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LICHEN PLANUS: A MUCO-CUTANEOUS, NAILS AND SCALP DISORDER – CASES REPORT

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Abstract: *Lichen planus is an autoimmune, itching disorder that involves T lymphocytes, which may interest the skin, mucous membranes and exoskeletons. The presence of lichen manifestations both at the skin, mucous membranes and at the level of the skin attachments (hair and nails) is rarely mentioned in the literature. Although lichen planus may have a self-limiting evolution, early treatment is important in shortening the evolution of the disease and in controlling the symptoms. The therapeutic effects of retinoids (immunomodulatory, anti-inflammatory, anti-proliferative effects) make them an appropriate choice in the treatment of lichen with (cutaneous-mucosal, nail and planopilaris lesions. We will present the cases of 2 patients with muco-cutaneous, nails and scalp disorder due to lichen planus, and the time of presentation to the doctor made the difference between the complete remission of the disease and the evolution towards severe, extended, definitive alopecia in the scalp and genital lichen sclerosus.*

INTRODUCTION

Lichen planus is an autoimmune disorder that can affect the skin, mucous membranes, nails and hair. Lichen planus affects 1-2% of the general population, palmo-plantar, cutaneous-mucosal and nail association being very rare. Mostly, this pathology affects the age groups of 30-60 years, without a gender predilection.

The etiopathogeny of lichen planus is not exactly known. Mediated by immune mechanisms, in the pathogenesis of the disease, the involvement of the T cells is observed with the activation of the cytotoxic lymphocytes, after the recognition of the specific antigens, with the apoptosis of the keratinocytes. Etiologically, regarding the triggering factors, we can mention infections (in particular, HCV - 5x higher prevalence than in the rest of the population, syphilis, HSV2, HPV, HIV, H.pylori etc.), genetic predisposition (HLA B7 have been reported, -Aw19, -B18, -Cw8 in family and non-family cases- HLA-A3, -A5, -A28, -B8, -B16, -Bw35), mental illness (anxiety and depression), medications (IECA, thiazide diuretics), NSAIDs, quinidine, beta-blockers, TNF-alpha inhibitors, smoking (lichen with oral damage), gold salts, mercury, copper (contact allergy to these metals found in dental restorations).(1,2)

Most lichen planus cases start insidiously, usually with symmetrical affectation of the flexion areas of the extremities, radiocarpal joints, dorsal hands, the sacral region, ankles and legs, with possible evolution towards generalization in 2-16 weeks.(3) The abbreviation of 4P is used: *papules, polygonal, purple, pruritus* to summarize the main clinical manifestations of the disease.(2) Another abbreviation is of the 6P: *purple, polygonal, papules, pruritus, plaques, plane.*(1)

Papules are usually grouped into plaques, annularly, linearly or with an actinic pattern and tend towards coalescence, with the presence on the surface of fine, transparent, adherent squamas. The characteristic component of the Wickham strips is a whitish, glossy network placed on the surface, easily visible after applying oil, xylene or water. Less characteristic is the Koebner phenomenon, an isomorphic response induced by scratches, traumas or other injuries on the skin. Gray-brown hyperpigmentation is found in most patients after lesions disappear due to melanin deposits in the superficial dermis.(1,2)

Lichen planus can affect the oral, vaginal, esophageal, nasal, laryngeal, conjunctival, urethral and anal mucosa. At the oral mucosa there are described the following forms: reticular, plaque-type, atrophic, papulose, erosive-ulcerative and bullous. The most common is this reticular form, clinically described as a whitish reticular network "in the fern leaf" pattern, with spontaneous remission in 40% of cases. The reticular form is usually asymptomatic, and in the atrophic and erosive (ulcerative) forms - the burning pain completes the clinical picture. Most frequently, the oral mucosa, tongue, lips, palate and buccal floor are affected, with predominant symmetrical damage. Oral lichen planus may persist for many years, improving and exacerbating, the erosive and atrophic form presenting a risk of malignant transformation into squamous cell carcinoma.(3,4)

Half of the patients with oral lichen planus develop lichen planus of the vulvo-vaginal mucosa. Clinically, erosions, asymptomatic desquamative vaginitis can be noticed or complains of pain, pruritus, burns, leukorrhea that can evolve towards leukoplasty and vulvar intraepithelial neoplasia.(5)

