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**Running head:** OLP in childhood



**Abstract**

**Background**: Although the exact incidence of pediatric oral lichen planus (OLP) is unknown, the oral mucosa seems to be less commonly involved and the clinical presentation is often atypical. The aim of the study is to present a case series of oral lichen planus in childhood.

**Methods:** From our database, we retrospectively selected and analyzed the clinical data of oral lichen planus patients aged under 18 where the diagnosis had been confirmed by histopathological analysis.

**Results:** The case series from our database shows eight patients, four males and four females. The mean (± SD) age at the time of diagnosis of the disease was 13.5 (±2.73) years, ranging in age from 9 to 17. Clinically, a reticular pattern was present in six patients (75%) and the tongue was the most commonly involved oral site (6 cases, 75%). We also report the first case of OLP in a 9 year-old girl affected by autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (APECED).

**Conclusions:** We report the largest case series of pediatric OLP so far published in literature. Differences in the disease between adults and pediatric patients have been detected but further investigation and a larger case series are needed to establish any detailed differences in clinical outcomes.

**Introduction**

Lichen planus (LP) is a common chronic inflammatory disorder that affects the skin, mucous membranes, nails, and scalp; the etiology of this condition is still unknown although an immune-mediated pathogenesis has been hypothesized.[1] The age at onset is usually between the third and sixth decade of life and it is predominantly seen in females.[2]

Although the exact incidence of LP is unknown, it seems to vary between 0.1% to 1.2%. Children represent only 1% to 4% of patients with LP and the clinical presentation is often atypical.[3]

Clinically, cutaneous LP is characterized by purple, polygonal, pruritic papules frequently covered by a lacy network of white scales on their surface, known as Wickham striae. The flat-topped papules are often located on the flexor surface of the wrist, the shins, the trunk, and the medial thighs, subdivided into one of the following variants: linear, hypertrophic, annular, follicular, actinic, vesciculobullous and pemphigoid-like. The disease often resolves within 8 to 12 months of treatment and it is not believed to be capable of malignant transformation.[4,5]

Any nail involvement may appear as a thinning of the nail plate, longitudinal fissuring, or distal splitting. Any hair follicle involvement is called lichen planopilaris and if untreated can lead to scarring alopecia. Any involvement of the mucous membranes can affect the oral mucosa, conjunctivae, larynx, esophagus, tonsils, bladder, vaginal vault, vulva, and anus.[5]

In contrast to skin LP, oral lichen planus (OLP) demonstrates a clinical variability[6] and the oral manifestation in adults is more frequently resistant and persistent than the cutaneous type.[7]

The oral lesions are categorized as reticular, papular, plaque-like, atrophic, erosive, or bullous.[8]

The hyperkeratotic variants are commonly asymptomatic while the atrophic/erythematous variant, the erosive/ulcerative variant and the bullous type often have persistent symptoms of pain or stinging aggravated during eating and drinking.[9,10]

The clinical differential diagnosis depends on the age of the patient, the clinical form of OLP, and the severity and persistence of the lesions and includes: lichenoid drug reaction, leukoplakia, lupus erythematosus, candidiasis, graft-versus-host disease (GVHD), frictional keratosis, autoimmune bullous diseases, erythema multiforme, allergic gingivostomatitis, and gluten sensitivity enteropathy.[10–12]

In challenging cases more sensitive diagnostic techniques could be useful to achieve diagnosis such as direct and indirect immunofluorescence. [13]

The aim of this study is to provide an update of the oral involvement of the disease in children through the report of a retrospective analysis of pediatric patients referred to our Department during the last four years for whom lichen planus has presented in the oral cavity as the single or as an additional site of involvement. We also conducted a literature review of the topic in order to highlight the similarities and differences between our data and the previously published clinical cases.

**Subjects and Methods**

From our database, we retrospectively selected and analyzed the clinical data of pediatric OLP patients in the outpatient clinic of the Department of Neurosciences, Reproductive and Odontostomatological Sciences, Federico II University of Naples. The selection was based on the following inclusion criteria:

- age <18 years old at the time of diagnosis
- a clinical and histological diagnosis of OLP

The exclusion criteria were

- GVHD lichenoid lesions
- the lack of a confirmatory histology

- oral lichenoid drug reaction

- the lack of any results of routine hematological testing including tests for hepatic and kidney functionality, markers of hepatitis A, B and C viruses, and a red and white blood cell count and platelet count.

From our database we collected the following data: age at time of diagnosis, sex, preexisting medical conditions, presence of a positive family history of immunological disorders, concomitant or previous assumption of drugs, concomitant oral predisposing or iatrogenic factors, confirmatory histology, clinical pattern, oral sites involved, oral symptoms reported, extra-oral sites involved, and the treatment and resolution of oral lesions.

