



Case Report

Juvenile Oral Lichen Planus: A Report of 2 Cases

Victoria L. Woo, DDS¹ • Akanksha Manchanda-Gera, BA² • Derek S. Park, BA, MA³ • Angela J. Yoon, DDS, MPH, MA⁴ • David J. Zegarelli, DDS⁵

Abstract: *Lichen planus is a mucocutaneous disease that predominantly affects older patients and occurs much less frequently in the pediatric population. Furthermore, oral lichen planus is extremely rare in childhood with very few cases cited in the literature. The intention of this paper is to contribute two clinically and histologically documented cases of juvenile oral lichen planus cases to the literature. Although a rare occurrence, early recognition and diagnosis of this condition by dental practitioners can have a significant impact on the oral health of affected patients. (Pediatr Dent 2007;29:525-30) Received November 16, 2006 / Revision Accepted March 4, 2007.*

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Lichen planus (LP) is an inflammatory disease that may affect a wide variety of sites, including the skin and mucous membranes.¹ It is estimated that 50% to 70% of adult LP patients have both skin and oral lesions² and approximately 25% of patients present with oral lesions alone.³ Cutaneous LP is characterized by purple, pruritic, polygonal papules with overlying reticular striations that tend to localize on the extremities and lower back.¹ Involvement of other sites, including the scalp, nails, and nasal, esophageal, and genital mucosa, also have been described.^{1,4} In contrast to skin LP, oral lichen planus (OLP) demonstrates clinical variability. Although OLP is historically divided into 6 subgroups based on lesion characteristics⁵ (Table 1), in practice most clinicians prefer 2 clinical designations: (1) reticular OLP; and (2) erosive OLP. Reticular OLP typically presents as asymptomatic white keratoses, while the erosive form is erythematous and frequently painful.^{4,6} Both forms generally present in a bilateral, symmetrical distribution, and the disease course is characterized by periods of quiescence and exacerbation.^{1,7} The histopathology of LP shows variable hyperkeratosis, irregular rete ridge elongation, basal cell degeneration, and a band-like predominantly lymphocytic infiltrate in close proximity to surface epithelium.

Table 1. ANDREASEN'S CLASSIFICATION OF ORAL LICHEN PLANUS⁵

1. Papular
2. Reticular
3. Plaque-like
4. Ulcerative (erosive)
5. Erythematous (atrophic)
6. Bullous

Although a common disease, LP's exact cause remains unknown. An autoimmune basis has been proposed, as LP often occurs in association with other autoimmune diseases, such as lupus erythematosus,⁸ pemphigus,⁹ Sjogren's syndrome,¹⁰ autoimmune liver disease,¹¹ rheumatoid arthritis,¹ and dermatomyositis.¹ There is evidence suggesting, however, that LP is not a true autoimmune disease but rather a chronic, cell-mediated immune disorder involving activated lymphocytes and upregulated cytokine production.^{1,4} Roles for genetic predisposition, stress, and environmental factors, such as infectious agents and systemic illnesses, also have been proposed.^{1,6}

While LP is widely recognized in adults, its occurrence in children is uncommon. The exact incidence of pediatric LP is unknown, as percentages vary greatly from practice to practice. Several retrospective reviews, however, have estimated that only 1% -16% of LP patients are younger than 15 years old.^{3,12} Moreover, juvenile OLP, which is defined as OLP in patients younger than 20 years old, has rarely been documented in the medical/dental literature. Proposed factors responsible for this paucity of reports include a lack of

Drs. ¹Woo and ⁴Yoon are assistant professors and ³Dr. Zegarelli is professor, Division of Oral Pathology, and ²Ms. Manchanda-Gera and ³Mr. Park are DDS candidates, all at Columbia University College of Dental Medicine, New York, NY.

Correspond with Dr. Woo at vlw2104@columbia.edu

patient and parent awareness of lesions, and misdiagnosis or lack of recognition by practitioners.⁶ The latter can occur when poor oral hygiene is superimposed upon affected mucosa.^{6,13} Other factors responsible for the rarity of juvenile OLP include a low incidence of autoimmune diseases, systemic diseases, precipitating factors such as stress, and LP-related infections in this very young population.^{6,7}

The purposes of this paper were to report 2 children with 2 distinct clinical variants of oral lichen planus and provide a brief literature review pertaining to juvenile oral lichen planus.

