Review Article

Management Of Childhood Lichen Planus

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Abstract

Childhood lichen planus (LP) is a rare entity, with less than 2–3% of all cases seen in patients under 20 years of age. LP in childhood is common in subtropical countries such as India. The most common clinical type of LP in Indian children is the classic form. Approximately 1–15% of patients with LP demonstrate nail involvement, but disease of the nails without skin involvement is rare. LP is diagnosed by history, clinical findings, biopsy results, and, in some cases, features on direct immunofluorescence (DIF). LP tends to have a chronic course. Depending on disease severity, however, LP may respond to a combination of topical or systemic therapies. The response to therapy may be similar to that seen in adults. Moderately potent or super potent steroids are the treatment of choice. Topical steroid can be combined with oral steroid in tapering doses over 2-12 weeks period. This is useful for children with widespread involvement or cutaneous LP lesions associated with significant morbidity. Intralesional steroid is effective for hypertrophic LP unresponsive to topical steroid. Topical steroid in adhesive base used several times a day for several months is a treatment of choice for symptomatic oral LP. Topical steroid in combination with systemic steroid can be given in a tapering dose over 3-6 weeks in very symptomatic cases in early stages. In severe unresponsive cases of both cutaneous and oral LP, oral retnoid are the preferred option. Treatment options for the nail LP in young children are oral steroid given as tapering dose over 4-12 weeks and oral retinoid. Intralesional steroid as nail matrix injection are the third option for older children. Most pediatric patients with LP respond to treatment with full clearance over 1-6 months. Poor response to treatment is a feature of hypertrophic LP and lichen planopilaris.

Key words

Children, Lichen planus, Nail lichen planus, Oral lichen planus

Introduction

Lichen planus is a pruritic papulosquamous dermatoses of unknown etiology characterized by presence of violaceous, polygonal, flat-topped papules, affecting the skin, nails and mucous membranes. It usually resolves after a variable period of time leaving behind residual areas of hyperpigmentation.

Epidemiology

Lichen planus, mainly a disease of adults, affecting upto 1% of the general adult population,[1] is only rarely encountered in children with less than 5% of cases occurring in childhood.[2] Though the world wide prevalence of childhood lichen planus is rare, for reasons unknown, it is not that uncommon in India, based on the fact that the largest case series on childhood LP are published

from India.[3-6] In a study from UK also it was found that children originating from the Indian subcontinent represented 80.8% of children with lichen planus suggesting that children from South Asian origin are more susceptible to developing LP.[7] This could be attributed to the difference in the genetic background and environmental triggers or underreporting in other areas. The rarity of associated autoimmune conditions, exposure to drugs and dental restorative materials, infective agents and other environmental triggers that have been known to initiate lichen planus and the overall rarity of LP in children may be responsible for the overall rarity of LP in children. Maximum proportion of children had disease onset between 5 and 9 years with a mean age at onset of 7.1-8.4 years. The youngest documented case is a three-month-old child and the reported earliest age at onset has been two weeks.[3-6,8] Majority of the studies, either the sexes were equally affected or there was marginal male preponderance.[4] Familial LP occurs in 1-2% of all childhood cases and these differ from the classical form clinically, with earlier age at onset, more generalized involvement, more common mucosal involvement and an increased tendency for erosive, ulcerative and linear forms, with prolonged course and frequent relapses.[4]

Etiology

LP is a T-cell-mediated autoimmune damage to basal keratinocytes, which have been altered by viruses, medications, or other unknown allergens. Several exogenous antigens thought to trigger LP in children include infections like hepatitis C infection or HHV 7 infections, vaccination like hepatitis B vaccination, DPT, MMR, polio vaccine, dental amalgam fillings and drugs. Autoimmune diseases such as chronic active hepatitis, primary biliary cirrhosis, autoimmune thyroiditis, myasthenia gravis, alopecia areata, thymoma, celiac disease, atopic dermatitis and autoimmune polyendocrinopathy have been reported to coexist with lichen planus.[7,9,10]

Clinical features

The primary lesion of lichen planus is a violaceous, flat topped, polygonal, pruritic, papule, and represents commonest among all the morphologies of lichen planus in all age groups. Classical LP was the most common variant observed in all the reported studies (42-76%).[3-9] The second most common variant differed between studies: lichen planus hypertrophicus (12%), actinic LP (11.5%), eruptive LP (13%). Actinic LP is common in tropical and sub-tropical countries including India. Initially linear LP was thought to be more common in children as compared to adults, but recent studies have shown results on the contrary. Postinflammatory hyperpigmentation is considered to be more intense in childhood LP. Koebner's phenomenon is considered to be common in children with LP, varying between 24 and 28%. Scalp involvement is rare (0-9%) in children. Lichen planus can be associated with or appear after generalized lichen nitidus. Nail involvement is rare in children and occurs upto-8.7% of patients. Longitudinal ridging is the most common finding followed by pitting, thinning of nail plate, patients, trachyonychia, discoloration, nail dystrophy, subungual hyperkeratosis, onycholysis, nail splitting, thickening of nail plate and leukonychia. Nail lichen planus in children can have two peculiar clinical features - twenty nail dystrophy and idiopathic atrophy of the nails apart from the other classical features. Nail LP in children is under estimated due to lack of skin and mucosal lesions which makes clinical diagnosis difficult and due to the reluctance to perform nail biopsies in children.[11,12]

