Bilateral Onychodystrophy in a Boy With a History of Isolated Lichen Striatus

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Practice Points

- Lichen striatus (LS) is a relatively rare and self-limited linear dermatosis of unknown etiology and diagnosis usually is made clinically.
- Nail involvement is uncommon in LS but also may be underreported. When present, nail changes may appear before, after, or concurrently with skin lesions.
- If a patient presents with a similar case of isolated onychodystrophy, the clinician should inquire about history of cutaneous LS and should consider the possibility of LS in the differential diagnosis.

Lichen striatus (LS) is a relatively rare and self-limited linear dermatosis of unknown etiology. It primarily affects children, with more than 50% of cases occurring in patients aged 5 to 15 years. The case of a 2-year-old boy who presented for evaluation of a nonpruritic linear rash on the left side of the lower abdomen of 3 weeks' duration is discussed. A diagnosis of lichen striatus (LS) was made and the lesions completely resolved within 6 months. At 5 years of age, the patient returned for evaluation of bluish discoloration and thinning of the nails on the left middle and ring fingers, which ultimately fell off and started to regrow 2 months prior to the second evaluation. The rare diagnosis of isolated onychodystrophy as a late manifestation of LS was made.

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Lichen striatus (LS) is a relatively rare and self-limited linear dermatosis of unknown etiology. Lichen striatus primarily affects children, with more than 50% of cases occurring in patients aged 5 to 15 years.[1,2] It presents clinically as a single unilateral linear band consisting of scaly, 1- to 3-mm papules that coalesce to form long streaks.[3,4] The diagnosis usually is made clinically based on the characteristic appearance of skin lesions and a pattern of distribution that follows the lines of Blaschko.[5,6] The papules usually are asymptomatic; however, if the patient is symptomatic, pruritus is the most common concern. Lichen striatus may resolve with postinflammatory hyperpigmentation or hypopigmentation that may last for several months to years.

Nail involvement is uncommon in LS; a review of the literature has shown that 30 cases have been reported in the world literature since 1941.[7] Nail changes may present before, after, or concurrently with the skin lesions.[4,8] On rare occasions, nail involvement may be the only area of involvement without the presence of typical skin lesions.[8] The involved nails may show longitudinal ridging, splitting, hyperkeratosis of the nail beds, thinning or thickening of the nail plate, nail pitting, and overcurvature of the nail plate, and rarely the nails may fall off completely.[8-10]

We report the case of a boy who was diagnosed with isolated LS at 2 years of age. The lesions spontaneously resolved within 6 months. Three years later

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the patient presented with a rare manifestation of LS in the form of bilateral onychodystrophy.

Case Report

An otherwise healthy 2-year-old boy presented for evaluation of a nonpruritic linear rash on the right lower side of the abdomen of 3 weeks' duration. A review of systems was negative for any other constitutional signs or symptoms. No sick contacts were reported at the patient's home, and his immunizations were up-to-date. His medical history was remarkable for a burn on the left hand from contact with a hot object at 11 months of age that required skin grafting.

Dermatologic examination revealed a linear band of small, 1- to 3-mm, flesh-colored lichenoid papules. Many of the papules had a scaly appearance and some had a vesicular component or were flat topped. The band ranged from 2- to 3-cm wide and was 25 cm in length, extending from the right anterolateral part of the lower abdomen to the right upper lateral part of the buttocks (Figure 1). No abnormalities were noted on the rest of the skin. A diagnosis of LS was made.

At 5 years of age, the patient returned for evaluation of bluish discoloration and thinning of the nails of the left middle and ring fingers of several months duration. The patient was afebrile and appeared to be healthy. There was no lymphadenopathy or hepatomegaly and the rest of the physical examination by a pediatrician was unremarkable. The nails of the 2 affected fingers had fallen off 2 months prior to presentation and had started to regrow. On dermatologic examination, it was noted that the regrown nails showed some residual longitudinal ridging, thinning, and dark discoloration of the proximal nail folds (Figure 2). On examination of the other toenails and fingernails there was evidence of bilateral pitting, ridging, and discoloration (Figure 3). The left great toenail was predominantly affected. The patient's guardians were not aware of the toenail changes and denied any history of trauma to the fingers. When asked about the course of the prior abdominal linear rash, they reported that the lesions had completely resolved within 6 months. The rare diagnosis of isolated onychodystrophy as a late manifestation of the prior LS was made.