Nail damage is estimated in 10-15% of patients with lichen planus. The matrix and the nail bed are affected by the chronic inflammatory process, leading to an evolutive destruction with the formation of lateral thinning, longitudinal ridging and striations (onychorrhexis), fissuring, distal splitting (onychoschizia), subungual hyperkeratosis, onycholysis,

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trachyonychia, erythematous patches of the lunula up to onychatrophy and pterygium unguis - the most common form in which all nails are sometimes affected, leading to complete nail dystrophy.(6)

Palmoplantar lichen planus is a form rarely described in the literature, with different morphology compared to the classical forms, being difficult to diagnose. The following clinical variants have been described: erythematous scaly form (which is the most common), pitted plaques, ulcerative lesions, vesicle-like papules, umbilicated papules, punctate keratoderma, diffuse keratoderma, perforating palmar LP and petechiae-like lesions, hypertrophic, keratotic plaque. Frequently, the lesions are well delimited, with the possibility of multiple forms in the same patient, located at the level of the internal plantar arch, usually without fingers damage.(7,8)

Laboratory tests do not reveal specific changes. A leukopenia with lymphopenia, dyslipidemia, HVC positive serology or other infections may be encountered. Direct immunofluorescence in the lichen planus shows IgM deposits and complement with the presence of keratinocytes destroyed at the dermal-epidermal junction.(3,9)

Skin biopsy confirms the diagnosis of lichen planus. Histopathologically, epidermis is hyperkeratotic with irregular acanthosis and colloid bodies (degenerative keratinocytes or Civatte bodies) in the epidermis, focal thickening in the granular layer, destruction of the basal layer, linear or shaggy deposits of fibrinogen and fibrin of basemen membrane, "lichenoid" infiltrate of lymphocytes (primarily helper T) and histiocytes with Langerhans cells in the upper dermis.(3,10)

The lichen planus is a self-limited pathology, with the remission of the lesions in 1-2 years. Treatment depends on the location of the lesions, on the severity; the therapeutic target being the shortening of the time of manifestation of the disease and the cutting of the symptomatology, of most interest being the pruritus. The first line of therapy includes class I and II potency dermatocorticosteroids. Local therapy second line includes: UVB, local retinoids, tacrolimus/ pimecrolimus locally.(11) Systemic treatment is performed with retinoids, corticosteroids, metronidazoles, griseofulvin, cyclosporine. Third-line therapy is represented by: PUVA, talomide, mycophenolate mofetil, low molecular weight heparin and iontophoresis. Systemic therapy is reserved for recurrent cases in combination with local corticosteroid therapy or phototherapy.(12)

In the medical literature there is not yet a guide for the treatment of palmo-plantar lichen planus. Systemic therapies with acitretin, cyclosporine, corticosteroids, enoxaparin and topical therapies with dermatocorticoids, retinoic acid, cyclosporine, tacrolimus/ pimecrolimus and various combinations of systemic and/ or local therapies have been shown to be effective.(13,7)

CASES REPORT

Case 1. Female patient, 52 years old, smoker, with a history of depressive disorder, is admitted to the dermatovenerology department for the appearance of a disseminated erythematous-papular rash for approximately 1 month. At the palm-plantar level, she presents areas of hyperkeratosis and marginal erythematous-papular lesions, at the level of the oral mucous membranes - whitish networks with a lacy appearance, and vulvar - whitish plaques with intense cutaneous pruritus.

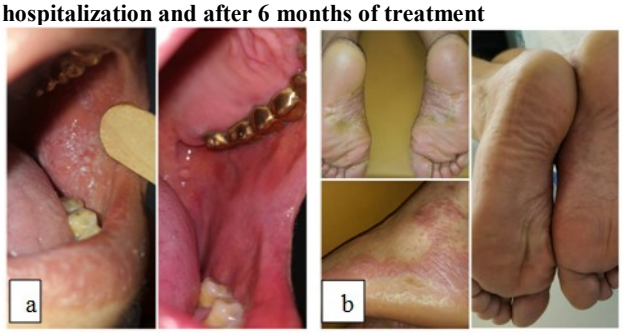
Upon hospitalization, at palmo-plantar level, she shows an erythematous-papulo-squamous rash with lesions coming together in plaques, with areas of hyperkeratosis, accompanied by intense skin pruritus (figure no. 1b). In a disseminated manner, she presents polygonal purple papules, D 0.3-0.7 mm, flat, showing "Wickham network", accompanied by moderate skin pruritus. At the level of the feet and hands, she presents thin, friable nails, with onycholysis, leukonychia onychorrhexis, melanonychia. At the level of the jugal and semi-mucous membranes of the lips, there were whitish networks with a lacy, fren-leaf appearance (figure no. 1a), and at vulvar level, she presented multiple whitish plaques, some confluent.

