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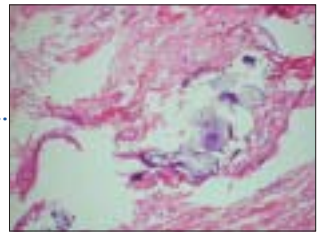
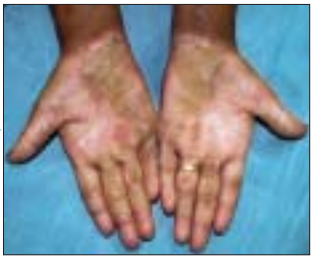
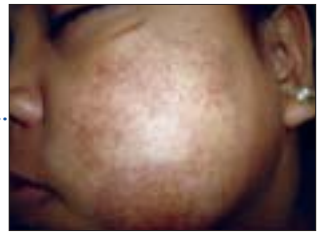
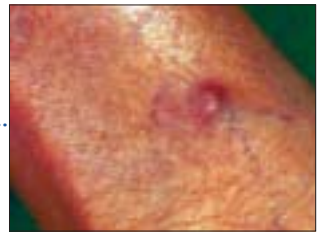
CONTENTS

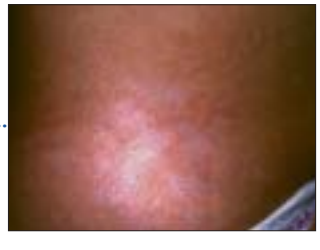


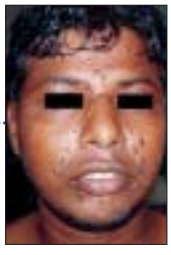


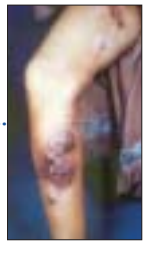


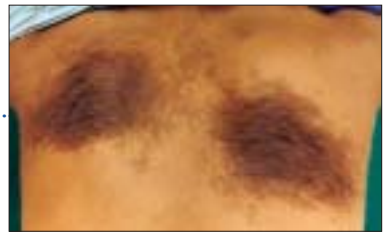


LETTERS TO THE EDITOR









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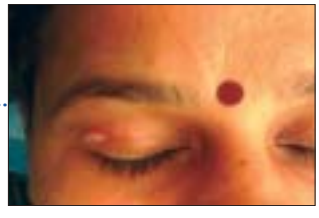

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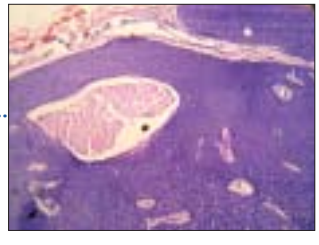

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Acitretin for Papillon-Lefèvre syndrome in a five-year-old girl

Sir,

Papillon-Lefèvre Syndrome (PLS) is a rare genetic disorder with an autosomal recessive mode of inheritance and is characterized by symmetrical transgradient palmoplantar keratoderma and severe periodontal disease. Other features may include tendency to frequent pyogenic infections of the skin and internal organs, and calcification of the dura, particularly the tentorium and falx cerebri. The disease prevalence is one to four persons per million. Both sexes are equally affected. Consanguinity has been observed in one-third of the cases described.[1,2] A few clinicians have previously reported that acitretin treatment is effective.[2-5]

We present experience of acitretin therapy in a case of PLS, who had an excellent response in palmoplantar skin lesions within 2 weeks.

A five-year-old girl presented with palmoplantar lesions that started at the age of 5 months. She also had periodontal problems with loss of multiple incisor and premolar teeth. The patient suffered from teeth problems for seven to 8 months. The parents were first cousins. She had no family history of similar disease. She had no response to previous therapy with topical corticoid, urea and salicylic acid ointments. On dermatologic examination, she had bilateral well-demarcated hyperkeratotic plaques with fissures on the palmoplantar region [Figure 1]. Oral examination revealed red and swollen gingivae associated with premature loss of deciduous teeth. Her weight was 15 kg with normal growth parameters. A pediatrician examined the patient, found no physical abnormality other than the above-mentioned. The patient was referred to a dentist who advised good dental care and bimonthly dental examination. Routine blood count, liver and kidney function tests, cholesterol, triglycerides and urinalyses were within normal limits. No abnormality was found on abdominal ultrasound examination and cranial tomography. X-ray of left hand was compatible with the chronological age of the patient. A skin biopsy specimen from the plantar region revealed hyperkeratosis, irregular acanthosis, dilated vessels within the papillary dermis and perivascular lymphocytic infiltrates.