Literature review

A PubMed search was carried out of articles published between 1966 and 2015 using the keywords “lichen” OR “lichenoid” alternatively matched with “oral” OR “lip” AND “juvenile” OR “child*” OR “familial” OR “pediatric”.

The selection of the studies was based on the following inclusion criteria:

- the English language

- a case series or case reports

- age <18 years old at the time of diagnosis

- clinical and histological diagnosis of OLP

- an accurate description of the oral sites and clinical features

The exclusion criteria were

- lack of clarity in reporting data about the clinical form(s) of OLP and/or the oral site(s) involved

- GVHD lichenoid lesions

- lack of confirmatory histology

- oral lichenoid drug reaction

The study was approved by the Ethics Committee of the University of Naples "Federico II" in July 2014 and it conforms to the provisions of the Declaration of Helsinki (as revised in Tokyo 2004).



**Results**

The case series from our database shows eight patients, four males and four females. The mean (± SD) age at the time of diagnosis of the disease was 13.5 (±2.73) years, the patients ranging in age from 9 to 17.

A positive familial history of immunological disorders was found in seven cases (87.5%). Seven patients had been submitted to hepatitis B virus (HBV) vaccination and three (37.5%) patients presented concomitant oral factors. Findings and/or a history of an immune disorder were present in seven patients (87.5%).

For each patient a confirmatory histology was obtained and in no case was dysplasia reported.

A reticular pattern was the one most frequently reported, present in six (75%) patients, followed by the atrophic (50%), plaque-like (37.5%), erosive (12.5%) and bullous (12.5%) patterns; no patient showed a papular pattern. A simultaneous multiple clinical pattern was observed in six cases (75%) and in one patient a mucosal pigmentation was detected.

The tongue was the most commonly involved oral site (6 cases, 75%), followed by the buccal mucosa (4 cases, 50%), gingiva (3 cases, 37.5%), retromolar fossae (1 case, 12.5%), palate (1 case, 12.5%) and lip (1 case, 12.5%). No patient showed any floor of the mouth or extra-oral involvement. Four patients (50%) were symptomatic.

The most commonly used drugs were topical antifungal medications prescribed in order to avoid overlapping fungal overgrowth. Topical steroids were also associated in three cases.

Two patients (25%) showed a complete disappearance of the oral lesions.

The analysis of the literature yielded 344 articles published between 1966 and 2015. After the application of the inclusion criteria, 12 articles were included in our study. We also searched among the references of the aforementioned articles and found a further six articles making a total of 18 articles that fulfilled our inclusion criteria, which described a total of 26 patients.

The flow chart is reported in Table 1.

Data from our cases concerning the epidemiology, predisposing factors, clinical features, diagnosis and treatment are recorded in Table2.

Data from the PubMed search concerning the epidemiology, predisposing factors, clinical features, diagnosis and treatment are recorded in Table 3.

A comparison between our cases and the review of literature is provided in Table 4.

**Discussion**

Many previous studies have reported that in LP among pediatric patients the oral mucosa seems to be less commonly involved with a prevalence of approximately 0.03%[17] compared with 1% to 2% of the general population.[2]

The present case series confirms the epidemiological data previously collected concerning the pediatric LP population with a balanced M:F ratio[25] and a greater prevalence for familial LP in children (25%) than in adults. In fact, although LP is usually a sporadic disorder, there is a rare familial form more prevalent in the pediatric population ranging from 1% to 4.3%, with childhood familial LP considered to occur at an earlier age and with a greater severity.[3] An autosomal dominant mode of inheritance with a variable penetration has been suggested and a linkage of familial LP with HLA-B7 and HLA-BR10 has been observed.[3]

The exact etiology of LP is unknown but it appears to be complex and multifactorial. Possible cofactors of OLP, such as a hypersensitivity to dental restorative materials (e.g. amalgam and gold), local trauma (the Koebner phenomenon) and several kinds of infections (plaque-causing microorganisms and hepatitis B or C virus infection) have been reported.[10,25]

Furthermore childhood LP has been documented as a complication of hepatitis B virus (HBV) vaccination, where the recombinant proteins of the HBV vaccine – specifically the viral S epitope – may trigger a cell-mediated autoimmune response targeted at the keratinocytes.[11]

For these reasons, in Tables 2 and 3 we have recorded the presence of any concomitant systemic and oral factors that could have had a role in the OLP pathogenesis or in its exacerbation; however to date these associations are still unclear. [7]

The medical histories collected from our cases confirm the presence of an increased association between OLP and auto-immune diseases[1,3,7,10,11,22] with seven of our cases showing associated immunological disorders. Among our patients we also report the first case of an oral lichenoid lesion related to autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (APECED) also known as polyglandular autoimmune syndrome (PGA) type I, in a 9 year-old girl. Until now there has been only one other case in literature of OLP associated autoimmune polyendocrine syndrome type II[26] in a 42 year- old woman, reinforcing the suggestion of a common immune-mediated pathogenesis between OLP and PGA. (Fig. 1)

