Clinical Features, Diagnosis and Management of Oral Lichen Planus in Children

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ABSTRACT

Lichen planus in children is chronic autoimmune disease, affecting the skin and mucous membrane. An immune mediated pathogenesis is recognized in lichen planus although the exact immunology is unknown. The prevalence of oral lichen planus in children is rare. In this paper, etiopathogenesis, features, management of oral lichen planus is reviewed.

Keywords: Oral lichen planus, Children, Wickham's striae, Corticosteroids, Retinoid.


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INTRODUCTION

Lichen planus is a chronic inflammatory mucocutaneous disease, first described in the literature by Erasmus Wilson in 1869.1-3 It is predominantly a disease of the middle aged or older, and has prevalence of 0.5 to 2% in adults.4 It is more common in females than in males ratio of approximately 2:1. Oral lichen planus (OLP) is also seen in children, although it is rare, and they may present with atypical findings. Childhood lichen planus is more common in the tropics and that children of Asian origin may be prone to the condition. There is limited literature available reporting the occurrences of oral LP in children. The exact incidence of pediatric LP is unknown. In view of the prevalence of OLP and the potential of this chronic disease to cause significant discomfort, it is important for clinicians to be aware of its clinical presentation and management.

ETIOLOGY

Although the etiology and pathogenesis of OLP are not fully understood, OLP has been associated with multiple disease processes and agents, such as viral and bacterial infections, autoimmune diseases such as ulcerative colitis, myasthenia gravis, lupus erythematosus, medications, vaccinations and allergy to dental restorative materials such as gold and amalgam, local trauma (Koebner phenomenon) and systemic disease, such as diabetes mellitus and hypertension (Griinspan syndrome).5 The association between OLP and chronic liver disease is still controversial.6 It was first suggested by Mokni et al in 1991. Carrozzo et al have demonstrated a strong association between hepatitis C viral infection and OLP.7

Recently, several studies have reported a relationship between Helicobacter pylori and OLP.8 Moravvej et al in 2007 found statistically significant differences in H pylori infection between patients with lichen planus and a control group. Numerous studies have investigated the prevalence of candidal infection in erosive lichen planus.9 Recently, a study was carried out on 50 patients with OLP and 35 healthy patients to investigate presence of candida; no significant difference was reported between the two groups. According to another study, there were no significant differences between the 21 healthy individuals and 21 patients with erosive lichen planus regarding candida presence. However, candida infection is not currently considered an etiologic factor for OLP.

Exacerbations of OLP have been linked to periods of psychological stress and anxiety. In contrast, Humphris and Field and MacLeod reported no statistically significant association between OLP and anxiety.5

PATHOGENESIS

The pathogenesis of lichen planus is not completely understood but a T-lymphocyte infiltrate suggests cell-mediated immunological damage to the epithelium. Modified Langerhans’ cells and keratinocytes possibly trigger an immune response and the recruitment of T-lymphocytes, encouraged by expression of cell-surface adhesion molecules. Both CD4 (helper) and CD8 (cytotoxic) cells are present but increasing numbers and activation of the CD8 cells is thought to contribute to the characteristic damage to the basal epithelium. Childhood lichen planus has been documented as a complication of hepatitis B vaccinations (HBV) where the recombinant proteins of the HBV vaccine, specially the viral S epitope, may trigger a cell-mediated autoimmune response targeted at keratinocytes giving rise to a lichenoid reaction.10-12 It is also found in association with predisposing conditions, such as graft vs host disease and chronic active hepatitis C. Studies of children with mucocutaneous lichen planus have shown a very low incidence of oral involvement.13

CLINICAL FEATURES

OLP in childhood was 1st described in 1920. The family history of lichen planus is more commonly positive in
patients with lichen planus in childhood than in adulthood. The prevalence of oral lichen planus in children is about (0.03%). The low prevalence can be partially be explained by a low number of associated systemic diseases, autoimmune phenomenon, infection (hepatitis B and C virus), drugs and dental restorations in children, which may reduce the risk for developing OLP. The diagnosis of childhood OLP can sometimes be missed due to irregular dental check ups, lack of symptoms, and ignorance.

