Severe Atypical Evolution of the Cutaneous Lichen Planus: Case Report

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Abstract

Lichen Planus (LP) is a chronic inflammatory disease of immunological character. It attacks the skin and mucous membranes with lesions ranging from pruritic papules associated with Wickham’s striae to erythematous and ulcerative lesions. It has unknown etiology and treatment based on immunosuppression with the use of corticosteroids. The objective of the present study was to report a case of Lichen Planus with ocular, cutaneous, oral and genital manifestations in a female patient of 40 years. A 40-year-old female patient with a diagnosis of cutaneous LP who started with small ulcerations in the oral cavity and developed papular, ulcerative and erythematous lesions on skin, conjunctiva, oral and genital mucosa within a 2-month period. The patient was initially treated with corticosteroids, showing an improvement in the overall condition, but died due to the spread of the disease. It can be concluded that the case is uncommon, and that the LP should be closely monitored for its severity, progression and chronicity.

Keywords: Lichen Planus; Chronic disease; Corticosteroids

Introduction

Lichen Planus (LP) is a chronic inflammatory disease that is relatively common, affecting skin and mucous membranes including mouth, genitals, nails, and scalp [1,2]. Despite the uncertain etiopathogenesis, it is believed that LP is originated from genetic and immunological factors [3]. Evidence shows a modification of antigens in the skin and mucosa results in an immune reaction of T lymphocytes, leading to a degeneration of the basal cells. Some studies have been done suggesting the relation of emotional state, use of medicines and systemic diseases with beginning or course of the disease [2,4].

Most patients affected by LP is female and middle-aged, with rare child involvement. Cutaneous lesions are described as polygonal, purple and pruritic papules with the presence of fine white lines interlaced on the surface (Wickham’s striae). While mucosal lesions can be divided into reticular, with asymptomatic papules and striae, and erosive when there is presence of erythematous and atrophic areas with central ulceration [1,2,5]. Ocular involvement of the LP usually presents as papular and erythematous lesions on the eyelid, rarely affecting the conjunctiva [3].

The diagnosis of LP is mostly clinical due to the presence of intertwined striae, as it is a sign almost pathognomonic of the disease [1]. However, histological findings should be considered to aid in diagnosis, especially in suspicion of erosive LP due to characteristics of lesions, similar to other diseases [1,3,5]. The differential diagnosis of LP may involve from leukoplakia, candida, and systemic lupus erythematosus to pemphigus, pemphigoid, paraneoplastic pemphigus and erythema multiforme [2,3].

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LP management varies according to the clinical picture and the severity of the disease. Due its immune character, corticosteroids are often effective in wound healing and can be used in topical or systemic route of administration. Being a chronic pathology and is considered a cancerable disease, follow-up is essential to diagnose recurrences, advances and malignant transformation, although the potential for malignancy is still a matter under discussion [1,4].

Thus, the aim of this study was to report a case of Lichen Planus with ocular, cutaneous, oral and genital manifestations in a woman patient of 40 years.

Case Report

Patient R.D.S., female, age 40 perceived lesions in oral cavity (similar to aphthae) associated with whitish plaques on tongue. She sought the Emergency Care Unit where she was treated symptomatically. A few days later, she developed worsening of lesions and sought again the service of Emergency Care Unit and she was treated with Fluconazole for oral candidiasis. At the same time, the patient presented conjunctival hyperemia and pruritus, evolving with worsening of oral lesions, decreased bilateral visual acuity, dysuria, lesions in genital area.

Patient sought hospital service, which was admitted. At the time of admission denied comorbidities and allergies, she reported previous use of Prednisone 40 mg/day for 07 days, without improvement of the lesions - passed to Herpes Zoster treated in 2013. She also reported to be an anxious person and to be going through a period of emotional stress. After hospitalization, the patient was diagnosed with bilateral keratoconjunctivitis, using Dexamethasone Eye drops 5ml, Ocular Lubricant, Ofloxacin 3 mg/ml and Dexpentenol 50 mg/g. During the first 15 days of hospitalization the patient was submitted to treatments with Rocefin, Clindamycin, Aciclovir and Fluconal, without improvement of the condition.