From the reported data in our case series the reticular pattern appears to be the most common in childhood followed by the erosive one, in accordance with the literature. Interestingly, we instead recorded a clear difference, when comparing our eight cases to the literature (Table 4), concerning the oral site predilection with 75% of our eight pediatric patients showing lesions on the tongue. Previous studies have reported the buccal mucosa as the most commonly involved oral site in pediatric OLP with the next most common location being the tongue.[25] Finally, confirming the hypothesis of a less common oral involvement in pediatric patients with LP[25] previously reported at a rate of 12.6%[27], none of our eight cases showed any extra-oral involvement.

The histology of OLP has revealed that parakeratosis is the most frequent type of keratosis while the erosive variety has involved the acanthotic epithelium in more than 50% of cases; the rete pegs are predominantly of a wavy pattern while basal cell degeneration and band-like subepithelial lymphocytic infiltration seem to be present in all cases. Our data are consistent with the previous literature with basal cell degeneration and band-like subepithelial lymphocytic infiltration present in all of our eight cases.[25]

The treatment of juvenile OLP does not differ significantly from the treatment of adult OLP and is often unnecessary in asymptomatic patients. Oral symptoms are relatively frequent when the erosive and/or atrophic pattern occurs; although patients with a keratotic form can report a roughness, treatment is rarely necessary. In the present paper we have considered prescribing topical antifungal medications to all patients in order to avoid overlapping fungal overgrowth in patients undergoing contemporary topical steroidal therapy. The analysis of previously published studies confirms that topical corticosteroid therapy is the most commonly used treatment in symptomatic OLP, reported in connection with 12 of the 18 symptomatic pediatric patients (66.6%), even if the chronic use of topical steroids can lead to

oral candidiasis; an association with retinoid therapy and a plaque control regimen in children have shown favorable responses.[11] Systemic steroid therapy and dapsone are typically reserved for refractory and recurrent cases; extreme caution is employed because significant long-term effects are of concern in this young patient population. Of note, tacrolimus ointment, topical tretinoin and topical cyclosporine have also been used with success in some cases[6] but the safety of any long term continuous use of some of these drugs in pediatric patients has not been adequately evaluated.[25] The effect of the treatment of OLP in children seems to be more favorable than in adults for whom the symptoms usually persist for many years in spite of intensive treatment and a thorough investigation of any associated factors.[17] Considering our case series and the previously reported papers, a complete resolution of oral lesions has been observed in 38.2% of cases (Table 4).

In Table 5 we show a summary of the most important similarities and differences in OLP between children and adults so far reported in literature.

In conclusion, our case series mostly mirrors previous epidemiological, clinical and therapeutic knowledge about pediatric OLP but a larger case series is needed to confirm the possibility of a different oral site predilection between adults and children as suggested by our findings. Clinicians must be aware that OLP in childhood may also have a simultaneous or future involvement of the skin and other mucosal sites[6] and, due to a more frequent positive familial history of LP in childhood, close relatives should be examined. Although there have been no OLP-related malignancies described to date in the pediatric population, most previous studies suggest that the schedule of follow-up of pediatric OLP should be of at least one or two examinations per year as long as the OLP persists[25] even if the prognosis seems to be more favorable. [19]

What is new:

- the present paper represents the largest case series so far published

- we report the first case of an oral lichenoid lesion related to APECED in a 9 year-old girl

- our cases suggest a different site predilection in OLP between children and adults

- the accurately tabulated review of the literature relating to pediatric OLP may facilitate further studies on the topic

**Limitations of the study:**

even if the present paper represents the largest case series so far published, further studies are needed to establish the epidemiological and clinical features in this population of patients. The review of the literature conducted may be conditioned by previous biases in reporting exceptional cases on PUBMED and therefore the results of the present comparison with previous studies should be considered critically.

**References**


1 Patel S, Yeoman CM, Murphy R. Oral lichen planus in childhood: a report of three cases. Int J Paediatr Dent 2005;15:118–22.

2 Munde AD, Karle RR, Wankhede PK, Shaikh SS, Kulkurni M. Demographic and clinical profile of oral lichen planus: A retrospective study. Contemp Clin Dent 2013;4:181–5.

3 Pandhi D, Singal A, Bhattacharya SN. Lichen planus in childhood: a series of 316 patients. Pediatr Dermatol 2014;31:59–67.

4 Madke B, Gutte R, Doshi B, Khopkar U. Hyperkeratotic Palmoplantar Lichen Planus in a child. Indian J Dermatol 2013;58:405.

