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Case Presentation

Congenital malalignment of the great toenail. Report of two cases.

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Abstract

Congenital malalignment of the great toenail is characterized by lateral deviation of nail plates, which are not parallel to the distal phalanx. We report two cases of congenital malalignment of the great toenail in two otherwise healthy children. Because this entity is probably underdiagnosed, a high index of suspicion is required to prevent from diagnostic errors and unnecessary treatments.

Keywords: Congenital abnormalities, Nail.

Introduction

Congenital malalignment of the great toenail is a nail disorder characterized by lateral deviation of nail plates, which are not parallel to the distal phalanx. This entity is probably underdiagnosed and there are few reports in the dermatological literature regarding this condition.[1] We report two cases of congenital malalignment of the great toenail in two otherwise healthy children.

Case synopsis

Case 1. A 3-year-old girl was referred to our dermatological unit owing to congenital and asymptomatic alteration of the big toenail of the right foot. Previous topical antifungal treatments had not been effective. There was no history of recent infectious diseases or previous trauma and no other relatives suffered from a similar condition. Physical examination revealed thickening, transverse ridging, onycholysis, and nail plate discoloration varying from green to yellow-brown and reddish. The nail plate was also laterally deviated (**figure 1**). No involvement of other toenails, fingernails, palms, or soles was noticed. Hair and teeth were normal and there were no other cutaneous alterations.



Figure 1. Nail plate thickening, green-brown-reddish discoloration, and pointed shape on the right toenail

Case 2. A 5-year-old boy presented with abnormalities of both big toenails. That alteration had been present since birth and was also asymptomatic. Potential triggering factors were absent and family history for nail diseases was negative. On physical examination, bilateral external deviation of great toenails was evidenced. Onycholysis and nail plate discoloration to brown-green in the distal third of the nail plate were observed (**figure 2**). The remaining examination of the skin, mucous membranes, and adnexal structures did not reveal alterations.



Figure 2. Bilateral deviation of great toenails, distal nail plate discoloration and transverse ridging

In both patients, mycological culture of the nail plate and bacteriological smears from the nail folds were negative. Histological examination of specimens from the nail with Periodic-Acid-Schiff and Gram stains did not reveal abnormalities. The diagnosis of congenital malalignment of the great toenail was made in both cases. Because nail deviation was mild and asymptomatic, and there were no complications, a conservative approach was followed in both children.

Discussion

There are three types of unguis deviation: traumatic, iatrogenic, and congenital. Congenital great toenail deviation or congenital malalignment of the great toenail is probably an underreported nail disorder characterized by lateral deviation of the nail plate frequently associated with nail dystrophy [1-2].

The etiology and pathogenesis are still unclear. It seems to be an autosomal dominant disorder with variable expressivity [1, 3]. Reported cases in monozygotic and dizygotic twins also suggest a genetic process [4]. Exogenous factors including intrauterine pressure, amniotic bands, or vascular abnormalities during fetal life may also play a role in the etiology [3-4]. The pathogenesis has been related to the lateral rotation of the nail matrix. This fact results in deviation of the longitudinal axis of the nail plate that is not parallel to the corresponding axis of the distal phalanx of the hallux [1-3, 5]. An increased traction by a hypertrophic extensor tendon of the hallux pulling the lateral portion of the proximal matrix has been suggested as the responsible cause of the matrix displacement. A desynchronized growth between the nail apparatus and the distal phalanx has also been proposed [3].

The diagnosis of congenital malalignment of the great toenail is usually at birth or in early infancy [1]. It could be possible that only slight displacement of the nail apparatus without changes in the nail plate occurs. This finding could remain unnoticed until a delayed appearance of nail dystrophy during childhood or puberty. It usually appears with simultaneous involvement of both great toenails, but unilateral cases have also been reported, being most frequent on the right foot [3]. It is more commonly an external deviation; medial deviation is extremely rare [1, 4-6]. Involved nails are shorter and show hyperkeratosis, transversal grooves, or ridges giving an oyster-shell-like appearance that favor nail plate breakage. Onycholysis and onychomadesis contribute to transverse or longitudinal elevation of the nail plate. Nail discoloration is also a common feature. In general, nails are darker than the uninvolved nails. They are grey, brown, or black, in the case of microhemorrhages or fungal colonization, and green in the case of *Pseudomonas* superinfection. The thickened nail plate may also be responsible for color alterations. The nail plate usually acquires a triangular or trapezoidal shape [1, 3, 6]. Onychocryptosis and onychogryphosis with accompanying inflammation are the main complications of great toenail deviation [1, 5]. Recurrent paronychia, increasing hypertrophy of the nail plate, or inability to cut the nail can also occur [3]. Associations with Rubinstein-Taybi syndrome [2] and ocular melanosis [6] have been documented.

The characteristic clinical presentation in combination with congenital involvement of both toenails and lateral deviation of the nail plate are the main clues for the diagnosis [3]. The differential diagnosis may include onychomycosis, connective tissue disorders, neuropathies, nail apparatus tumors, dermatoses with nail involvement such as psoriasis, or ectodermal dysplasia [1].

Spontaneous improvement is expected in approximately one-half of the patients before the age of 10 years, but irreversible damage of the nail plate may develop if surgical treatment is delayed [6]. To select the correct therapy, the degree of deviation should be considered. In the case of minimal deviation, a conservative and expectant attitude is indicated, based on prevention and treatment of complications. In the case of severe deviation or early complications, surgical treatment is indicated; better results are achieved by surgery before the age of 2 years [3-5]. Surgical correction consists of alignment of the nail matrix and nail plate by medial rotation in the case of lateral deviation and external rotation in the case of medial deviation [1]. In addition to positional correction, elongation of the extensor tendon of the hallux is also recommended [3].

In conclusion, a high index of suspicion is necessary for the diagnosis of congenital malalignment of the great toenail. The recognition of this entity could prevent diagnostic errors and unnecessary treatments.

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