A diagnosis of PLS was made after dermatological, dental and histopathological examinations. The patient was given acitretin 10 mg PO every day, omitting the third day. The lesions improved with a marked reduction in the hyperkeratosis 2 weeks after the treatment [Figure 2]. The treatment was maintained for 5 months. The patient's skin remained almost lesion-free following the therapy. Her gingivae also showed remarkable improvement. Her unstable deciduous teeth were extracted and replaced by a dental prosthesis. The biochemical tests were performed every month. X-ray of the left hand was repeated at the end of the treatment. Neither skeletal nor biochemical side-effects were seen. The acitretin therapy was discontinued, followed by the rapid recurrence of the palmoplantar keratoderma within 2 weeks. Intermittent therapy is planned during exacerbations of the disease, especially in the winter months.

A multidisciplinary approach including the dermatologist, pediatrician and dentist is important for the therapeutic management of PLS. Different therapeutic options have been used for the management of the PLS-associated palmoplantar keratoderma. Topical keratolytics containing salicylic acid and urea have been used. Especially in winter, palmoplantar hyperkeratosis can worsen with painful fissures limiting routine activities and necessitating systemic treatment.[3] Extraction of the primary teeth combined with oral antibiotics and professional oral hygiene care are measures to improve periodontitis.[6,7] On the other hand, these therapies do not frequently achieve protection of permanent teeth.[7]

A number of authors used systemic acitretin in the treatment of palmoplantar keratoderma and, reported that this therapy is effective at a dose of ranging between 0.4 and

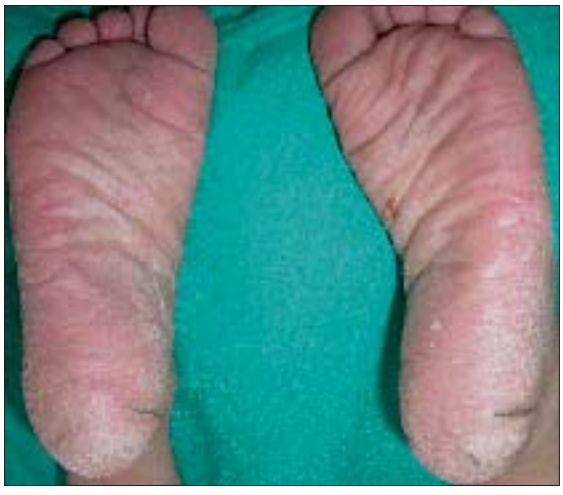

Figure 1: Plantar keratoderma with fissures before acitretin treatment

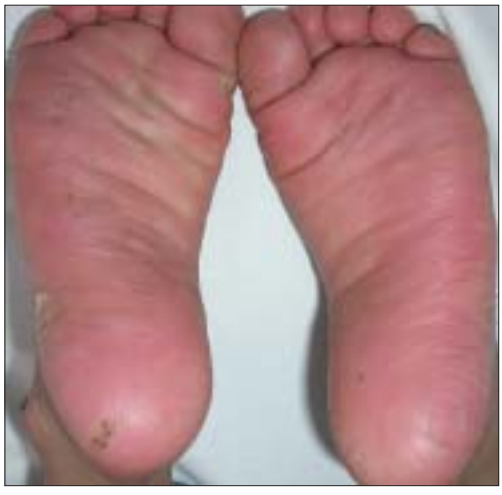

Figure 2: Marked improvement of the plantar keratoderma and fissures 2 weeks after acitretin treatment

1 mg/kg/day.[2-4] The authors noted significant improvement after four to 6 weeks of this therapy. The dose of acitretin was gradually tapered in most of these reports. Lee *et al.,* also reported improvement of periodontal disease after 12 month-therapy with acitretin.[4] Nazzaro *et al.,* reported satisfactory improving of palmoplantar keratoderma and periodontal disease in all three patients which was continued for 16 months of therapy.[5]

According to the above mentioned reports, palmoplantar keratoderma usually recovers rapidly whereas periodontal disease tends to improve later. In our case, the palmoplantar keratoderma had markedly improved in the second week of the therapy. The patient's skin remained almost lesion-free during the 5 month-therapy. Her gingivae also showed remarkable improvement. However, a rapid recurrence of the palmoplantar keratoderma was observed when the acitretin therapy was discontinued.

Many authors have suggested that use of oral retinoids for prolonged periods is useful to prevent loss of permanent teeth in children with PLS.[4,5] The safety of oral retinoids in children remains controversial due to their side-effects on skeletal development. However, a review of the use of acitretin (mean dosage of 0.47 mg/kg) in 46 children for a cumulative period of 472 months revealed that it is a safe and effective treatment in children with keratinization disorders.[8]

In conclusion, the use of low-dose of acitretin in treatment of PLS-associated palmoplantar keratoderma is extremely useful but not curative. The current report suggests that low dosage of acitretin is safe and effective in the treatment of PLS.

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