practice.

In spite of the polymorphic clinical forms, the histopathological aspect remains evocative for the diagnosis of lichen planus, the main features being orthokeratosis hyperkeratosis, focal thickening of the granulous layer, irregular acanthosis, liquefaction degeneration of the basal layer and subepidermic lymphatic infiltrate. In addition, in the lichen planus pigmentosus the dermal pigmentary incontinence [10, 11] additionally interferes.

Positive diagnosis is generally based on clinical examination due to the morphology and typical distribution of intense pruritus lesions. The histopathological examination is necessary to confirm clinical suspicion, especially in atypical forms, such as the case of the patient presented.

The lichen planus pigmentosus must be differentiated by erythema discromicum perstans due to the many clinical and histological similarities between the two pathologies: dermal pigmentary incontinence, epidermal atrophy. The differences are distinguished by a more widespread distribution of lesions in erythema discromicum perstans, which do not respect the boundaries of areas exposed to the sun, and hyperpigmentation usually appears late in the course of the disease. Histopathologically, erythema discromicum perstans is characterized by basal vacuole degeneration with dermal mononuclear infiltrate which is not lichenoid. Hyaline corpuscles are typical of lichen planus pigmentosus and occur only occasionally in erythema discromicum perstans. Also, some authors consider deeper pigmentation in the dermis compared to that in the lichen planus pigmentosus.

Other pathologies for differential diagnosis with lichen planus pigmentosus are macular discoid lupus erythematosus, acanthosis nigricans, fixed drug eruptions (biseptol), post-inflammatory pigmentation and dermal focal elastosis.

The treatment of lichen planus pigmentosus is a challenge, as most lesions are not responsive to classical therapy. The treatment principles include topical dermocorticoid therapy or with calcineurin inhibitors in combination with photoprotection. Systemic corticotherapy is also a viable treatment option. Other therapeutic options suggest the use of substances such as dimethylsulfoxide, griseofulvin, chloroquine, vitamin A, but also laser therapy for hyperpigmentary lesions in lichen planus.

In conclusion, lichen planus pigmentosus may be presented in a linear pattern and therefore it is necessary to include it in the differential diagnosis of linear skin hyper-pigmentation.

The particularity of this case is the blaschkoid distribution of typical lesions of lichen planus pigmentosus and subsequent pigmentary macules in the left hemifacies and left area under the chin. The onset coinciding with an endobucal trauma attests to the possible involvement of the proximity trauma in the etiopathogenicity of the case, a relatively rare occurrence in current clinical practice.

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Conflict of interest
NONE DECLARED

Correspondance address: Tatiana Țăranu, MD, PhD
University Clinical Hospital C.F. Iași
Garabet Ibrăileanu Street, No 1