Title:

Late onset malalignment of the great toenails

Short running title:

Late onset great toenail malalignment

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This is the author manuscript accepted for publication and has undergone full peer review but has not been through the copyediting, typesetting, pagination and proofreading process, which may lead to differences between this version and the [Version of Record](#). Please cite this article as [doi: 10.1111/AJD.13101](#)

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Article type : Brief Reports

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Abstract:

Great toenail malalignment is characterised by lateral deviation of the longitudinal axis of the nail plate with respect to the hallux, and is usually posttraumatic, iatrogenic or due to congenital malalignment of the great toenails. We present cases of great toenail malalignment with onset in adolescence or young adulthood without preceding nail surgery or acute trauma. We postulate that this may represent a late onset presentation of congenital malalignment of the great toenails.

Learning points:

- Late onset malalignment of the great toenails should be considered in adolescents and adults presenting with apparent onychogryphosis affecting the great toenails, especially if lateral nail plate deviation is evident
- The dystrophic appearance of the great toenail is a result of an underlying nail plate malalignment exacerbated by repeated stress and damage from tight footwear, exercise and sports

Keywords

Nail, dystrophy, congenital malalignment, great toenail

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Introduction

Great toenail malalignment has three main causes; congenital malalignment of the great toenails, post-traumatic and iatrogenic. Traumatic nail malalignment usually arises from an acute event such as laceration to the nail matrix or fractures of the terminal phalanx. Iatrogenic nail malalignment may arise from wide lateral nail biopsies and excisions¹. Congenital malalignment of the great toenail typically presents in early infancy with lateral deviation of the long axis of the nail plate and onychodystrophy resembling an “oyster-shell” in appearance.

We present a series of patients with great toenail dystrophy resembling congenital malalignment of the great toenails with onset in early adolescence or young adulthood, but without an inciting traumatic or iatrogenic precipitant. This may represent a late onset variant of congenital malalignment of the great toenails.

Main text

We performed a manual search of all clinical encounters during a three-year period (November 2015 to January 2019) at a specialised nail disorder clinic (Skin & Cancer Foundation Inc., Carlton, Australia). We screened potential cases for documentation of malalignment of the great toenail(s) during the clinical encounter. Two dermatologists reviewed clinical photographs of all potential cases. We included cases based on the presence of lateral deviation of the great toenail, onychodystrophy with transverse ridging and discolouration. Cases were excluded if photographic evidence was not available for review. All clinical data was collected retrospectively from the electronic medical record.

Our initial search yielded 13 potential cases from the three-year period. A total of seven cases were included; five were excluded due to the absence of clinical photographs, and one case was excluded due to an incorrect diagnosis after review of clinical photography.

Of the seven patients, five were female and the median age of presentation was 21 (range 12 to 35). None of the patients reported an acute traumatic event or toenail surgery preceding the onset of the deformity, however a history of chronic mechanical stress and damage from regular exercise/sports, tight footwear and pubertal growth spurt was common. Five cases had bilateral involvement of the great toenails. There was no involvement of other toenails in all of our patients. Two patients (cases 1 and 7) were previously diagnosed with onychocryptosis and underwent nail avulsion surgery, but the nail dystrophy and lateral deviation persisted after nail regrowth. Clinical and demographic data is summarised in table 1.

None of our patients opted for surgical intervention at the time of diagnosis. All patients were recommended conservative measures including podiatry review for burring of the thickened nail, maintaining shorter toenail length and appropriately fitting footwear. One patient (case 2) was followed-up, and there was some cosmetic improvement with the aforementioned conservative measures.

In 1978, Samman reported cases of great toenail dystrophy in neonates with features of discolouration, transverse ridging and overcurvature². A year later, Baran et al. attributed this congenital onychodystrophy to minor trauma against the shoe in the setting of a laterally deviated longitudinal axis of the great toenail and coined the term “congenital malalignment of the big toenail”^{3,4}. Nail discolouration occurs from haemorrhage (grey to brown from) and/or bacterial infection (green), whilst transverse ridging reflects episodic damage to the nail matrix³. Resultantly, the dystrophic nail resembles an “oyster-shell” in appearance (see figure 1). Shortening and thickening of the nail plate, hypertrophy of the lateral nail fold (due to the obliquely growing nail burrowing into the lateral nailfold), transverse overcurvature and onycholysis may also be present^{3,5}. The abnormal nail is predisposed to complications including onychomycosis, bacterial infection, paronychia and onychocryptosis. Lateral deviation of the distal phalanx of the hallux from the metatarsal-proximal joint is also common and is accentuated by the nail malalignment¹.

Recently, Decker et al. described two cases of great toenail malalignment with onset in adolescence and termed this “acquired congenital malalignment of the great toenails”⁶. It is

unclear whether this late onset onychodystrophy is related to an underlying congenital malalignment or whether this is an unrecognised clinical entity. It is plausible that a mild variant of congenital malalignment with minimal nail plate deviation goes unnoticed in infancy and childhood, and then presents later in life with onychodystrophy following repeated mechanical stresses and microtrauma^{6,7}. Indeed, most of our patients were young adults (median age of 21) and had chronic low-grade damage to the toenail from sports and exercise, pubertal growth spurt and tight footwear. None of our patients reported a preceding acute traumatic event or surgery to suggest an iatrogenic or traumatic malalignment either. Nevertheless, to avoid confusion and given the undetermined aetiology, we suggest 'late onset malalignment of the great toenail' is a more appropriate term.