During this period of hospitalization the patient developed erythematous papules in the skin, edema and diffuse hyperemia, with friable areas in vagina, mild bilateral conjunctival hyperemia with crusts on the eyelid (Figure 1), malar and perioral rash with scaling and crusts throughout the region upper lip and granulation tissue, purulent secretion in the lower lip and ulcerated lesions on oral mucosa (Figures 2 and 3A and 3B). Patient with complaints of dyspnea at minimal effort without thoracic pain complaints and laboratory tests within normal limits.

Figure 1: Frontal image evidencing ocular involvement causing crusts and eyelid scaling.
Skin biopsy right cervical region was made and treatment with corticosteroids and prednisone 60 mg/day and five days later was added to treatment Cefepime 1g 08/08h due to dyspnea and changes in pulmonary auscultation suspected of infection nosocomial respiratory tract. After 10 days from the beginning of Cefepime was observed maintaining the above dyspnea and pulmonary auscultation and persistence of purulent secretion in the lower lip. Therefore, it was decided to replace the antibiotic Meropenem 1g 08/08h and 12 Vancomycin 1g/12h, keeping the corticosteroid therapy.

**Figure 2:** Frontal image showing skin rash and scaling in malar region, crusts and purulent secretion on the lips.

**Figure 3A and 3B:** Clinical aspect of lips and perioral region with crusts on lips and perioral rash with scaling and crusts.
The diagnostic hypotheses for the case were lupus erythematosus, pemphigus vulgaris, erosive lichen planus, Behçet’s disease, erythema multiforme, pharmacoderma and seborrheic dermatitis. The result of skin biopsy indicated Dermatitis suggestive Lichenoides of Cutaneous Lichen Planus. Thirty days after the start of corticosteroid therapy, the patient evolved with improvement of conjunctival and vaginal hyperemia, reduction of malar rash and crusts on lower and upper eyelids and lips (Figures 4 and 5) and persistence of cutaneous lesions and oral mucosa ulceration (Figures 6A and 6B).

**Figure 4:** Clinical aspect of the ocular region 30 days after initiation of cortotherapy.

**Figure 5:** Clinical aspect of lesion in face 30 days after the beginning of corticotherapy exhibiting malar rash and crusts in lower and upper lips.
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Severe involvement of oral region prevented a detailed clinical examination of oral cavity. Treatment with corticosteroids associated with Meropenen and Vancomycin showed improvement of the overall picture, without total remission of lesions previously described. However, after 2 months and 10 days from the start treatment, disease spread and the patient died.

Discussion

LP is a chronic mucocutaneous disorder that may involve keratinized and mucosal skin. Often affects the oral and genital mucosa and is associated with pruritus, pain, burning and destruction of the vulvar and vaginal architecture [6,7]. Spanemberg., et al. [8] evaluated the correlation of 274 patients with LP, found that 40.1% had oral lesions, skin lesions 38.7%, 7.7% genital lesions and only 2.6% in three regions lesions. The case reported is similar to literature regarding the affection local, due to the fact that patient has lesions on oral, genital and skin muco sa. However, ocular involvement, as can be observed, is not characteristic, being a more common sign in other diseases such as paraneoplastic pemphigus, systemic lupus erythematosus and erythema multiforme [3].

Soares., et al. [5] and Werneck, Miranda, Junior [7] report that the LP has a slow evolution, often asymptomatic, as well as periods of exacerbation and remission. What defines the clinical picture of our patient as unusual, since there is no previous report of symptomatology and that in two months the lesions evolved from ulcerations in oral mucosa to severe involvement of all regions that the LP can affect.

Usually, clinical features are described as polygonal, purple and pruritic papules associated with Wickham's striae on skin whereas on mucosa, lesions may present as reticular, plaque, papular, bullous, atrophic and erosive [3,5]. The ulcerated and erythematous areas found in patient showed an erosive and atrophic appearance of disease.