Case 1

A 9-year-old Caucasian female was referred to the Division of Oral Pathology, Columbia University College of Dental Medicine, New York, NY, for evaluation of bilateral tongue lesions. She reported that her tongue lesions caused her discomfort when stimulated by certain foods and liquids. Her parents described a completely negative medical history, and she was on no medications. Questioning revealed that there were no family members with LP and no history of hepatitis B vaccination. On intraoral examination, bilateral and symmetric ulcers were evident involving the right and left lateral ventral tongue surfaces (Figure 1), each measuring approximately 9 cm². The ulcer margins showed an adherent, white reticular and papular pattern. There were no adjacent dental restorations and no similar lesions elsewhere on the oral mucosa or skin. A differential diagnosis included juvenile oral lichen planus and oral lupus erythematosus (LE). Routine blood studies were within normal limits, except for a slightly positive antinuclear antibody titer (<1:40). Further LE testing via the family pediatrician was negative. The histopathology from a right lateral tongue biopsy revealed lichenoid mucositis consistent with lichen planus (Figure 2). The patient was placed on a beclomethasone dipropionate inhaler and dexamethasone elixir and advised to return every 6 months for periodic evaluation. Despite repeated efforts to contact the patient, however, she has not returned for follow-up.



Figure 1. Clinical photograph depicting a large ulcer with white, reticular margins.

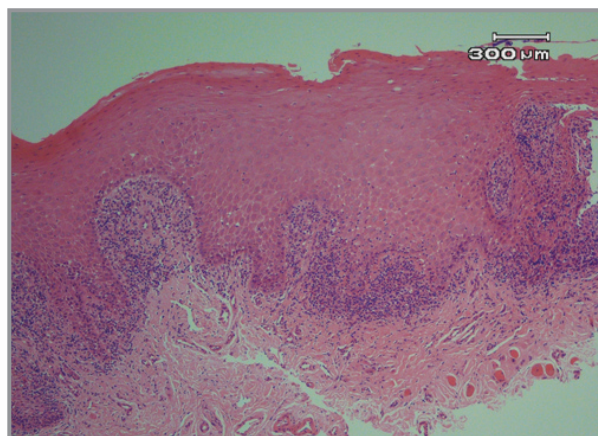


Figure 2. Medium power photomicrograph depicting a band-like infiltrate of lymphocytes subjacent to hyperkeratinized epithelium (hematoxylin and eosin, original magnification X 100).

Case 2

An 11-year-old Caucasian female presented to the Division of Oral Pathology, Columbia University College of Dental Medicine, New York, NY, for evaluation of bilateral, asymptomatic mucosal tongue lesions. As with the former patient, she had a completely negative medical history, was on no medications, and had no family members with LP. Interestingly, the patient had received 3 hepatitis B vaccinations at 1 month, 2 months, and 6 months of age. On intraoral examination, classic white, lacy striations were noted on the patient's right and left lateral tongue surfaces and left posterior buccal mucosa (Figure 3). The patient had metal orthodontic brackets that did not contact the lesions. There were no adjacent dental restorations and no similar lesions elsewhere in the mouth or on the skin. Due to the oral lesions' classic appearance, a diagnosis of juvenile oral lichen planus was strongly suspected. A confirmatory biopsy of the right lateral tongue was read as lichen planus (Figure 4). Since the patient was asymptomatic, no further treatment was given at that time. Follow-up 1 year later revealed similar asymptomatic lichen-



Figure 3. Clinical photograph depicting classic white, lacy lesions.