The reported prevalence of oral LP in childhood is 0.03%, significantly lower than that seen in adults. This difference can be attributed to the lesser association with systemic diseases, autoimmune phenomena, drugs and dental restorations in childhood. Moreover, as the onset of oral LP is insidious, the diagnosis may be

missed because of irregular dental visits, a low level of symptoms and ignorance. Oral lichen planus in childhood seems to occur preferentially in those of Asian race. The clinical features resemble those of oral lichen planus in adults. However, generally the prognosis of oral lichen planus and the effect of treatment in childhood seems to be more favorable than in adults.[13-15]

Differential diagnosis

LP can be a reaction pattern to various exogenous antigens.[9,10] Thus, identifying potential triggers such as viruses, medications, vaccinations, immunizations and dental materials is important, before labeling the condition idiopathic LP. Other diseases that resemble LP include lupus erythematosus, lichen nitidus, lichen striatus, lichen sclerosis, pityriasis rosea and psoriasis.

Investigations

Lichen planus is LP is predominantly a clinical diagnosis based on the history and typical clinical features. However, histopathology and sometimes DIF may be required to confirm the diagnosis in doubtful cases.[9,10]

The classical histopathological changes include hyperkeratosis, wedge-shaped hypergranulosis, variable acanthosis with saw-toothing of the rete ridges, band-like lymphohistiocytic infiltrate at thedermal–epidermal junction, basal layer damage causing Caspary–Joseph spaces, Civatte bodies and colloid bodies, variable melanin incontinence.[9,10]

Direct immunofluorescence of biopsy specimen from either skin or mucous membrane reveals shaggy fibrin deposit at the dermo-epidermal junction and colloid bodies.[9,10]

Nail biopsy is not required in every child with nail LP where clinical manifestation is characteristic like nail plate thinning with longitudinal ridging and fissuring with or without pterygium. The presence of skin lesions along with nail changes suggestive of nail LP such as trachyonychia or idiopathic atrophy of the nails does not require nail biopsy.[4,12]

Treatment

Lichen planus may be completely asymptomatic or associated with significant pruritus.[17,18] In either cases, and even if it subsides spontaneously or is not associated with symptoms, post-LP, violaceous or brownish hyperpigmentation may persist for a long time and can be associated with significant cosmetic disability, particularly in pigmented races. Therefore, early aggressive treatment of LP is reasonable approach rather than waiting for spontaneous subsidence to effectively suppress pruritus and to quickly restitute quality of life. The recommended treatment goal for most cases of lichen planus is controlling the severity of symptoms because there is no established cure. Depending on disease severity, LP may respond to a combination of topical or systemic therapies and the response to therapy may be similar to that seen in adults.[7,18]

Cutaneous lichen planus

Topical corticosteroid are the first line of treatment in localized classic form of lichen planus but, till date, no consensus exists regarding standardized therapy regimens for exanthematous LP in childhood.[4,5,8-10,16,17]

Topical corticosteroid (class II-IV) with or without occlusion along with antihistamines is the standard therapy for childhood lichen planus.[4,5,8-10,16,17] The parents should be forewarned that occlusion increases the steroid strength and cautioned about the steroid side effects. They should be reassured that pruritus may require 3 weeks of therapy to subside and the lesions themselves take 6 weeks to begin to flatten. Intralesional steroid like triamcinolone, or class I topical steroid under occlusion can be used in hypertrophic lichen planus. Topical calcineurin inhibitor (tacrolimus 0.03% ointment) have been reported to be efffective in cases recalcitrant to

topical steroid and for some cases of refractive oral LP.

Short courses of systemic steroid (prednisone 1mg/kg/day) over 2-6 weeks can be used in severe, extensive and recalcitrant cases to ameliorate associated pruritus and hasten clearance.[4,5,8-10,16,17]

Topical steroid can be combined with systemic steroids in very symptomatic cases in early stages. This combination is also useful for children with widespread involvement or cutaneous LP lesions associated with significant morbidity like unresponsive hypertrophic LP, follicular LP on the scalp with scarring alopecia and bullous LP. In severe unresponsive cases of both cutaneous and oral LP, oral retinoid (acitretin 0.5-1mg/kg) for 8 weeks are the preferred option, but the side effects like premature epiphyseal closure needs to be taken into account.

Dapsone was found to be a safe and effective alternative in controlling chronic and recurrent disease. With the risk of premature epiphyseal closure and other systemic side effects associated with systemic steroid and retinoid, dapsone can be considered cheaper, easily available and safe alternative.