Figure 6A and 6B: Intraoral clinical aspect 30 days after corticotherapy showing ulcerations in oral mucosa.
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Approximately 1% of the population may have cutaneous Lichen Planus, while buccal LP affects from 0.1% to 4% of it. It affects an age group of 30 to 70 years and about 60% of the people affected by LP are female [1,4]. It is believed that the pathogenesis of LP occurs through the destruction of basal keratinocytes by T lymphocytes that are recruited to the dermoepidermal junction to induce apoptosis of these cells [6]. However, there are several factors that can trigger this condition. It is suggested that there is connection between the LP and certain systemic diseases, medications and anxiety.

Neville., et al. [1] and Soares., et al. [5], to trace the epidemiological profile of LP patients, found that most of these one were nervous, annoying and anxious people, attesting to the importance of the psychological factor for the development of the disease. Yang., et al. [11] analyzed psychological problems of patients with oral mucosal diseases, including LP. With this, they can notice that almost half of sample had anxiety or depression. Therefore, they show that psychological stress plays an important role in LP, thus interfering in patient’s quality of life and course of disease. Therefore, it is possible to suggest an explanation for the greater affection of feminine gender, because there is a great incidence of emotional stress among women [5].

It is also known that drugs can induce the formation of mucosal ulcerations and erosions [1]. These reactions may occur mainly in middle-aged individuals due to numerous medications such as beta-blockers, non-steroidal anti-inflammatories, methyldopa, antimalarials, penicillamine, among others, which act as triggering antigens [6,7].

Although the medications used by the patient do not fit in described classifications, this information should not be disregarded, since the use of empirical drugs, at the beginning of treatment, coincided with period of her worsening. Alvarez., et al. [10] observe involvement of oral LP, verified that 45.8% of evaluated patients were under antidepressant and anxiolytic treatment, and 32.3% used two or more drugs, which evidence the relation of the ingestion of with LP, as well as the higher incidence of psychological disorders in these patients. With replacement of medications by corticosteroids, a satisfactory result may not be noticed immediately after the exchange or removal of these drugs. Likewise, lesions can also occur months after the onset of use, which makes it more difficult to relate the lesions to the medication used [7].

The ulcerative and erythematous character of lesions associated with Wickham’s striae are often enough to reach the diagnosis of LP. However, biopsy of lesions should be performed to confirm the suspicion and rule out other pathologies [3], especially when there is atypical involvement of disease as in case reported. Ocular involvement and severity of oral and perioral lesions are uncommon for LP, which has increased the number of disorders that could be involved, making it essential to perform the biopsy. The diagnostic hypotheses raised by the medical team resemble those described as differential diagnosis of LP [1-3].

For be a chronic inflammatory disease, treatment of LP is directed to the symptoms that vary according to the overall picture and severity of lesions [3,4]. According to Schmitz., et al. [12] spontaneous remission of cutaneous LP occurs in 64% to 68% of patients, therefore, the case presented is shown to be uncommon due to persistent symptomatology. Initial management is performed by potent topical corticosteroids such as clobetasol 0.05% and if there is no response, systemic corticosteroid oral corticosteroids can be started [6]. The worsening of condition at beginning of patient’s treatment showed that immunosuppressive medication was necessary, even without biopsy result, due to the autoimmune characteristics of disease. Therefore, corticoetherapy with Prednisone 60 mg/day was used, resulting in improvement of patient’s condition, even if discrete, after first month of treatment.

LP is considered a cancerable condition, and although cases of malignant transformation are not well documented. Some authors affirm that the lesions have a malignant potential and that atrophic epithelium of the LP is more susceptible to the action of carcinogens [1,15]. Some authors claim that atrophic epithelium of LP is more susceptible to the action of carcinogenic agents [1]. Thus, monitoring of patient should be long-term. Even in milder cases, patients should be reassessed by a multidisciplinary team at least every three months [1,5].

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Conclusion

It was possible concluded that the present case coincides with clinical aspects of Cutaneous Lichen Planus, but non-standard and severely. Diagnosis should be made through clinical findings and biopsy of lesions. Treatment should be started as soon as possible with corticosteroid therapy, avoiding its evolution. In addition, the patient’s emotional character is an important factor related to the onset and severity of disease. Thus, monitoring of the case in multidisciplinary way is necessary for a long time, mainly because it is a chronic disease.

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Bibliography


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