Mucosal lesions are classified into six clinical forms, described by Andreason: reticular, papular, plaque (White Forms), atrophic, erosive, and bullous (Red Forms). Atrophic, erosive form is malignant in nature. No malignant transformation of OLP has yet been been reported. In the majority of patients with OLP there is no associated cutaneous lichen planus or lichen planus at other mucosal sites. This may be called ‘isolated’ OLP. The most commonest type is reticular pattern which is seen as fine white striae known as ‘Wickhams striae’. It is usually bilateral and symmetrical in nature. Patients with reticular lesions are often asymptomatic, but atrophic (erythematous) or erosive (ulcerative) OLP is often associated with a burning sensation and pain, and aggravated during eating and drinking. Atrophic OLP may appear as a mixture of clinical subtypes like for, e.g. white or gray streaks which may form a linear or reticular pattern on an erythematous background. Erosive OLP can present as an irregular erosion or ulceration covered with pseudomembrane or a fibrinous plaque. Periphery of the lesion is usually surrounded by reticular or finely radiating keratotic striae. When erosive OLP involves the attached gingival tissue, it is called desquamative gingivitis. The lesions of erosive OLP migrate over time and tend to be multifocal. Patients with this form of OLP often present with symptoms ranging from episodic pain to severe discomfort that can interfere with normal masticatory function. Plaque type OLP appears as an homogeneous white patches which resemble leukoplakia, which usually affects dorsum of the tongue and buccal mucosal. It may also be slightly elevated or smooth or slightly irregular in form. It is usually seen in tobacco smokers. The papular type of oral lichen planus is rarely seen. This is usually seen as small white raised papules with white striation seen at the periphery of the lesion. Bullous type of lichen planus is the least common type of OLP. Usually the diameter of bullae is few millimeter to few centimeter, which tends to rupture leaving behind ulcerated and painful surfaces. The periphery of the lesion is usually surrounded by reticular or finely radiating white keratotic striae. Oral lichen planus is chronic, usually persists for several years with periods of exacerbation and quiescence. During the periods of exacerbation, the areas of erythema or erosion increases with slight increase in pain and sensitivity. During the period of quiescence, the area of erythema or erosion decreases, with decreased pain and sensitivity. Usually exacerbations are associated with psychological stress, anxiety and mechanical trauma (Koebner phenomenon).2

HISTOPATHOLOGY

The histology of OLP is characterized by a dense subepithelial lymphohistiocytic infiltrate, increased numbers of intraepithelial lymphocytes and degeneration of basal keratinocytes. Degenerating basal keratinocytes form colloid (civatte, hyaline, cytoid) bodies that appear as homogeneous eosinophilic globules. The ultrastructure of colloid bodies suggests that they are apoptotic keratinocytes and recent studies using the end-labeling method demonstrated DNA fragmentation in these cells. Epithelial basement membrane changes are common in OLP and comprise breaks, branches and duplications. In addition, the basal keratinocyte anchoring elements (hemidesmosomes, filaments and fibrils) are disrupted. Degeneration of basal keratinocytes and disruption of the epithelial basement membrane and basal keratinocyte anchoring elements produce weakness at the epithelial-connective tissue interface which may result in histological cleft formation (Max-Joseph space) and clinical blistering of the oral mucosa (bullous lichen planus). Parakeratosis, acanthosis and ‘saw-tooth’ rete peg formation may be seen. B-cells and plasma cells are infrequent in OLP and immunoglobulin and complement deposits are not a consistent feature. The presence of a mixed and sometimes more diffuse infiltrate should alert the pathologist that the condition may be drug related or perhaps lichenoid, rather than true idiopathic lichen planus. Some cases show fibrinogen and fibrin deposition in a linear pattern in the basement membrane zone. Colloid bodies may be positive for fibrin, IgM, C3, C4 and keratin. Laminin and fibronectin staining may be absent in areas of heavy fibrin deposition and colloid body formation, suggesting basement membrane damage in these areas. Immunofluorescent findings in OLP are not diagnostic.13,14

DIAGNOSIS

A complete history and physical examination by a multidisciplinary group of healthcare providers may be required to investigate oral, skin, nail, scalp, genital, esophageal, laryngeal and conjunctival involvement. The history, typical oral lesions and skin or nail involvement are usually sufficient to make a clinical diagnosis of OLP. However, biopsy is required to differentiate between OLP and other chronic white or ulcerative oral lesions including...
reactive keratoses, chronic hyperplastic candidosis, epithelial dysplasia, discoid lupus erythematosus, gastrointestinal disease (including oral Crohn’s disease) or anemic states. Direct immunofluorescence can help distinguish erosive, ulcerative or the very rare bullous form of OLP from pemphigus vulgaris, benign mucous membrane pemphigoid, dermatitis herpetiformis and linear IgA bullous dermatosis. There are no consistent serological changes associated with OLP but some patients do present an elevated ANA titer.13,14

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of OLPc may be quite extensive and, of course, depends on the age of the patient, the clinical variant of OLPc, and the severity and the persistence of the lesions and includes candidosis, morsicatio buccarum, leucoplaikia, lingua geographica, autoimmune bullous diseases, lupus erythematosus, several viral infections (herpes simplex, Epstein–Barr, Coxsackie, HIV), recurrent aphthous stomatitis (caustic) traumata, erythema multiforme (major), allergic gingivostomatitis, gluten sensitivity enteropathy, and less commonly Crohn and Behçet diseases, oral lesions in immunodeficiencies, dyskeratosis follicularis, pachyonychia congenita, dyskeratosis congenita, and white sponge nevus.