5 Sharma A, Białynicki-Birula R, Schwartz RA, Janniger CK. Lichen planus: an update and review. Cutis 2012;90:17–23.

6 Woo VL, Manchanda-Gera A, Park DS, Yoon AJ, Zegarelli DJ. Juvenile oral lichen planus: a report of 2 cases. Pediatr Dent 2007;29:525–30.

7 De Moraes PC, Teixeira RG, Tacchelli DP, Bönecker M, Junqueira JLC, Oliveira LB. Atypical case of oral lichen planus in a pediatric patient: clinical presentation and management. Pediatr Dent 2011;33:445–7.

8 Andreasen JO. Oral lichen planus. 1. A clinical evaluation of 115 cases. Oral Surg Oral Med Oral Pathol 1968;25:31–42.

9 GunaShekhar M, Sudhakar R, Shahul M, Tenny J, Ravikanth M, Manikyakumar N. Oral lichen planus in childhood: A rare case report. Dermatol Online J 2010;16:9.

10 Laeijendecker R, Van Joost T, Tank B, Oranje AP, Neumann H a M. Oral lichen planus in childhood. Pediatr Dermatol 2005;22:299–304.

11 Pendyala G, Joshi S, Kalburge J, Joshi M, Tejnani A. Oral lichen planus: a report and review of an autoimmune-mediated condition in gingiva. Compend Contin Educ Dent 2012;33:e102–8.

12 Sanjaya P, Hallikeri K, Angadi P. Disseminated form of childhood lichen planus. East J Med 2013;16:72–5.

13 Van der Meij EH, van der Waal I. Lack of clinicopathologic correlation in the diagnosis of oral lichen planus based on the presently available diagnostic criteria and suggestions for modifications. J Oral Pathol Med 2003;32:507–12.

14 Chandna P, Adlakha VK, Singal G, Sharma R. Pediatric oral lichen planus: review and case report. Curr Pediatr Rev 2014;10:292–6.

15 Khandelwal V, Nayak PA, Nayak UA, Gupta A. Oral lichen planus in a young Indian child. BMJ Case Rep 2013;2013.

16 Moger G, Thippanna CK, Kenchappa M, Puttalingaiah VD. Erosive oral lichen planus with cutaneous involvement in a 7-year-old girl: a rare case report. J Indian Soc Pedod Prev Dent 2013;31:197–200.

17 Padmini C, Bai KY, Chaitanya V, Reddy MS. Ulcerative lichen planus in childhood. Case Rep Dent 2013;2013:874895.

18 Chaitra TR, Telgi RL, Kishor A, Kulkarni AU. Juvenile oral lichen planus: a clinical rarity. BMJ Case Rep 2012;2012.

19 Anuradha C, Reddy GS, Nandan SRK, Kumar SR, Reddy BVR. Oral mucosal lichen planus in nine-year-old child. N Y State Dent J 2011;77:28–30.

20 Mohan Das U, Jp B. Oral lichen planus in children. Int J Clin Pediatr Dent 2009;2:49–51.

21 Singal A. Familial mucosal lichen planus in three successive generations. Int J Dermatol 2005;44:81–2.

22 Alam F, Hamburger J. Oral mucosal lichen planus in children. Int J Paediatr Dent 2001;11:209–14.

23 Scully C, de Almeida OP, Welbury R. Oral lichen planus in childhood. Br J Dermatol 1994;130:131–3.

24 Borrego Hernando L, Vanaclocha Sebastián F, Hergueta Sánchez J, Ortiz Romero P, Iglesias Diez L. Lichen planus pemphigoides in a 10-year-old girl. J Am Acad Dermatol 1992;26:124–5.

25 Chatterjee K, Bhattacharya S, Mukherjee CG, Mazumdar A. A retrospective study of oral lichen planus in paediatric population. J Oral Maxillofac Pathol 2012;16:363–7.

26 Mignogna MD, Fedele S, Lo Russo L, Ruoppo E, Lo Muzio L. Polyglandular autoimmune syndrome type II associated with oral lichen planus. Int J Dermatol 2002;41:244–6.

27 Handa S, Sahoo B. Childhood lichen planus: a study of 87 cases. Int J Dermatol 2002;41:423–7.

28 Lodi G, Carrozzo M, Furness S, Thongprasom K. Interventions for treating oral lichen planus: a systematic review. Br J Dermatol 2012;166:938–47.

29 Barbosa NG, Silveira ÉJD, Lima EN de A, Oliveira PT, Soares MSM, de Medeiros AMC. Factors associated with clinical characteristics and symptoms in a case series of oral lichen planus. Int J Dermatol 2015;54:e1–6.

30 Carbone M, Arduino PG, Carrozzo M, et al. Course of oral lichen planus: a retrospective study of 808 northern Italian patients. Oral Dis 2009;15:235–43.

