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Case Study

EVALUATION OF FAMILIAL ORAL LICHEN PLANUS : PRESENTATION OF 3 FAMILIES

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ABSTRACTS

Lichen planus (LP) is a relatively common disorder of the stratified squamous epithelia, which is, in many ways, an enigma. Lichen planus is probably of multi-factorial origin, sometimes induced by drugs or dental materials, often idiopathic, and with an immune-pathogenesis involving T-cells in particular. The occurrence of OLP appears to be complex, with interactions between and among genetic, environmental, and even lifestyle factors. Familial occurrence of alo has always been a debated topic with a limited literature available to support the relevance. In this case report series a total of 112 OLP cases were examined, out of which 3 families with a total of 7 patients were found to be having the familial correlation. The main objective of this paper is to present a case report series of 7 patients based on their familial correlation.

KEYWORDS: Auto-Immune, Lichen, Familial, Mucocutaneous

INTRODUCTION

Delineated and named in 1869 by Erasmus Wilson lichen planus is defined as a common chronic immunological inflammatory mucocutaneous disorder that varies in appearance from keratotic (reticular or plaque like) to erythmatous and ulcerative.[1] The name lichen planus is derived from the Greek word defined lichen as tree moss and planus as flat. Etiopathogenesis of OLP has always remained with different etiologies to be associated with the condition. Though cell mediated immune dysregulation has been associated with the pathogenesis of OLP. However, current data suggests that OLP is T-cell autoimmune disease in which auto-toxic CD8+ T cells trigger the apoptosis of oral epithelial cells. The nature of antigen is uncertain; however there are various predisposing factors that have been implicated in the pathogenesis of OLP. [2]

The role of genetic factors in OLP is still undetermined, as the attempts to identify predisposing human leukocyte antigen (HLA) types have not proved to be conclusive. Copeman et al. found an association of HLA-B7 in familial cases but not in patients with sporadic LP.[3] However, no significant associations were found between HLA and LP in other studies.[4] LP has been reported in families and monozygotic twins, with an incidence of between 1% and 11% of all LP cases.[5] Oral lichen planus in familial cases has a characteristic occurrence with its tendency to develop at an early age, become severe or chronic, and to have widespread atypical manifestations.[5],[6] Oral lichen planus is a chronic inflammatory disease, which is often characterized by bilateral white striations, papules, or plaques on the buccal mucosa, tongue, and gingiva. The clinical presentation of these cases has been variable according to the age sex and severity of the symptoms. The six P's of lichen planus characterize the lesions of lichen planus which are

planar, polygonal, purple, pruritic, papules and plaques. The lesions often occur bilaterally, intra-orally and on the flexor surfaces of the extremities. [7]

CLINICAL CASES

A total of 112 cases were diagnosed in the department of Oral Medicine PGIDS, Rohtak from April 2012 to March 2013. All the patients were examined according to the clinical & family history, age, sex, site extent of the lesion in the oral cavity. Other associated symptoms like pain, burning sensation and discomfort were considered. Biopsy was done to confirm the diagnosis. Final diagnosis of Lichen planus was made only after both the clinical and histo-pathological criteria specified by World Health Organization (WHO) 1978, modified by Van Der Meij et al. in 2003 were fulfilled. [8] Family members of all the patients were examined to rule out the familial predisposition. Within these 3 families a total of 7 patients showed the familial occurrence of this condition. The other family members who were tested positive for OLP in clinical examination were also marked for biopsy for the confirmation of diagnosis except for one. All the patients with familial predisposition were further divided in family 1(A), family 2(B) & family 3(c).They were evaluated according to the age, location, duration, associated clinical features and their treatment. (Table 1)

Table 1

S no	Sex	Age	Relation-ship	Location	Type of lichen	Associated symptoms	Diagnosis
A	Female (pt 1)	46 yrs	Sister	Bilateral buccal mucosa	Reticular	Pain & discomfort	Clinical & histological
	Female (pt 2)	39 yrs	Sister	Bilateral buccal mucosa & Desquamation of gingiva	Erythmatous / Atrophic	Burning sensation while eating food	Clinical & histological
B	Female (pt 3)	43 yrs	Mother	Bilateral buccal mucosa & abdominal folds	Reticular/ Papular	Burning sensation	Clinical & histological
	Male (pt 4)	12 yrs	Son	Ankle area of both legs	Violaceous plaque	Itching	Clinical
C	Female (pt 5)	59 yrs	Mother	Buccal mucosa, tongue & gingival	Reticular Erosive	Burning sensation	Clinical & histological
	Male (pt 6)	62 Yrs	Brother	Scalp & Buccal mucosa	Reticular	Discomfort while eating	Clinical
	Female (pt 7)	42 yrs	Daughter	Buccal mucosa	Reticular	Asymptomatic	Clinical & histological