Paraclinical investigations were within normal limits. Skin biopsy confirmed the diagnosis of lichen planus, the histopathological examination showing orthokeratotic hyperkeratosis, hypergranulose, acanthosis, hydropic degeneration at the basal layer and lymphohistiocytic infiltrate placed "in the band" in the upper dermis.

Initially, the patient underwent systemic treatment with corticosteroids 0.5mg/ kgc/ day, with progressive decreasing of doses within 1 month, antihistamines H1, antidepressants, anxiolytics, gastric protector and local treatment with class III potency dermatocorticoids, keratolytic, mouthwash with disinfectant solution. The evolution was favourable, but with recurrence approx. 2 weeks after discontinuation of Methylprednisolone.

Systemic corticosteroid therapy (0.5 mg/ kgc/ day with progressive dose reduction within 1 month) was resumed in combination with Acitretin (initially 20 mg/ day, 4 months, with progressive dose reduction, 8 months). At 1-year follow-up, lichen planus lesions were absent.

Figure no. 1. a) Oral Lichen planus at hospitalization and after 6 months of treatment; b) Plantar Lichen planus at hospitalization and after 6 months of treatment

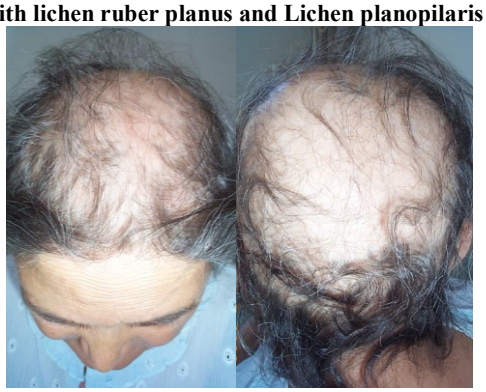


Case 2. Female patient, 80 years old, with associated cardiac pathology is admitted to the dermatovenerology ward for the recurrence of lichen planus manifestations, currently with muco-cutaneous, nails and scalp extension (cutaneous-mucous and hair). Upon hospitalization, she presented typical, disseminated erythematous-papular rash, affecting the mouth and genital mucosa (whitish, reticulated network, with a characteristic fern-leaf appearance at the buccal level and with genital sclerotrophic appearance). Initially, at the level of the scalp, she had multiple small alopecic plaques with follicular papules on the surface.

Over time, the alopecic plaques have expanded through confluence, with the skin of the scalp slightly atrophic and with hair isolated on the surface. Unfortunately, in our patient, during the time, the most serious manifestation of lichen planus was the extended alopecia with the appearance of pseudopelada Brocq. Repeated systemic therapies with corticosteroids have allowed temporary stabilization of the disease, and the intralesional injections with corticosteroids in the alopecic areas have stopped the evolution of hair loss. The evolution of the disease for over 20 years, the late presentation of the patient to the doctor allowed the evolution towards a severe form, with muco-cutaneous, nails and scalp disorder, with a pseudopeladic state, which is definitive in lichen planus

(figure no. 2).

Figure no. 2. Pseudopeladic state with extended alopecia in a patient with lichen ruber planus and Lichen planopilaris



DISCUSSIONS

Palmoplantar lichen planus is a clinical variant, rarely described in the literature, with unknown prevalence and incidence. Most commonly, this form of lichen appears in the 3-5th decade of life. The association of the palmoplantar lesions with the cutaneous-mucous and nails affection, as in our case, is unusual, therefore, being a diagnostic challenge. The presence of typical lichen planus lesions on the preferred typical areas helps the clinician in establishing the diagnosis. At the plantar level, our patient presented the erythematous-squamous form, with pruriginous areas of hyperkeratosis, this being the most common form of plantar lichen planus.(8)

The etiology in the lichen planus is multifactorial and often not elucidated. In our patient, depression and smoking were the main triggers. The literature indicates that stress, anxiety, depression are triggers that are frequently involved in the appearance of the oral lichen planus. The association between smoking and oral lichen planus is still debated, but many studies support it.(14,15)

Skin biopsy is a useful tool in the diagnosis of lichen planus, including palmoplantar form. Histopathologically, the same elements are found in the palmoplantar form as the cutaneous one.