The aetiology and pathogenesis of congenital malalignment of the great toenails is not well understood either. Some propose an underlying heritability in an autosomal dominant manner with variable expression, due to occurrences in monozygotic and dizygotic twins and several generations of a single family⁷. Based on MRI evidence, tension from the extensor tendons of the hallux pulls the proximal nail plate laterally resulting in displacement of the longitudinal axis of the nail. Others have suggested desynchronised growth of the nail apparatus in relation to the distal phalanx, resulting in larger nail plates that must grow laterally to fit into the underlying bony space⁷.

Recognition of the underlying malalignment is important to avoid unnecessary treatment and its associated harm. As aforementioned, two patients underwent nail avulsion surgery for an initial diagnosis of onychocryptosis but the nail malalignment persisted after regrowth. The main differentials diagnoses are early onychogryphosis, onychocryptosis and onychomycosis. Onychomycoses is a common cause of nail dystrophy, however it does not usually cause nail malalignment. Nevertheless, we recommend fungal cultures for all patients as onychomycoses may complicate the underlying malalignment. All of our patients had nail clippings cultured, and most had negative culture results. In the two patients with positive fungal culture results, treatment with antifungals in these patients did not significantly improve the malalignment or the appearance of the nail. Onychogryphosis is characterised by thickening of the nail plate with gross hyperkeratosis and transverse curvature resembling a "ram horn" in appearance⁸. In onychogryphosis, the axis of deviation is usually upwards or downwards rather than lateral⁴. We believe that some cases of onychogryphosis, particularly those limited to the great

toenails and associated with lateral deviation of the nail plate, are misdiagnosed and the underlying problem is congenital malalignment. Indeed, Baran et al. used the term “hemionychogryphosis” to describe onychogryphosis-like changes in long-standing congenital malalignment of the great toes after chronic microtrauma or if there is insufficient attention paid towards the care of the nail^{3,8}. Ultimately, the diagnosis of late onset or congenital malalignment is clinical. There is no established role for nail biopsy, but it may be indicated if a tumour of the nail unit is suspected.

Based on limited observational data, around 50% of children with congenital malalignment experience spontaneous resolution and realignment of the nail by ten years of age, especially in those with minimal deviation^{1,5,7}. Accordingly, it may be reasonable to begin with a conservative approach in adults if the deformity is not psychologically distressing to the patient. As aforementioned, podiatry review for burring of the thickened nail, regularly maintaining shorter nail length and appropriately fitting footwear should be advised. Toe taping, sometimes used in the management of onychocryptosis⁹, may be a non-surgical option that could counter the lateral pull exerted by the extensor tendons of the hallux and ameliorate distal nail wall hypertrophy.

Nevertheless, persisting or severe nail malalignment may necessitate surgical intervention. Rotation of the whole nail unit, nail matrixectomy, ungueodermal flap repair, and surgical elongation of the distal phalanx have been reported as potential treatment options^{1,5,10}. It is unknown whether these surgical interventions would be effective in adults with long-standing nail malalignment.

Conclusion

We present several cases of nail dystrophy with lateral nail plate deviation. We believe an underlying congenital malalignment is present from birth, but onychodystrophy is only evident later in life after repeated stress and damage to the nail. Therefore, we suggest that the diagnosis of late onset malalignment of the great toenails should be considered in adolescents or young adults with great toenail dystrophy without preceding acute trauma or nail surgery. Further studies would be useful in exploring treatment methods and outcomes for this

condition. In particular, it would be worthwhile exploring the efficacy of surgical interventions and conservative measures such as podiatry review combined with toe taping.

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Figure legend

Figure 1 depicts characteristic features of congenital malalignment of the great toenails. Note the presence of lateral deviation of the nail plate with respect to the longitudinal axis of the nail plate and an oyster shell-like appearance due to transverse ridging and discolouration.

Case	Age	Sex	Medical history	Exacerbating factors	Involvement	Duration	Fungal cultures	Response to antifungals
1	27	M	Seborrhoeic dermatitis	Regular exercise/sports	Bilateral	4 years	Nail clippings x3 negative	Oral terbinafine and fluconazole - no improvement
2	35	F	Recurrent herpes simplex virus infection	Tight footwear	Bilateral	6 years	Nail clippings x1 negative	Oral terbinafine - no improvement
3	20	M	Nil	Nil	Bilateral	3 years	Nail clippings x1 positive	Oral terbinafine for 12 months - no improvement
4	34	F	Nil	Nil	Unilateral	5 years	Nail clippings x2 negative	Oral terbinafine - no improvement
5	21	F	Hypothyroidism	Nil	Bilateral	6 years	Nail clippings x1 negative	Oral terbinafine for 6 months - no improvement
6	12	F	Hallux valgus deformity	Growth spurt	Bilateral	2 years	Nail clippings x1 negative	No treatment
7	21	F	Acne	Tight footwear	Unilateral	4 years	Nail clippings x2 (x1 positive)	Oral terbinafine for 3 months - no improvement

Table 1. Clinical and demographic data for all six cases of acquired congenital malalignment of the great toenails

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Date:

2019-11-01

Citation:

Wang, C. Y., Kern, J. & Howard, A. (2019). Late-onset malalignment of the great toenails. AUSTRALASIAN JOURNAL OF DERMATOLOGY, 60 (4), pp.315-317. <https://doi.org/10.1111/ajd.13101>.

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