Case Report 1

In Family A, the first patient (Pt 1) to report was a female of 46 years of age, who complained of pain and discomfort while eating food in the right & left buccal mucosa region from the past 2 years. On examination bilateral reticular striations extending from the mid - buccal mucosa to the retro molar pad area were seen. (Figure 1) Striations were diffuse and accompanied by the grayish discoloration. Diagnosis of "Oral Lichen Planus" was made upon the basis of history, clinical examination and histo pathological investigations.(Figure 2) On taking the family history patient reported of her elder sister (Pt 2) suffering from the same problem with the more intense burning sensation. On examination erythmatous/atrophic type of lichen planus was seen in bilateral buccal mucosa along with the desquamation of the gingiva.

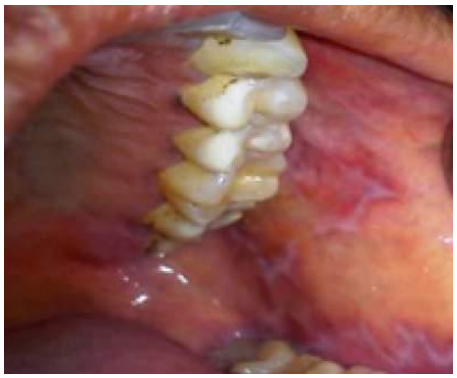

Figure 1: *Reticular striations on buccal mucosa*

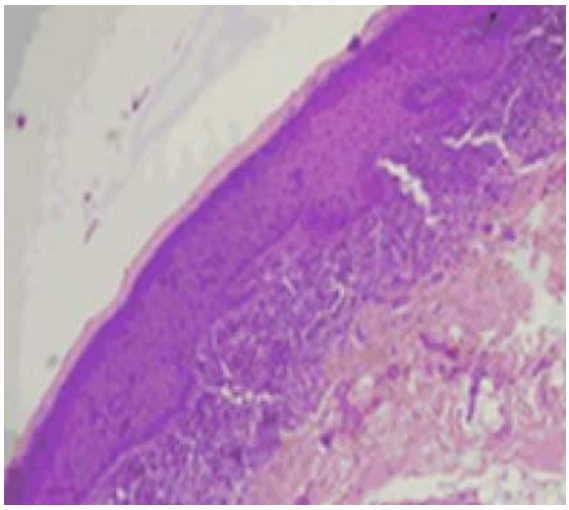

Figure 2: *Photomicrograph 10x view*

Case Report 2

In family B, The first patient (Pt 3) to report was a female of 43 years old who reported with the complaint of burning sensation in the mouth. On examination reticular striations were seen in the bilateral buccal mucosa along with the papular patches in the abdominal fold region. She was diagnosed to be a case of lichen planus on the basis of history, clinical examination and histo-pathological investigations. She also reported about her 12 years son (Pt 4) on having the similar type of lesion of his legs. On examination violaceous plaque was seen on the ankle region of his right leg. (Figure 3)

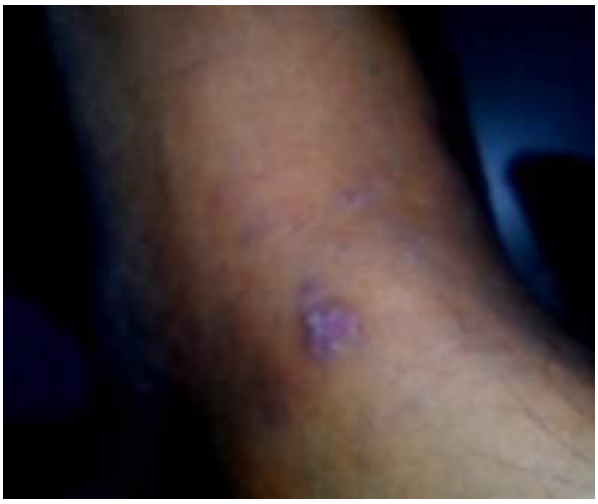

Figure 3: *Violaceous plaque on ankle*

Case Report 3

In family C, a 59 year old female patient (Pt 5) was the first to report with the chief complaint of burning sensation in the mouth. On examination erosive type of lichen planus was diagnosed on bilateral buccal mucosa, which was later confirmed by the histo-pathological investigations as well. On taking family history patient revealed that her brother and daughter as well were suffering from the similar type of problem. Her brother (Pt 6) reported with the reticular striations of buccal mucosa and keratotic papules associated with alopecia of the scalp(Figure 4) Daughter (Pt 7) reported with reticular striations on the buccal mucosa but she was apparently asymptomatic.