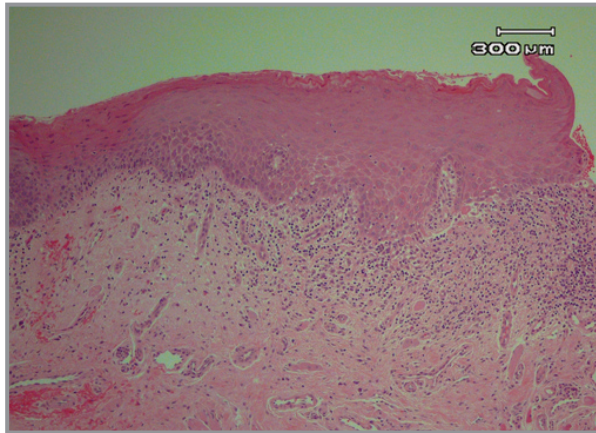


Figure 4. Medium power photomicrograph depicting a lichenoid infiltrate subjacent to hyperkeratinized epithelium (hematoxylin and eosin, original magnification X 100).

oid lesions involving bilateral tongue and buccal mucosa. It also was noted that the patient's orthodontic brackets had been removed. No additional treatment has been given and the patient continues to return for annual evaluations.

Discussion

There is very little dermatology literature on the subject of juvenile lichen planus, and even fewer reports in the dental literature. We performed an extensive literature search for juvenile/childhood OLP to summarize this illness' pertinent demographic and clinical features. The inclusion criteria for our summary are presented in Table 2.

Table 2. INCLUSION CRITERIA FOR SUMMARY OF JUVENILE ORAL LICHEN PLANUS STUDIES

1. Less than or equal to 20 years old.
2. Clinical evidence of oral lichen planus (OLP).
3. Oral biopsy confirmation of lichen planus, lichenoid lesion, or lichenoid mucositis.
4. If no oral biopsy was performed, a clinical description of "reticular" or "reticulate," "striae," or "striated," and/or "lacy" oral lesions was required (considered to represent reticular OLP).
5. No evidence of mucosal contact with dental restorative materials, no exposure to medications known to induce oral lichenoid reactions, and no documented history of graft-versus-host disease.

Our search yielded 18 studies, 10 of which provided the demographic data and clinical information we were seeking (Table 3).^{2,6,7,13-19} Five additional studies were not included in the summary but are noteworthy; 4 reported oral involvement in 5 pediatric patients with biopsy-proven dermal LP.^{12,20-22} Specific clinical descriptions of the oral lesions were not provided, however, and oral biopsies were not performed. The fifth study described 9 OLP children, 6 of whom exhibited white "lacy" lesions of the buccal and/or gingival mucosa.²³ Several of the children in this series, however, had

preceding upper respiratory tract infections, and oral biopsies were not performed.

Thus, our literature review from 1990 to 2005 yielded 42 juvenile OLP patients who fulfilled the aforementioned criteria. Table 4 summarizes the pertinent findings of these cases. The nature of the review articles precluded their inclusion in the gender, age, and ethnicity summaries. Juvenile OLP occurred slightly more frequently in male patients (60%), particularly those 11 and 15 years old (60%). There was no ethnic predilection noted. The buccal mucosa was the most commonly affected site (55%), although synchronous involvement of 2 or more sites (usually the buccal mucosa and tongue) was frequently observed. Most patients were affected by reticular OLP (64%), although it was not unusual to see 2 or more types occurring in a single patient.

According to previous studies, the incidence of pediatric LP among all LP patients is low. Furthermore, the rarity of oral involvement in pediatric LP patients is even more noteworthy. Kumar et al reported OLP in 1 of 25 LP children (4%); this patient also had 20-nail dystrophy.³ Similarly, Luis-Montoya et al documented oral lesions in 1 of 16 LP children (6%).¹² Handa and Sahoo studied 87 LP children, 12 (14%) of whom demonstrated oral findings;¹¹ had both skin and oral involvement, and 1 had oral involvement only.² Sharma and Maheshwari⁸ reported OLP in 15 of 50 LP children (30%), and Nanda et al²³ found OLP in 9 of 23 LP children (39%). The rare occurrence of juvenile OLP also is apparent when large cohorts of OLP patients of all ages are analyzed. Xue et al presented 674 patients with OLP; 4 (<1%) were children between 10 and 13 years old.¹⁹ Similarly, Eisen evaluated 723 OLP patients and only 5 (<1%) were children younger than 15 years old.⁷ All 5 had atrophic and erosive OLP, and all developed dermal LP within 2 years of oral onset. Of note, Eisen reported that each patient was initially misdiagnosed and treated incorrectly for other oral conditions, such as herpes simplex, candidiasis, and recurrent aphthous stomatitis.