Table 1

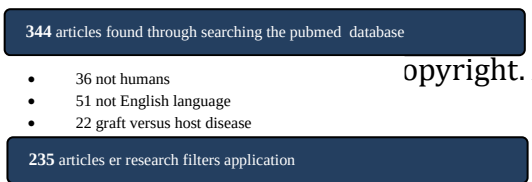


Table 2

| Case | Age | Sex | Preexisting Medical conditions | Family history | HBV vaccination status | Drugs | Concomitant oral factors | Confirmatory histology | Clinical pattern | Oral sites | Oral symptoms | Extra-oral sites | Treatment | Resolution of oral lesions |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 | 17 | F | allergy to NSAIDs † | atopy (mother) | **vaccinated** | none | parafunction | HE ‡ | plaque-like + erosive<br>plaque-like + erosive | margins of tongue bilaterally<br>ventrum of tongue bilaterally | **burning with spicy foods** | none | topical steroids<br>topical antimycotics | none |
| 2 | 17 | M | atopy, ↓ Hb § | OLP (father) | **vaccinated** | none | none | HE | reticular<br>plaque-like | hard palate<br>retromolar fossae bilaterally | **dryness** | none | topical antimycotics | none |
| 3 | 14 | F | none | none | **vaccinated** | none | none | HE | plaque-like<br>plaque-like + atrophic<br>plaque-like + atrophic<br>plaque-like + pigmentation | buccal mucosa right side<br>margins of tongue bilaterally<br>ventrum of tongue bilaterally<br>lower lip | **none** | none | topical antimycotics | **complete** |
| 4 | 14 | M | Hg ($NH_2$) C patch test ++<br>IgG-EBV ¶ 159 U/l<br>erythema discromicum perstans | none | **vaccinated** | none | orthodontics | HE | reticular<br>reticular<br>reticular + atrophic<br>reticular + atrophic | buccal mucosae bilaterally<br>gingiva<br>dorsum of tongue bilaterally<br>margins of tongue bilaterally | **none** | none | topical antimycotics | none |
| 5 | 15 | M | autoimmune thyroiditis<br>atopy | autoimmune thyroiditis (mother) | **vaccinated** | none | none | HE | reticular<br>reticular | buccal mucosae bilaterally<br>ventrum of tongue bilaterally | **none** | none | topical antimycotics | complete |
| 6 | 9 | F | APECED†† | N/A ‡‡ | **N/A** | N/A | none | HE | bullous + atrophic + reticular<br>bullous + atrophic + reticular<br>reticular<br>bullous + atrophic + reticular | dorsum of tongue<br>margins of tongue bilaterally<br>gingival<br>buccal mucosae bilaterally | **pain** | none | topical steroids<br>topical antimycotics | none |
| 7 | 11 | F | atopy | OLP (father) | **vaccinated** | none | parafunction | HE | reticular | gingiva | **none** | none | topical antimycotics | none |
| 8 | 11 | M | atopy<br>immune carrier for Mediterranean anemia | Chron's disease<br>rheumatoid arthritis | **vaccinated** | none | none | HE | reticular + atrophic | dorsum of tongue | **roughness** | none | topical steroids<br>topical antimycotics | none |

