Childhood lichen planus with nail involvement: a case

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Childhood lichen planus is a very rare entity which is characterized by violaceous, scaly, flat-topped polygonal papules commonly involving the flexor aspects of the wrists and legs, and oral and genital mucous embranes. The incidence of lichen planus peaks between the ages of 30 and 60 years, where as children comprise only 2% to 3% of reported cases. We report a nine-year-old girl with widespread lichen planus, involving the nails, with no mucosal involvement and regressing after treatment with a starting dosage of 20 mg/day systemic flucortolon (Ultralan ®) therapy.

Key words: lichen planus, childhood, nail.

Lichen planus, first described by Erasmus Wilson in 1869, is a pruritic dermatosis of unknown etiology that affects the skin and mucous membranes. It is characterized by violaceous, scaly, flat-topped polygonal papules commonly involving the flexor aspects of the wrists and legs, and oral and genital mucous membranes. Lesions regress with postinflammatory hyperpigmentation. Scarring may occur if hairy areas are involved. There is no sex predilection[1]. However Sharma et al.[2] found the male: female ratio as 2:1 in their 50 childhood lichen planus patient series. The incidence of lichen planus peaks between the ages of 30 and 60 years, where as children comprise only 2% to 3% of reported cases[3].

In this report, we present a nine-year-old child with lichen planus who showed classical generalized pattern regressing after systemic corticosteroid therapy.

Case Report

A nine-year-old Caucasian girl applied to our outpatient clinic suffering from itching papules localized mainly on her trunk for about one month. The papules had started on her limbs and forearms and later became more widespread.

Nail examination revealed longitudinal ridging and pittings over the nail plate. On her left thumb, the nail plate was broken longitudinally and had lost its luster (Fig. 1). Skin lesions were violaceous, flat-topped polygonal papules spread throughout her body (Fig. 2). No mucosal involvement was detected.

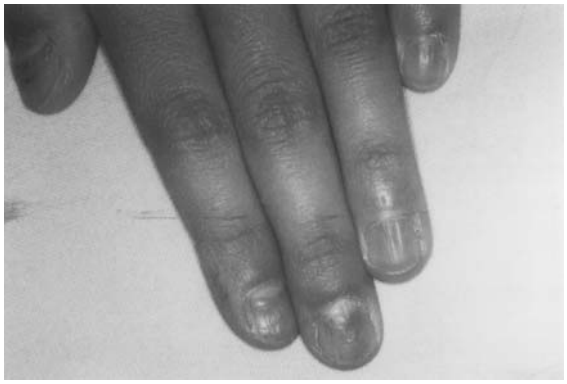

Fig. 1. The pittings and longitudinal ridging of the

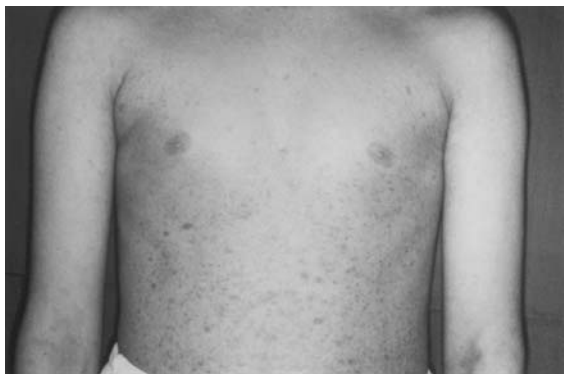

Fig. 2. Widespread papules on the trunk.

There was neither drug nor family history and her blood pressure was normal for age. Routine tests including liver enzymes were normal and hepatitis markers were negative. A biopsy specimen which was taken from a papule showed focal parakeratosis, wedge-shaped hyper-granulosis, saw-tooth acanthosis, band-like dermal lymphocytes infiltrating the epidermis and damage to the basal cells (Fig. 3). This confirmed our diagnosis of lichen planus.

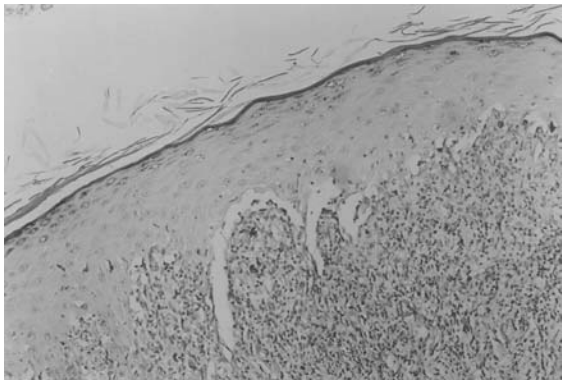

Fig. 3. Histopathology of the lesion showing focal parakeratosis, wedge-shaped hypergranulosis, saw-tooth acanthosis, band-like dermal lymphocytes infiltrating the epidermis and damage to the basal cells.