Comment

The etiology of LS remains unknown, but there have been several hypotheses suggesting environmental triggers such as trauma[11] or infection.[12] Others have suggested a possible autoimmune response[13] or genetic components.[6] Reports of simultaneous occurrences of LS in siblings as well as in a mother and her son[14,15]; outbreaks of LS among children who are not biologically related but in a shared living environment; and a possible seasonal variation suggest an environmental infectious agent (eg, a virus) as the possible

Figure 1. A linear band of small, 1- to 3-mm, flesh-colored, lichenoid, scaly papules, some with a vesicular component. The band was 2- to 3-cm wide and 25-cm long, extending over the right side of the lower abdomen.

Figure 2. Longitudinal ridging, splitting, and thinning of the proximal nail folds on the regrown nails.

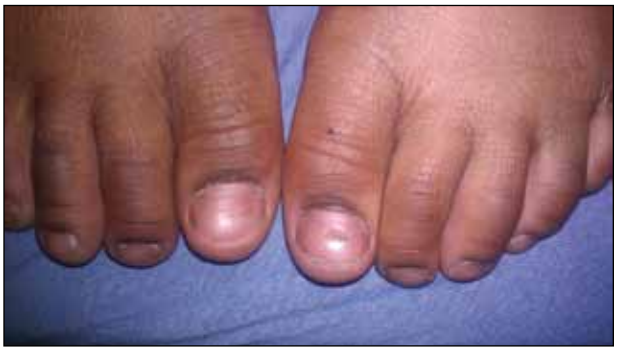

Figure 3. Bilateral pitting, thinning, and overcurvature of the nail plates of the toenails with more involvement of the toenails on the left foot.

triggering factor. However, laboratory testing for viral etiology in LS has not been helpful.

Many of the reported cases of LS have described a pattern of distribution along the lines of Blaschko.[5,6,16,17] Lines of Blaschko are thought to be embryologic in origin and caused by the segmental growth of clones of cutaneous cells or the mutation-induced mosaicism of cutaneous cells, which led to the theory that mosaicism is involved in LS. Lichen striatus needs to be differentiated from other conditions with similar cutaneous appearances (eg, lichen nitidus, linear lichen planus of the digits, linear psoriasis, linear keratosis follicularis, linear epidermal nevus).

Skin biopsy to confirm the diagnosis rarely is necessary, as LS is a self-limited disorder and generally no treatment is recommended. Topical and intralesional steroids do not routinely impact the resolution of LS; however, emollients and topical steroids may be used to treat associated dryness and pruritus, if present.[18] Immunomodulators such as tacrolimus and pimecrolimus have been successfully used in treating persistent and pruritic LS lesions on the face and extremities.[19,20] Tacrolimus also has been successfully used to treat nail abnormalities in LS.[21]

Guardians and family members should be reassured that LS is a benign condition that generally resolves spontaneously within 3 to 12 months. Also, guardians should be counseled regarding the possibility of postinflammatory hyperpigmentation or hypopigmentation, which may last for several months to years, particularly in children with darker skin types. Lichen striatus of the nails may have a more protracted course, lasting from 6 months to 5 years,[22] but usually resolves spontaneously and without deformity.

Our patient developed a rare case of isolated LS at 2 years of age. Reports have suggested later onset of the condition, with more than 50% of all LS cases occurring in children aged 5 to 15 years.[1,2] Despite the earlier onset in our case, the patient still presented with the classic nonpruritic single linear band of papules that is characteristic of LS.

The nail involvement in our case is quite intriguing because of its rarity, timing, and extent of involvement. Nail involvement is generally uncommon in LS, with approximately 30 cases reported worldwide since 1941.[7] The nail changes in our patient were unique in their timing, with the isolated onychodystrophy developing 3 years after the initial skin lesion. This subtle timing may pose a diagnostic challenge in patients with LS if treating physicians are unable to link the presenting onychodystrophy to the earlier cutaneous component of the condition. Two reports have shown that nail changes in association with LS may occur at any time before, after, or concurrently with the skin lesions,[4,8] suggesting that on rare occasions, as in our case, nail involvement may be the only area of involvement without the presence of typical LS skin lesions.[8]

The nail involvement in our patient also showed a greater severity than prior reports,[8,9] as he lost 2 fingernails completely before regrowth. Also, the bilateral distribution of onychodystrophy in our patient involving both the fingernails and toenails appeared to be consistent with a report by Al-Niaimi and Cox.[22]

Nail involvement in cases of LS may be underreported when, as in our case, nail dystrophy presents as the only area of involvement without the presence of the typical skin lesions characteristic of LS. It is reasonable to recommend that clinicians facing similar presentations of isolated onychodystrophy should include the possibility of LS in the differential diagnosis before committing patients to a more common diagnosis (eg, onychomycosis). Clinicians should inquire about any history of cutaneous LS and counsel patients to return for treatment should skin lesions develop that are suggestive of LS.

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