MANAGEMENT

OLP occurs more frequently than the cutaneous form and tends to be more persistent and more resistant to treatment. Topical corticosteroids are the mainstay in treating mild to moderately symptomatic lesions. Options (presented in terms of decreasing potency) include 0.05% clobetasolpropionate gel, 0.1 or 0.05% betamethasone valerate gel, 0.05% fluocinonide gel, 0.05% clobetasol butyrate ointment or cream, and 0.1% triamcinolone acetonide ointment.5,13,15

Patients are instructed to apply a thin layer of the prescribed topical corticosteroid up to 3 times a day, aftermeals and at bedtime. The gel or ointment can be prescribed topical corticosteroid up to 3 times a day, but adverse effects are possible even with short courses. Topical corticosteroids are widely used in the treatment of OLP to reduce pain and inflammation. Triamcinolone acetonide is commonly used either in Orabase or as lozenges. An oral suspension of triamcinolone has also been used with beneficial effects.15

OTHER APPROACHES

Twice-daily topical application of compounded 0.1% tacrolimus ointment was recently reported to be effective in controlling symptoms as well as clearing lesions of OLP. Tacrolimus is a macrolide immunosuppressant with a mechanism of action similar to that of cyclosporine, but it is 10 to 100 times more potent and is better able to penetrate the mucosal surface. Treatment with topical tacrolimus 0.1% ointment four times daily induced a better initial therapeutic response than triamcinolone acetonide 0.1% ointment in patients with symptomatic OLP.17

RETINOIDS

Retinoids are metabolites of vitamin A. They have been noted to have antikeratinizing and immunomodulating effects retinaldehyde 0.1%, isotretinoin gel 0.1% have been tried in OLP and they showed good clinical efficacy. OLP has been treated with fenretinide and tazarotene gel 0.1% successfully. These studies suggested that topical retinoid might be a suitable therapeutic agent in the treatment of hyperkeratotic OLP.4,5,13

ULTRAVIOLET IRRADIATION

Photochemotherapy with 8-methoxypsoralen and long-wave ultraviolet light (PUVA) has been used successfully in the treatment of skin lesions and cutaneous lichen planus. Some studies have indicated that PUVA therapy might also have therapeutic effects. To avoid PUVA side effects, photosensitization with topical 0.01% trioxsalen can be used for the treatment, although oral mucosa seems more resistant to phototoxic damage in comparison to skin. PUVA with
8-methoxypsoralen has various side effects, such as nausea, dizziness, eye symptoms, paresthesia, and headache. Moreover, one matter of concern is that PUVA therapy has been shown to have oncogenic potential. Further studies on PUVA therapy for OLP are needed. LASER THERAPY

In 2004, Barclay used 308 nm excimer laser radiation in OLP patients. This technique was suggested as a viable, palliative treatment option with high patient acceptance in those with symptomatic OLP. In 2005, Soliman et al used diode laser (980 nm) in the management of OLP as an easy, fast and safe technique. It could be used in an outpatient clinic with local anesthesia. In 2008, van der Hem et al found good results in the treatment of OLP lesions with CO2 laser evaporation. In this study, there were no problems with wound healing. In every case there was complete epithelialization within 3 weeks. They found out that when there is no further improvement with steroids, CO2 laser evaporation seems to be a good treatment option for OLP and may even be considered a first choice. DISCUSSION

There is a limited literature available reporting the occurrences of OLP in children. Studies of children with mucocutaneous lichen planus have shown a very low incidence of oral involvement. Kumar et al in a series of 25 children with cutaneous lesions, reported only a single patient with oral mucosal lesions and Kanwar et al described only 1 patient out of 17 with mucosal lichen planus involving the lips. Familial lichen planus has been reported as being uncommon. Milligan reported a family history present in 1 to 2% of cases. Childhood familial lichen planus is said to occur at an early age and with greater severity. It is not possible to draw firm conclusion about many aspects of OLP especially associated factors, treatment, prognosis, age of preference, and the boy to girl ratio, because of the low numbers of children affected with OLP. Childhood lichen planus is usually atypical with a positive family history often has asymptomatic lesions. The treatment in symptomatic OLP most commonly consists of local regimens because of both general improvement and possible side effects of systemic therapy. The schedule of follow-up of OLP in children should be at least 1 to 2 visits/year as long as OLP persists and even more frequently in symptomatic OLP.

CONCLUSION

OLP is rare in children and therefore at present it is not possible to draw conclusions about this disease, though the clinical presentations resemble those of adult OLP. However, generally the prognosis of OLP in childhood seems to be more favorable. Clinicians are invited to submit the cases of OLP in children in order to develop consensus on the clinical presentation, diagnosis and its management.

REFERENCES

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