| Authors | Age | Sex | Preexisting Medical conditions | Family history | Drugs | Concomitant oral factors | Confirmatory histology | Clinical pattern | Oral sites | Oral symptoms | Extra-oral sites | Treatment | Resolution of oral lesions |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Chandna et al. [14] (2014) | 7 | F | none | none | none | none | HE ‡ (oral) | reticular<br>papular + reticular + pigmentation | buccal mucosae bilaterally<br>upper and lower lips | **none** | skin | oral hygiene<br>topical steroids | **partial** |
| Khandelwal et al. [15] (2013) | 10 | F | ↓ Hb §<br>(10.2 g) | none | none | N/A ‡‡ | HE + DIF §§<br>(oral) (oral) | reticular + pigmentation<br>reticular + pigmentation | buccal mucosae bilaterally<br>retromolar fossae bilaterally | **burning with spicy foods** | none | topical retinoids | none |
| Moger et al.[16] (2013) | 7 | F | none | none | none | caries at 36,63,65,74 | HE + DIF<br>(oral) (oral) | reticular+ atrophic + erosive + papular + pigmentation | buccal mucosae bilaterally | **burning with spicy and hot foods** | skin | topical antimycotic<br>topical steroids<br>topical anesthetic | partial |
| Padmini et al.[17] (2013) | 12 | M | none | N/A | none | none | HE (oral) | erosive | dorsum of tongue | **burning with spicy foods** | none | topical steroids<br>topical anesthetic<br>topical antimycotic | complete |
| Chaitra et al. [18] (2012) | 9 | F | none | N/A | none | none | HE‡ (oral) | reticular + erosive + pigmentation | buccal mucosae bilaterally | **burning with spicy foods** | N/A | topical steroids<br>topical anesthetic | none |
| Pendyala et al.[11] (2012) | 17 | M | none | none | none | none | HE (oral) | bullous | gingiva | **burning exacerbated by acid and spicy foods** | none | topical steroids<br>topical NSAIDs †<br>antioxidants<br>Chlorhexidine | complete |
| De Moraes et al.[7] (2011) | 7 | F | none | N/A | none | parafunction | 2 X HE<br>(oral) | reticular<br>reticular + atrophic | buccal mucosae bilaterally<br>upper lip | **discomfort (upper lip)** | N/A | topical steroids<br>topical CNIs ¶¶ | complete |
| Anuradha et al.[19] (2011) | 9 | F | none | none | N/A | N/A | HE + DIF<br>(oral) (oral) | reticular<br>reticular | buccal mucosae bilaterally<br>retromolar fossae bilaterally | **burning with spicy foods** | none | N/A | none |
| Sanjaya et al.[12] (2011) | 9 | M | none | none | none | N/A | 2 X HE<br>(oral + skin) | reticular<br>reticular<br>reticular<br>reticular | buccal mucosae bilaterally<br>hard palate<br>lower and upper lips<br>tongue | **burning with spicy foods** | skin | systemic steroids | complete |
| GunaShekar et al.[9] (2010) | 7 | M | none | none | none | caries at 36,37 | HE (oral) | reticular + atrophic<br>reticular + atrophic<br>reticular + atrophic<br>reticular + atrophic<br>reticular + pigmentation | buccal mucosa right side<br>margins of tongue bilaterally<br>dorsum of tongue<br>floor of mouth<br>upper and lower lips | **soreness with spicy foods (tongue)** | none | chlorhexidine<br>topical steroids | none |
| Mohan Das & Jp[20] (2009) | 12 | F | ↓ Hb<br>(9 g) | none | none | none | HE (oral) | reticular + erosive + pigmentation | buccal mucosae bilaterally | **burning on consuming food** | none | topical retinoids | none |
| Woo et al.[6] (2007) | 9 | F | ANA + ††† | none | none | none | HE (oral) | reticular + papular + erosive<br>reticular + papular + erosive | margins of tongue bilaterally<br>ventrum of tongue | **dryness** | none | topical steroids<br>systemic steroids | N/A |
| Woo et al.[6] (2007) | 11 | F | none | none | none | orthodontics | HE (oral) | reticular<br>reticular | buccal mucosae bilaterally<br>ventrum of tongue bilaterally | **none** | none | orthodontics removal | none |
| Singal [21] (2005) | 11 | M | N/A | OLP ( father)<br>OLP (grandmother) | N/A | N/A | HE (oral) | plaque-like + pigmentation | dorsum of tongue | **burning with hot and spicy foods** | none | topical steroids | complete |
| Laeijendecker et al. [10] (2005) | 11 | F | fragrance mix 8% pet patch test ++ | none | none | four amalgam fillings | HE (oral) | reticular | buccal mucosae bilaterally | **none** | none | none | complete |
| Laeijendecker et al. [10] (2005) | 16 | M | none | none | none | eight amalgam fillings, poor oral hygiene | HE + DIF<br>(oral) | erosive<br>erosive | buccal mucosae bilaterally<br>gingiva | **pain and stinging** | none | topical steroids<br>topical anesthetic<br>topical CNIs<br>systemic steroids<br>systemic CNIs | complete |
| Laeijendecker et al. [10] (2005) | 14 | F | atopy<br>nickel sulfate 5% pet patch test ++ | none | cetirizine | orthodontics | yes | reticular<br>reticular | buccal mucosae bilaterally<br>margins of tongue bilaterally | **soreness** | none | systemic steroids<br>systemic CNIs | complete |
| Patel et al. [1] (2005) | 15 | F | ↓ferritin (8µg/L)<br>↓IgA ¶¶¶ (0.60 g/L) | none | none | none | HE (N/A) | erosive + reticular<br>erosive + reticular | margins of tongue bilaterally<br>dorsum of tongue | **pain** | skin | topical steroids | none |

