

Isolated nail lichen planus with primary sclerosing cholangitis in a child

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ABSTRACT

Lichen planus (LP) is an uncommon, inflammatory dermatosis with characteristic lesions affecting the skin, the nails, and the mucous membranes. It is rare in childhood. Although nail abnormalities have been reported in 1 - 10% of patients with LP, the prevalence of nail involvement in affected children is unknown. Here we report a 2-year-old child with isolated nail LP, in association with primary sclerosing cholangitis.

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Lichen planus (LP) is an uncommon, inflammatory dermatosis with characteristic lesions affecting the skin, the nails and the mucous membranes.¹ The LP is rare in childhood, as only 2 - 3% of all cases occur in persons under 20 years of age.² Although nail abnormalities have been reported in 1 - 10% of patients with LP, the prevalence of nail involvement in affected children has not been documented.^{3,4} Since 1969, only 15 proven pediatric cases of isolated nail lichen planus (NLP) have been reported.⁵ We report a child with isolated NLP associated with primary sclerosing cholangitis, to emphasize this association and to alert physicians to the possibility of liver disease, particularly primary biliary cirrhosis (PBC) and primary sclerosing cholangitis (PSC) in patients with NLP.

Case Report. A 2-year-old, Bahraini girl from Bahrain, was referred to us for consideration of liver

transplantation, with end-stage PSC. She was healthy until the age of 16 months when she started to have jaundice with pruritus, dark urine, lethargy, and an increasing abdominal distension. Two months after her illness, her fingers and toenails ceased to grow, and started to turn purple with swollen margins. On examination, she was jaundiced with hepatosplenomegaly and ascites. The nail examination revealed dystrophic nails in both hands and feet. All nails showed swollen red to violaceous proximal nail folds, with severe atrophy to complete absence of nail plates. Many nails showed pterygium formation (**Figure 1**). The mouth and the rest of the skin examination were unremarkable. The diagnosis of isolated NLP was rendered. Biochemical evidence of direct hyperbilirubinemia with elevated transaminases, and prolongation of prothrombin time and partial thromboplastin time were noted. All autoantibodies were negative, apart from the positive anti-smooth muscle antibodies. Computed tomography of the abdomen revealed cystic dilatation, and beading of intra- and extrahepatic biliary ducts. The liver biopsy showed micronodular cirrhosis, with marked bile ductular proliferation typical of PSC. She was started on topical mometasone furoate 0.1% (Elocom, Schering, USA) ointment 2 times daily, to be applied to nail folds and plates. Unfortunately, because of lack of a liver donor, she returned to Bahrain and was lost to follow-up.

Discussion. The LP is an inflammatory dermatosis affecting the skin, the nails, and the mucous membranes and is considered rare in childhood.^{1,2} Moreover, isolated NLP is exceedingly rare.³ In children, NLP can have 3 clinical presentation.⁵ The first form is typical NLP, which presents with diffuse nail ridging, thinning, and splitting.⁵ The second form is 20-nail dystrophy, also known as trachyonychia, which begins insidiously in early childhood and is characterized by excessive longitudinal striations, and loss of nail luster.³ It differs from the other manifestations of NLP because it does not cause permanent loss of nails.³ Twenty-nail dystrophy can be an expression of other diseases, including



Figure 1 - The left hand of the patient showing red violaceous and swollen nail folds, with multiple nail atrophy and pterygium formation.

alopecia areata, psoriasis, and atopic dermatitis.⁶⁻⁸ The third form of NLP is characterized by asymptomatic, progressive nail atrophy with painless nail destruction that may lead to pterygium formation, which, although not pathognomonic, is highly suggestive of NLP.^{1,3,5,9,10} Isolated NLP is considered very rare in children, and was first described in 1969, with only 15 biopsy-proven cases reported in the literature, most as single case reports.⁵ There is a large body of literature describing an association between LP and liver disease, including hepatitis C, PBC, and others.¹¹ The association between LP and PBC was first alluded to when Seehafer¹² reported 6 patients with PBC, who were treated with penicillamine and developed LP. Subsequently, Powell et al¹³ reported a total of 24 patients with LP and PBC, of whom 7 had never received penicillamine. These data suggest that in patients with PBC, LP can occur either spontaneously or in association with penicillamine therapy.¹ The association of LP and PBC is now well recognized, but the association of isolated NLP and PBC was reported only once before by Sowden et al.¹

The PSC is another chronic cholestatic liver disease of unknown etiology characterized by inflammation and fibrosis of the intrahepatic and extrahepatic bile ducts, and has a progressive course leading to sclerosis with eventual obliteration of bile ducts.^{11,14,15} The association of LP and PSC is scarce in literature, with only 5 patients reported.^{11,15} Our case reported here is considered the sixth patient with PSC and LP, and the first patient with isolated NLP.

We conclude that prompt diagnosis of NLP is imperative to prevent permanent nail destruction. The treatment of NLP is disappointing, and challenging especially in children. Topical steroids are mildly effective, while systemic or intralesional steroids are moderately effective.³ Since intralesional steroid injections are painful, systemic administration is usually required and justified even in children, to prevent permanent nail scarring in severe NLP.¹⁶ Given the rarity of LP, and even more the rarity of NLP and PSC in children, this occurrence is significant. We suggest that the findings of NLP should alert the physician to the possibility of liver disease, particularly PBC and PSC.

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