SELF ASSESSMENT QUIZ



Thickening and grooves in left great toenail

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CLINICAL FINDINGS

A-16-year-old girl was referred for left great toenail thickening and yellow discoloration. She was otherwise asymptomatic. The patient had no history of diabetes, infection, or trauma to her toenails. The condition started with gradual onset and slowly progressive course within two years. The patient did not complain of any pain in her left great toenail. And, the only complain was the disfigurement and discomfort during the walking. Physical examination revealed left great toenail to have marked thickening of the nail plate with closely spaced transverse superficial ridging, lack of luster, and brown tan-yellowish discoloration (Fig. 1). There was no subungual debris. There was mild separation of the nail plate from the underlying nail bed (onycholysis). Family history of the same condition was negative. Fungal study



Fig. 1 Thickened left great toenail plate with transverse grooves

including KOH test and fungal culture were negative. Dermoscopy examination showed transverse groove and scaly, yellow brownish thickening of nail plate (Fig. 2).



Fig. 2 Dermoscopy examination: Scaly, Yellowish to brownish thickening with transverse grooves with the nail plate

What is the clinical diagnosis?

- Congenital malalignment of the great toenail.
- Onychomycosis
- Pachyonychia congenita
- Congenital ectodermal dysplasia syndromes.
- Nail psoriasis
- Nail lichen planus
- Connective tissue disease

DIAGNOSIS

Acquired congenital malalignment of great toenail.

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The lateral deviation of the nail plate along the longitudinal axis as a result of lateral deviation of the nail matrix was initially characterized as congenital malalignment of the great toenail by Samman¹ in 1978 and later further illustrated by Baran *et al.*² in 1979. Late onset is unusual for this unusual condition, which often manifests in infancy or childhood.³

Nail plate lateral deviation along the longitudinal axis as a result of nail matrix lateral rotation is the hallmark of congenital malalignment of the great toenails.1 Also included is the median deviation.4 Hallucices and other toenails are frequently affected by malalignment, which can be unilateral or bilateral. Because of repetitive microtrauma to the nail matrix, the nail plate spreads out in transverse ridges. Commonly observed discoloration can be made worse by bleeding or infection.⁵ Pseudomonas and/or fungus infections are frequent opportunistic infections of the nail plate. In many cases, the misaligned toenail becomes embedded in the lateral nail fold, causing pain, erythema, and inflammation. Another common consequence that worsens ingrowth is acute or chronic paronychia. Although the exact cause of congenital malalignment is unknown, a number of theories have been proposed, including lateral rotation of the nail matrix, an anomaly in the ligament that joins the matrix to the distal phalanx's periosteum, and environmental factors (such as vascular or intrauterine injury during fetal development).6 The distal phalanx of the hallux and the nail may grow out of sync, resulting in bigger nail plates that must extend laterally to fit into the underlying bony space, according to a recent theory by Chaniotakis et al.6 Surgical or mechanical trauma are potential causes of acquired forms. The clinical characteristics of 21 individuals with acquired great toenail dystrophy were examined in a retrospective research

conducted by Kim *et al.*⁷ Of the 21 cases (57.1%) where dystrophy developed, 12 had a prior history of trauma. The authors speculate that in the cases mentioned above, trauma served as the catalyst for the development of acquired malalignment. Nail dystrophy is a common symptom of congenital ectodermal dysplasia syndromes, such as pachyonychia congenita. However, these disorders differ in that they also impact other ectodermal structures.

Depending on the consequences and the severity of the disease, there are many different therapy choices. Since about half of the cases will resolve on their own, it is best to start with conservative options. It should be advised to wear comfortable, well-fitting footwear to reduce the likelihood of repetitive injuries. In extreme situations, surgery is necessary to allow the nail plate to grow parallel to the distal phalanx by rotating the nail matrix to the correct orientation.⁸ A wedge-shaped excision is made in the distal toe's curve. The dorsal toe's lateral side is 3-4 mm below where the excision is located. After the nail unit is separated from the bone, the nail matrix is repositioned and stitched into position. Although this cutoff is adjustable, surgical correction is advised up until the age of two.8

A nail matrixectomy is an additional surgical technique in which the nail plate and matrix are completely removed. Although the symptoms are completely resolved when the nail unit is removed, the appearance may not be acceptable. 9,10 In the case study, the patient experienced symptoms during adolescence that were consistent with acquired malalignment. Given that the majority of cases occur in infancy, this is a rare entity. Treatment options are still available and vary from conservative management to surgical correction despite the delayed start. Surgical correction is a reasonable alternative, depending on the patient's



preference, as spontaneous correction is less likely to occur in these people.

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