Only 1 study has documented a gender predominance among juvenile OLP patients. Alam and Hamburger described 6 OLP patients, all males ranging from 6 to 14 years old.¹³ Based on their series, these authors proposed a possible male predilection for juvenile OLP.

Although reporting bias may be a contributing factor, there appears to be an increased incidence of juvenile OLP in patients from India, China, the United Kingdom, and Italy. A predominance of juvenile OLP in these geographic regions is of interest and suggests that environmental factors and/or genetics influence disease evolution. To date, there have been six studies originating from India documenting juvenile OLP cases.^{2,3,15,16,18,20} With the exception of Eisen's study⁷ and our current report, to our knowledge there have been no additional juvenile OLP studies originating from the United States.

Table 3. SUMMARY OF JUVENILE OLP STUDIES

Author	No. of Cases	Gender/ Age	Ethnicity	Location	Clinical type	Biopsy confirmed	Comments
CASE REPORTS							
Scully et al ¹⁴	2	F/10	Caucasian	FOM*	Erosive	Y	
		F/11	Caucasian	Tongue	Erosive	Y	
Alam and Hamburger ¹³	6	M/6	Asian	BM	Reticular	Y	
				tongue	Erosive		
		M/7	Asian	BM	Atrophic	Y	
		M/8	Caucasian	Gingiva	Reticular	N	“lichenoid”, “striae”
		M/11	Caucasian	BM	Reticular	N	“classical reticular”
				tongue	Papular		
		M/14	Asian	BM	Reticular	Y	
				tongue	Plaque-like		
Sandhu et al ¹⁵	1	F/12	NS	NS	Atrophic	Y	Familial
Singal ¹⁶	1	M/11	NS	Tongue	Atrophic	Y	Familial
Patel et al ¹⁷	2	M/6	Caucasian	Tongue	Plaque-like	Y	
		F/15	Caucasian	Tongue	Erosive	Y	
Laeijendecker et al ⁶	3	F/11	Asian	BM	Reticular	Y	
		F/14	Caucasian	BM	Reticular	Y	
				tongue			
		M/16	Asian	BM	Erosive	Y	
				gingiva			
REVIEW STUDIES							
Sharma and Maheshwari ¹⁸	10	NS/≤14	NS	BM	Reticular	Y (skin)	“classic lacy”
Eisen ⁷	5	NS/<15	NS	NS	Erosive, atrophic	Y	
Handa et al ²	8	NS/7-11.5	NS	BM (5)	Reticular	Y	“reticulate”
				Lips (3)			
Xue et al ¹⁹	4	NS/10-13	NS	NS	Reticular (2) Erosive (2)	Y	

* FOM = floor of mouth; BM = buccal mucosa; NS = not specified

Many illnesses and conditions associated with LP occur in older patients. Thus, explaining the occurrence of OLP in the younger population is a challenge. Important factors in the development of juvenile OLP include: (1) previous hepatitis B vaccination^{24,25}; (2) liver disease, including chronic active hepatitis²⁶; and (3) genetic predisposition, such as in

familial LP.¹ A familial history of LP deserves brief discussion, as it has been regarded as a relevant predisposing factor in pediatric patients. Milligan and Graham-Brown²⁷ reported a family history of LP in 1% to 2% of their juvenile OLP patients, whereas Cottoni et al²⁶ found 1 of 5 (20%) of their juvenile OLP cases had a positive family history. Singal documented OLP in 1 family over 3 successive generations: an 11-year-old Indian boy, his father, and his grandmother.¹⁶ Based on this report, the author suggested an autosomal dominant basis for familial LP. Mahood found that 12% of his familial LP patients manifested the disease before age 10.²⁸ Other notable features of familial LP include a higher incidence of oral lesions, frequent clinical relapses,¹⁵ and increased disease severity.²⁷ Interestingly, reticular OLP is the most common form overall, while erosive and ulcerative OLP tend to predominate in familial cases.¹⁵ A number of human leukocyte antigen types also have been associated with familial LP.^{15,28-30}