Table 3

| | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | hypothyroidism | | | | | erosive + reticular | floor of mouth | | | | |
| Patel et al. [3] (2005) | 6 | M | autism | none | none | N/A | HE (oral) | plaque-like | dorsum of tongue | **none** | none | none | none |
| Alam & Hamburger [22] (2001) | 6 | M | none | none | none | N/A | HE (oral) | reticular<br>reticular | buccal mucosa right side<br>dorsum of tongue | **N/A** | none | none | complete |
| Alam & Hamburger [22] (2001) | 7 | M | none | none | none | poor oral hygiene | HE (oral) | reticular + atrophic<br>erosive | buccal mucosae bilaterally<br>gingiva | **soreness** | none | chlorhexidine<br>topical steroids | none |
| Alam & Hamburger [22] (2001) | 14 | M | asthma | N/A | salbutamol and beclomethasone inhaler | N/A | HE (oral) | reticular<br>plaque-like | buccal mucosae bilaterally<br>margins of tongue bilaterally | **pain with spicy foods** | N/A | none | complete |
| Alam & Hamburger [22] (2001) | 14 | M | none | none | none | N/A | HE (oral) | reticular + atrophic + pigmentation | buccal mucosa left side | **none** | N/A | none | none |
| Scully et al. [23] (1994) | 11 | F | none | none | none | N/A | HE (oral) | erosive | margin of tongue | **soreness** | none | topical steroids | none |
| Scully et al. [23] (1994) | 10 | F | none | none | none | N/A | HE (oral) | erosive<br>erosive<br>erosive | buccal mucosae bilaterally<br>ventrum of tongue<br>floor of mouth | **N/A** | none | topical steroids<br>topical CNIs | complete |
| Borrego Hernando et al. [24] (1992) | 10 | F | IgG anti-BMZ ‡‡‡ 1:80 LPP §§§ | N/A | N/A | N/A | HE + DIF (skin) | reticular | buccal mucosae bilaterally | **N/A** | skin, vulvar mucosae | systemic steroids | complete |

| | CASE SERIES (n=8) | | REVIEW CASES (n=26) | | TOTAL (n=34) | |
|---|---|---|---|---|---|---|
| **AGE** | | | | | | |
| overall age range (years) | 9 - 17 | | 6 – 17 | | 6 – 17 | |
| mean (± sd)age | 13.5 | (± 2.73) | 10.41 | (± 3.24) | 11.18 | (± 3.40) |
| **GENDER** | | | | | | |
| males | 4 | (50%) | 11 | (42.3%) | 15 | (46.8%) |
| females | 4 | (50%) | 15 | (57.7%) | 19 | (55.8%) |
| **FAMILIAL OLP** | 3 | (37.5%) | 1 | (3.8%) | 4 | (11.7%) |
| **SYSTEMIC CONDITIONS** | | | | | | |
| **PREEXISTING MEDICAL CONDITIONS** | 7 | (87.5%) | 9 | (34.6%) | 16 | (47%) |
| **IMMUNOLOGICAL DISORDERS** | 7 | (87.5%) | 5 | (19.2%) | 12 | (35.2%) |
| **CONFIRMATORY HISTOLOGY** | 8 | (100%) | 26 | (100%) | 34 | (100%) |
| **CONCOMITANT ORAL FACTORS** | 3 | (37.5%) | 8 | (30.7%) | 11 | (32.3%) |
| **OLP CLINICAL PATTERN** | | | | | | |
| reticular | 5 | (62.5%) | 19 | (79.1%) | 24 | (70.5%) |
| papular | 0 | | 3 | (11.5%) | 3 | (8.8%) |
| plaque-like | 3 | (37.5%) | 3 | (11.5 %) | 6 | (17.6%) |
| atrophic | 3 | (37.5%) | 5 | (19.2%) | 8 | (23.5%) |
| erosive | 1 | (12.5%) | 10 | (38.4%) | 11 | (32.3%) |
| bullous | 1 | (12.5%) | 1 | (3.8%) | 2 | (5.8%) |
| mixed | 5 | (62.5%) | 10 | (38.4%) | 15 | (44.1%) |
| **OLP SITE INVOLVEMENT** | | | | | | |
| tongue | 6 | (75%) | 13 | (50%) | 19 | (55.8%) |
| buccal mucosae | 4 | (50%) | 19 | (73%) | 23 | (76.4%) |
| gingiva | 3 | (37.5%) | 3 | (11.5%) | 6 | (17.6%) |
| retromolar fossae | 1 | (12.5%) | 2 | (7.6%) | 3 | (8.8%) |
| lips | 1 | (12.5%) | 4 | (15.4%) | 5 | (14.7%) |
| palate | 1 | (12.5%) | 1 | (3.8%) | 2 | (5.8%) |
| floor of mouth | 0 | | 3 | (11.5%) | 3 | (8.8%) |
| **EXTRA-ORAL INVOLVEMENT** | 0 | | 5 | (19.2%) | 5 | (14.7%) |

| | | | | | | |
|---|---|---|---|---|---|---|
| **SYMPTOMS REFERRAL** | 4 | (50%) | 18 | (69.2%) | 22 | (64.7%) |
| **COMPLETE RESOLUTION OF ORAL LESIONS** | 2 | (25%) | 11 | (42.3%) | 13 | (38.2%) |