Fluocortolon (Ultralan ®) (20 mg/day) was started orally and significant improvement was seen in 10 days. The dosage was tapered down progressively and was continued at 5 mg/day for one month. The systemic treatment was then changed to topical corticosteroid ointments for a few persisting papules and resistant nail changes.

From the etiological point of view, child psychiatry consultation was performed. Psychiatric changes and stress had been found to aggravate the disease. Imipramine (Tofranil ®), 25 mg/day, was started and continued for three months. No relapse was detected in the following two years.

Discussion

Childhood lichen planus is a very rare entity. There are some reports that suggest childhood lichen planus is more common in the tropics, especially in Indians[4-6]. The explanation for this condition is still not known, the number of patients reported are very few and new reports are strongly needed. Lichen planus in children must be differentiated from other lichenoid eruptions in childhood, including lichen nitidus, lichen striatus and pityriasis lichenoides[1].

Children with lichen planus should be detected for the familial forms. Familial lichen planus, which is rarely described and uncommon, also occurs at younger ages. Familial lichen planus clinically appears as a more extensive disease with longer duration[3].

The most common morphology of childhood lichen planus is the classical pattern[1,2]. However, it may also present in a more atypical fashion including linear, annular, bullous and follicular variants, which are very rare[3]. Mucous sites can also be affected in lichen planus. Although mucosal involvement is extremely uncommon in children[2], there are also some reports about children with oral lichen planus[7,8]. Nanda et al.[9] reported 23 children with lichen planus, in which oral involvement was seen in 39% of patients. There appears to be a predisposition for some oral lichen planus to develop into carcinoma, and oral lichen planus should be considered as a premalignant condition[10].

The exact etiopathogenesis is unknown. An inborn error of metabolism has been cited, such as glucose-6-phosphate dehydrogenase deficiency, and long-lasting abnormal glucose metabolism with glucose intolerance. Viral infections were suggested but no true viral etiology has been proved. It is known that lichen planus may be associated with severe emotional trauma. Whatever the etiology is, the primary event starts with the damage of the basal cell layer of the epidermis. An unidentified allergen may have an originating effect on this immunological-based process[1]. There are some iatrogenic causes of lichen planus in childhood. A patient with dwarfism receiving human recombinant growth hormone, who developed lichen planus, was published recently[11].

Nail involvement is seen in only 1-10% of lichen planus patients[2]. Nail is the most common affected area when skin lesions appear. Two forms of nail dystrophy have been described. A uniform dystrophy simultaneously involving all 20 nails, usually with loss of luster, thinning of nail plates, bluish-brown discoloration and

longitudinal ridging may occur. An atrophic cicatrizing form in which the nails are involved randomly with pterygium formation and progressive nail loss can also be seen, especially in blacks and Asians[1]. Some nails may be more severely affected than others[12]. Patients with only nail involvement should also be examined for other nail disorders, especially for psoriasis. In order to make the correct diagnosis, longitudinal nail biopsy involving the proximal nail matrix is necessary. If correctly performed, this procedure heals with minimal scarring[13]. In our patient, longitudinal ridgings and pittings were detected. Her left thumbnail was affected more severely, and the nail plate was longitudinally broken with lost of luster.

Nail involvement is the most difficult site to treat in lichen planus. Topical steroid applications to the proximal nail fold are not effective enough and intralesional steroids may not be used in children. Because of the risk of permanent scarring of nails in lichen planus, oral prednisone at an early stage is indicated[3]. In our case, after systemic steroid treatment was started, nail changes healed moderately and her skin lesions flattened.

In the general treatment of lichen planus, oral antihistamines and topical corticosteroids are the main therapeutic agents. For extensive involvement, like in our case, oral steroids can be used at doses of 1-2 mg/kg/day. Adverse effects must be considered carefully. Dapson, griseofulvin and PUVA are alternative therapeutic agents in children. Recently ultraviolet B phototherapy was also found to be safe and effective, especially in patients with eruptive and widespread disease[9].

We wanted to report our case of lichen planus with nail involvement at the age of nine without family history, who improved with corticosteroids. Although lichen planus is not a life-threatening disease, it is known to be an intensive pruritic disorder. Lichen planus may also cause some irreversible scarring that cannot be accepted cosmetically. Such patients will have low quality of life. Since early treatment can avoid such complications, early diagnosis and correct treatment are very important for both dermatologists and pediatricians.

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