Differences in clinical presentation of adult and juvenile LP have been observed. Several authors have noted that LP in children may exhibit atypical features, such as a “linear” pattern not commonly seen in adults.^{2,6,13,21} Regarding OLP, it appears that the erosive form is relatively rare in children—as opposed to the adult population, in which erosive OLP is estimated to affect 39% of patients.¹⁹ A lack of exacerbating factors more commonly seen in adults, including periodontal disease,³¹ trauma from poor-fitting prostheses, irritation from dental plaque and calculus, increased stress, and contact with certain foods,^{4,7} may contribute to the low inci-

Table 4. SUMMARY OF DEMOGRAPHIC AND CLINICAL DATA OF JUVENILE ORAL LICHEN PLANUS CASES

Variable	Total no. of patients	No. of cases (%)
Gender	15	
Male		9 (60)
Female		6 (40)
Age* (ys)	15	
6-10		5 (33)
11-15		9 (60)
16-20		1 (7)
Ethnicity	15	
Caucasian		7 (47)
Asian		6 (40)
Unspecified		2 (13)
Lesion location †	42	
Buccal mucosa		23 (55)
Tongue		8 (19)
Lips		3 (7)
Gingiva		2 (5)
Floor of mouth		2 (5)
Unspecified		10 (24)
Clinical type †	42	
Reticular		27 (64)
Erosive		12 (29)
Atrophic		8 (19)
Plaque-like		2 (5)
Papular		1 (2)

* 27 patients from review studies excluded.
 † Several patients exhibited >2 sites of involvement or OLP types.

Table 5. COMMONLY USED TOPICAL STEROIDS IN ORAL LICHEN PLANUS TREATMENT

Dexamethasone elixir (0.5 mg/5 ml) Rinse and expectorate 1 teaspoon 2-4x/day as needed.
Beclamethasone spray (40 mg) Take 1-2 puffs 1x/day as needed.
Fluocinonide gel (0.05%) Apply to affected area 2x/day as needed.

dence of erosive OLP in children. The rarity of this form likely explains why most juvenile OLP patients are asymptomatic.^{2,6,17}

Treatment of juvenile OLP does not differ significantly from treatment of adult OLP. Pharmacologic treatment is often unnecessary in asymptomatic patients. For symptomatic lesions, topical corticosteroids are the most commonly used

agents (Table 5). The patient and parents should be informed, however, that chronic use of topical steroids can lead to oral candidiasis. Systemic steroid therapy and dapsone are typically reserved for refractory and recurrent cases.^{21,23} Extreme caution is taken when these agents are used, as significant long-term effects are of concern in this young patient population.¹⁸ Of note, tacrolimus ointment, topical tretinoin, and topical cyclosporine also have been used with success in some cases.^{1,6,15} Periodic follow-up is required in all OLP patients, typically every 6 months to every year. This is especially important in the pediatric population as malignant transformation has been described in a small percentage of adult OLP cases in follow-up studies.^{1,4,16}

In summary, we reported 2 children with clinically and microscopically documented oral lichen planus. Our first patient had predominantly erosive OLP, a rare finding in the pediatric population. Our second patient had classic reticular OLP and, interestingly, a positive history of hepatitis B vaccination. We do acknowledge that long-term follow-up information could not be provided for our first patient. Thus, the effectiveness of our treatment could not be assessed. The dental literature supports that, in young patients with suspected OLP, eliciting a family history of LP and prior hepatitis B vaccination is indicated. Additionally, inquiring about systemic medication use and careful examination for contacting dental restorative materials is recommended to rule out a lichenoid mucosal reaction. Clinicians must be aware that OLP children also may have simultaneous or future involvement of skin and other mucosal sites; if lesions are reported elsewhere, appropriate referrals are necessary. The asymptomatic reticular variant of OLP appears to predominate in children. Therefore, pharmacologic treatment is often not necessary. In symptomatic patients, good oral hygiene should be encouraged as a means of reducing irritating factors such as plaque and calculus.^{1,7,31} The prognosis of juvenile OLP is unclear at this time, as long-term studies have not been published. Although Laeijendecker et al⁶ reported no OLP-related malignancies to date in the pediatric population, it appears that careful follow-up of all OLP patients is warranted.

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