As a peculiarity, in the palmoplantar lichen planus, the horny layer is very thick, and clinically the lesions are yellow instead of purple.(16) There is also a thickening of the palmar and plantar shiny layer, which clinically correlates with the absence of Wickham striations.(17)

The first line of therapy includes class I and II potency dermatocorticosteroids, with anti-inflammatory and pruritus-improving effects.(17)

Second line therapy includes: UVB, systemic and local retinoids, systemic corticosteroids, tacrolimus/ pimecrolimus locally (11), metronidazole, griseofulvin, cyclosporine. UVB phototherapy produces apoptosis of the affected cells and interferes with the immunological functions that inhibit the expression of proinflammatory factors. For a better control of the symptomatology and a faster remission of the disease, systemic corticosteroids are indicated. They are indicated in extended forms of the disease, with acute exacerbations, in patients in whom the mucous membranes are affected, with an adverse response to local treatment.

Third line therapy is represented by: PUVA, thalidomide, mycophenolate mofetil, low molecular weight heparin, and iontophoresis. PUVA is not the ideal choice in the local treatment of lichen planus, but when patients do not respond favourably to UVB, it may be considered a therapeutic option.(12,19,20)

Retinoids are a good choice for the treatment of lichen with palmoplantar damage. Structurally, they are derived from A vitamin or have structural and/ or functional similarities with it. Retinoids bind to retinoid-binding proteins and retinoid nuclear receptors, which specifically activate regulatory regions of DNA involved in cell growth, differentiation and apoptosis. Immunomodulatory, anti-inflammatory, anti-proliferative mechanisms explain the therapeutic efficacy of this class of drugs. It is necessary to periodically monitor the patients treated with retinoids due to multiple adverse effects: cutaneous, musculoskeletal, neurological, ocular, gastrointestinal, psychic, teratogenic. Thus, it is recommended to evaluate liver function at the beginning of oral retinoid therapy and at 1-2 months of treatment, with therapy discontinuation if transaminases increase by> = 3 times. Teratogenicity is proven, so that fertile patients require contraceptive use 1 month before starting treatment and another 2-3 years in the case of acitretin treatment. Some studies claim that there is an association between depression and retinoids, others disprove it.

The mechanism of action is unclear, but it is assumed that retinoic acid may have an effect on areas of the brain involved in depression: striatum, hippocampus, frontal cortex and hypothalamus. Rapid recurrence after Medrol therapy prompted us to initiate retinoid line 3 therapy (Acitretin) after relieving depressive syndrome, the therapy being well tolerated.(21) In the specialized literature, there are several isolated cases, where the efficacy of Acitretin in the systemic treatment of the palmoplantar lichen has been proven.(22-25)

Only a few cases of palmoplantar, mucosal, nail and cutaneous lichen planus forms are published in the specialized literature. Sehgal VN et al describe a case of palmoplantar lichen associated with skin lesions in the flexion areas, nails, mouth in a 52-year-old patient.(26) Wei Chen et al published the case of a 67-year-old patient with palmoplantar and buccal lesions.(27) Arun Joshi et al describe the case of a 4-year-old child with palmoplantar lesions associated with lichen lesions of the trunk and limbs (28), and Mihn Sook Jue et al published the case of a 7-year-old patient with palmoplantar and cutaneous generalized skin damage.(29)

The palmoplantar lichen is a diagnostic challenge for the clinician as well. There is not yet a consensus on its treatment, in the literature being documented only case presentations, clinical studies are needed for the correct and complete management of this form of lichen planus. Associations of cutaneous lichen planus with oral, genital, palmoplantar and nail form as in the case of our patient are exceptionally rare. In the first case, there was no damage to the hair that was present in case 2 and which had as a consequence the evolution towards pseudopelada Brocq, with the irreversible destruction of the hair.

CONCLUSIONS

1. In the first case, depressive syndrome and smoking were the main triggers of the disease. The patient had a very good response to the systemic treatment with retinoids compared to the systemic corticosteroid, under which the rash recurred.
2. In the second case, late presentation to the doctor allowed the lichen to evolve into severe forms, with extended, definitive alopecia of the scalp, even if repeated treatments were performed with systemic corticosteroids and intralesional injections at the alopecic areas.
3. Generalized skin affection due to lichen planus is rarely described in the literature, with the concomitant presence of cutaneous-mucosal lesions and skin attachments (nails and hair), as it was our cases.

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