LP is not as chronic or relapsing as psoriasis and thus requires only lower doses of methotrexate for shorter duration making it an effective alternative to oral corticosteroids in the treatment of generalized LP. In a recent study, two children with LP have been treated with methotrexate (0.25 mg/ kg/week) and showed good response.[18] Phototherapy with narrowband or broadband UVB or PUVA may be used in older children or in generalized, severe, resistant cases. PUVA is contraindicated in children less than 11 years of age and NBUVB is a safe and effective option. Studies on phototherapy in lichen planus have encouraging results and have found that the complete response rate and the need for higher cumulative exposure doses are not influenced by sex, age, skin type, presence of additional diseases, failure of previous treatment or disease duration. The carcinogenicity associated with phototherapy should not be of much concern as relapses in lichen planus is much less frequent compared to other childhood dermatoses.

In difficult patients not responding to steroid and retinoid, agents like cyclosporine, griseofulvin, metronidazole, phenytoin, sulfasalazine, levamisole, thalidomide, and mycophenolate mofetil have been tried and have had reported success.

In lichen planus pemphigoides, systemic therapy with steroids and/or immunosuppressive agents is recommended whereas in bullous lichen planus topical therapy may alone suffice. However, in children, both the diseases were found to require high doses of systemic steroid along with dapsone or azathioprine. But there are reports of response to topical steroid alone, combination of oral erythromycin (30 mg/kg in four divided doses) and nicotinamide (150 mg three times a day) which was later replaced by dapsone 50 mg daily and topical steroid.[19-22] Actinic LP has responded to treatment with antimalarials and photoprotection.[23]

Nail lichen planus

Prompt diagnosis of lichen planus of the nails is essential because permanent destruction of the nail apparatus is likely to occur without adequate treatment.[11,12,16,17] Treatment includes systemic or intralesional steroids. With topical steroid being ineffective and intralesional steroid injections being very painful and associated with local complications, systemic steroid is usually required. Moreover, with the exception of twenty nail dystrophy, all cases of nail matrix lichen planus are associated with the risk of permanent nail scarring, thereby justifying the use of systemic steroid even in children. Both long-acting injectable steroid and oral prednisone can be given, but to reduce the chances of adrenal

suppression, oral prednisolone is preferred and given as a tapering dose over 4-12 weeks. Oral retinoids are second option and intralesional steroids as nail matrix injection can be a third option for older children. Still local application of potent steroids under occlusion at the nail base can be used if the disorder is largely limited to the nail.

Oral lichen planus

Children affected with oral LP are often asymptomatic or minimally symptomatic. Most patients with the reticular pattern of lichen planus are asymptomatic and require no active treatment.[13,14,16,17] Mucous membrane lesions should be treated if symptomatic, ulcerated, or eroded. The treatment in symptomatic oral LP in children most commonly consists of local regimens because of both the general improvement and the possible side effects of systemic therapy. Topical corticosteroids in adhesive base used several times a day for several months are the drugs of choice for treating mild to moderate symptomatic oral LP because they have fewer side effects. However, widespread, symptomatic lesions require systemic drugs. Topical anesthetics like diphenhydramine elixir, viscous lidocaine, topical steroid like triamacinolone acetonide in orabase or topical tacrolimus ointment given two to three times daily over three months may be beneficial when combined with chlorhexidine rinse, and a plaque control regimen. Plaque control is crucial because oral LP is an inflammatory condition and local irritants, such as plaque and calculus, consistently trigger exacerbations. An alcohol-free rinse should be prescribed to avoid desiccation and irritation of oral tissues. Furthermore, prophylactic use of 0.12% chlorhexidine gluconate rinse reduces the incidence of candidiasis during the corticosteroid therapy. If the child is very symptomatic, topical steroid can be combined with systemic steroid (0.5-1mg/kg) as a tapering dose over 3-6 weeks in the early stages of treatment. Other documented modalities of oral LP treatment include retinoids, cyclosporine rinse, PUVA treatment, dapsone, and cryotherapy.

Studies have related higher levels of anxiety and depression, mechanical trauma of dental procedures, lip chewing habit and intake of citrus and spicy foods causes exacerbation of oral LP. Chronic low-grade irritation from dental plaque and calculous has been suggested to cause exacerbation of gingival lichen planus. Hence avoidance of these factors also plays an important role.

Erosive forms are associated with an increased risk of malignancy and hence periodic follow up and biopsy of suspicious lesions is warranted.

Course and prognosis

Spontaneous resolution of LP in weeks is possible; two-thirds of patients self-resolve after 1year and 10–20% will have intermittent recurrences for many years and residual pigmentary changes can be disfiguring.[9,10,16,17] LP lesions may also persist for years, especially on the shins and in the mouth where the mean duration is 5 years. Poor response to treatment is a feature of hypertrophic LP and lichen planopilaris. Careful follow-up for oral LP is recommended since it has the potential for malignant transformation (1%–2%) especially in familial cases.

Conclusion

To conclude, lichen planus is a rare childhood dermatoses worldwide with a predisposition for patients of Asian origin more so in India. LP in children has similar manifestations as in adults with an increased occurrence of certain patterns like linear LP and rare involvement of oral and nail LP. The response to treatment is same as that in adults and has a favourable prognosis except for the disfiguring hyperpigmentation. The long term systemic side effects in children need to be borne in mind while dealing with immunosuppressants or retinoids for extensive lesions.

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