Table 4



Table 5

| **topic** | **childhood** | **adulthood** |
|---|---|---|
| OLP frequency | 0.03% [17] | 1-2% [2] |
| familial OLP | 1-4% [3] | 1.5% [21] |
| most common clinical pattern | reticular<br>erosive | reticular (83.5%) [2]<br>erosive (15-39%) [2,6] |
| most common oral sites involved | buccal mucosae and tongue [25] | buccal mucosae (88%)<br>tongue and gingiva (18.7%). [2] |
| involvement of both skin and oral | 12.6% [27] | 20-34%[2] |

| | | |
|---|---|---|
| mucosae | | |
| histology | no dysplasia, basal cell degeneration and band-like subepithelial lymphocytic infiltration [25] | no dysplasia, basal cell degeneration and band-like subepithelial lymphocytic infiltration [13] |
| treatment | symptomatic[6] | symptomatic [28] |
| resolution | more frequent than in adults[17] | 2-5% [29,30] |
| malignant transformation of OLP | never reported[10] | 0.4-5.3% [2] |



**Table legends**

Table 1: Flow chart

Table 2: Data on the 8 patients from our case series: epidemiology, predisposing factors, clinical features, diagnosis and treatment.

†:non-steroidal anti-inflammatory drugs ‡:hemotoxylin eosin §:hemoglobin ¶:Immunoglobulin G versus epstein-barr virus ††:autoimmune-polyendocrinopathy-candidiasis-ectodermal dystrophy ‡‡:not available

Table 3: Data on the 24 patients from the PUBMED search: epidemiology, predisposing factors, clinical features, diagnosis and treatment.

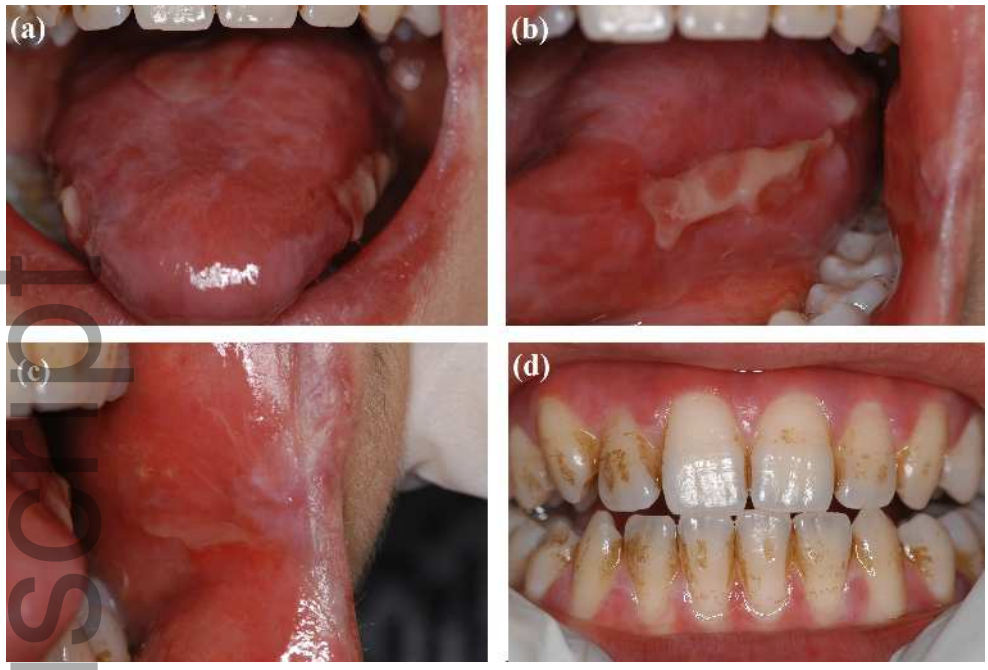
‡:hemotoxylin eosin §:hemoglobin ‡‡:not available §§:direct immunofluorescence †:non-steroidal anti-inflammatory drugs ¶¶:calcineurin inhibitors †††: antinuclear antibodies ¶¶¶:Immunoglobulin A‡‡‡:basal membrane zone §§§:lichen planus pemphigoides

Table 4: Comparison between our case series and the cases from the PUBMED search

Table 5: Comparison between OLP in childhood and adulthood

**Figure legends**

Fig 1 Case No.9 a 9-year-old patient affected by APECED.(a)bullous lesions involving the dorsum and margins of the tongue bilaterally and interlaced by reticular keratotic, erythematous and atrophic aspects of the epithelium;(b)bullous lesion involving the left margin of the tongue surrounded by reticular keratotic, erythematous and atrophic aspects of the epithelium;(c)bullous lesion involving the left buccal mucosa surrounded by reticular keratotic, erythematous and atrophic aspects of the epithelium;(d)reticular keratotic lesions of the upper and lower gingiva

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