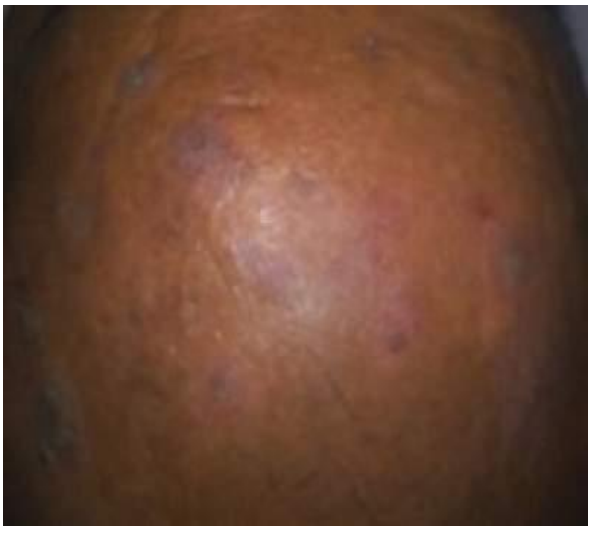

Figure 4: *Alopecia & popular lesions on scalp*

TREATMENT

Treatment plan for all the patients varied according to the severity of the lesion and discomfort of the patient. In general; all the patients were put on the initial antioxidants. Pt 1 who reported with who discomfort

while eating was put on immune-modulators e.g. Tab. Dicaris 150 mg once a day for 5 days. It was accompanied by topical Oint. Kenacort (Tacrolimus) 0.1% Local Application 3 times/day for 10 days. An antioxidant readily available in the market was also used for all the patients. Pt 2 who presented with more severe symptoms was put on Tab. Wysolone 30 mg once a day which was later tapered down subsequently after 10 days. Oint kenacort 0.1% was used for topical application as well. Pt 3 & Pt 4 was treated in harmony with dermatology department as there was presence of skin lesions as well. For the oral lesions pt 3 was put on topical ointment and anti-oxidants as used for the other patients. Pt 5 was put on systemic steroid therapy whereas Pt 6 was given the topical treatment only. Pt 7 was asymptomatic so no need for any aggressive treatment was felt.

DISCUSSION

Lichen planus is a common mucocutaneous disease which is thought to affect 0.5 to 1% of the population.[9] It is a disease of the middle-aged population and is more common among women.[10] Etiology of OLP has always been an enigma. However, an immunologically induced degeneration of the basal cell layer of the oral mucosa has been suggested.[11] In the past, speculation about the etiology covered a wide range of possibilities including trauma, specific bacteria, syphilis, parasites, viruses, mycotics, allergies, toxicity, neurogenic, hereditary and psychosomatic disorders.[11] Familial occurrence of LP is a well-recognized but rare event, with an incidence varying from 1% to 11% of all LP patientsIn this case report series a total of 7 patients out of 3 families were evaluated to be having a familial correlation.

Lowe et al first reported the significance of HLA in OLP patients, with the frequency of raised HLA-A3 in the patients with cutaneous OLP. In comparison with classic LP, familial LP is characterized by its early age of onset, atypical and widespread clinical presentation, and its higher tendency to become severe. Different HLA haplotypes were reported in familial lichen planus such as HLA-B7, -Aw19, -B18 and CW8. In non-familial lichen planus HLA-A3, -A5, -A28, -B8, -B16 and -BW35 are more common.[9] Familial occurrence is quite evident from the present case series as well as the cases reported by Bermejo-Fenoll & Jornet in 2006 .[12]

Due to the limited resources HLA (human leukocyte antigen) correlation could not be studied in this case report series, but the familial presence was confirmed on the basis of previous care reports and studies done in this aspect.

Clinical feature of all the cases in this case report were similar to all those described in other studies which has been previously done. OLP predominantly manifests in the fifth and sixth decades of life, and is more common in women.[13] It is well accepted that OLP is a chronic, possibly lifelong, disease that is characterized by remissions and exacerbations.[14],[15] Likewise in this case report all the cases showed the exacerbation as well as the remission at different stages which concur with the studies done by Bethke G et al & Piboonniyom SO et al in 2005. In the treatment, Corticosteroids have been the mainstay of management of OLP; yet, other modalities like calcineurin inhibitors, retinoids, dapsone, hydroxychloroquine, mycophenolate mofetil and enoxaparin have contributed significantly toward treatment of the disease. According to lavanya et al blocking IL-12, IFN-γ, , TNF-α,RANTES, or MMP-9 activity or up regulating TGF-β1 activity in OLP may be of therapeutic value in the future.[16] In this case report the treatment of the lichen planus was based upon the severity of the lesion and the symptoms elicited by the patients. Patients are currently under the regular follow up. Thus it can be concluded that familial co-relation of the patients might be rare, but the occurrence is evident and it can't be ruled out.

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