

## IN-DEPTH REVIEW

**Acquired Congenital Malalignment of the Great Toenails: A Case Series**

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**ABSTRACT**

Congenital malalignment of the great toenail is a dystrophic disorder that is characterized by lateral deviation of the nail plates. It is usually present at birth and diagnosed very early in life. Late onset of this condition, often referred to as, “acquired,” is very uncommon. Mechanical stress and repeated microtraumas to the nail can cause complications that can worsen the appearance of the nail and make the condition more obvious in older individuals, however, there are other triggers for this condition as well, such as drugs that can cause nail changes. Properly diagnosing this condition can prevent patients from undergoing unnecessary treatments and also facilitate the efficient initiation of the correct treatment in patients. Here, we discuss 3 cases of acquired congenital malalignment of the great toenail in patients during their 5<sup>th</sup>, 8<sup>th</sup>, and 7<sup>th</sup> decades of life.

**INTRODUCTION**

Congenital malalignment of the great toenail is a dystrophic disorder of unknown etiology that is characterized by lateral deviation of the nail plates, which are not parallel to the major axis of the distal phalanx. This condition is typically present at birth, or very early in life, and can be diagnosed by observing the toenail and noting its pattern of growth.<sup>1</sup> Certain diagnostic tests can be performed to rule out other possible diagnoses on the differential. It has been hypothesized that an abnormality in the ligament that connects the matrix to the periosteum of the distal phalanx may be the root cause of this disorder.<sup>2</sup> It is possible that,

in cases where the disorder is very mild during early life, it may go unnoticed initially. These children will not be diagnosed with this condition until they are older or until they develop complications related to repeated microtraumas to the toenail, both of which have the potential to make the condition more obvious.<sup>1,3</sup> In this case series, we present three cases of acquired congenital malalignment of the great toenail that did not become apparent until much later in life, with the disorder apparently triggered by various insults. We also discuss the outcomes for patients and the treatment options available.

**CASE SERIES**

**Case 1 Synopsis:** The patient is a 48-year-old female with no significant past medical history, who presented to our clinic with bilateral first toenail dystrophy. She reported that at 18 years of age, her right great toenail endured significant trauma from someone who stepped on it while wearing steel-toed boots, which resulted in subungual hemorrhage and subsequent loss of the toenail. Unfortunately, when they nail grew back, there was some degree of dystrophy and discoloration present. In her mid-20's, the right great toenail would reportedly exhibit a recurring pattern of partial regrowth followed by shedding of the entire nail. After this cycle occurred several times, the right great toenail stopped falling off and became persistently thickened. Podiatry was consulted and her toenails were tested for fungus, which she reported was negative. Nevertheless, the podiatrist treated patient with oral terbinafine without success.

In 2022, the patient reported noticing similar changes in her left great toenail. For this nail, she opted to try treating the condition with Kerasal nail repair. This over-the-counter treatment is meant to be applied topically to damaged nails to reduce discoloration, normalize nail thickness, and hydrate brittle nails. After a year of treatment, the patient reported improvement in the appearance of her nails with new growth at the proximal aspect of both great toenails. However, the patient subsequently got a pedicure, after which both great toenails became dystrophic again. In addition to the changes in nail appearance, the patient reported recurrent ingrown toenails and discomfort, which she managed by keeping her toenails short.

Physical examination at time of presentation to clinic showed yellow discoloration, full-length thickening of both halluces, and lateral deviation of the nail plate (**Figure 1**). Nail clippings, taken and sent for PAS staining of

patient's left great toenail, were negative for fungal infection. A diagnosis of acquired malalignment of the great toenails was made. The patient was grateful for a definitive diagnosis, but declined surgical correction.

**Case 2 Synopsis:** A 74-year-old female with no significant past medical history presented to dermatology clinic with a chief complaint of "toenail issues." She reported that she had toenail fungus on her left foot, and described gradual thickening and yellow discoloration that had continuously worsened over the course of the previous three years. Prior to this, her toenails were reportedly normal. The patient believed that the nail became ingrown initially after cutting the nail too short, which she thought may have initially triggered the nail dystrophy. She reported no prior treatment, as her primary care physician initially provided reassurance and informed the patient that the nail would likely eventually fall off.

Physical examination showed medial deviation of the left great toenail with onychogryphosis, discoloration, and retronychia, with milder medial deviation, hyperkeratosis, and discoloration on the remaining toenails on the left foot (**Figure 2**). The condition was diagnosed as acquired malalignment of the left great toenail, likely from repeated trauma. In this case, the patient did wish to pursue surgical intervention and was referred to podiatry for more definitive treatment.

**Case 3 Synopsis:** A 67-year-old female with a past medical history significant for breast cancer, melanoma in situ, and dermatitis herpetiformis (treated with dapsons) presented to dermatology clinic for a routine skin cancer examination. In 2014, she underwent a 4-cycle course of paclitaxel for recurrent breast cancer, at which time she first reported asymptomatic discoloration of



**Figure 1.a.** Bilateral acquired malalignment of the great toenails induced by initial traumatic event on the right toe.

**Figure 1.b.** Bilateral acquired malalignment of the great toenails arising idiopathically on the left toe.



**Figure 2.** Unilateral malalignment of the great toenail following repeated trauma.



three of her toenails. Paclitaxel is well known to cause cutaneous side effects, with up to 89% of patient reporting hair loss.<sup>4</sup> Nail changes are less common, but still a well-documented as a potential side effect of the medication.<sup>4</sup> Onycholysis, subungual hemorrhage, dystrophy, Beau's lines, and melanonychia have all been reported. Because of this, the discoloration in the patient's toenails was initially attributed to paclitaxel and it was suspected that the condition would resolve with time, after the course of chemotherapy was complete. The condition, however, persisted, with subjective increase in thickness and discoloration over time. Physical examination at the time of presentation in dermatology clinic showed bilateral brown thickening and increased curvature of the great toenails (**Figure 3**). The condition was diagnosed as acquired malalignment of the great toenails, likely secondary to paclitaxel treatment. The patient was reassured and agreed to continue monitoring.

## DISCUSSION

Malalignment of the great toenails typically presents as lateral deviation of the nail plate along the longitudinal axis due to lateral rotation of the nail matrix. While lateral deviation is more common, medial deviation has also been reported.<sup>5</sup> The halluces are the most commonly affected toenails, however, other toenails can be affected. Malalignment may also be unilateral or bilateral, as seen in this case series. This condition is usually present at birth, as most congenital conditions are, however, in cases of minimal deviation, it may not be noticed until later in childhood or during puberty when nail dystrophic changes occur from mechanical stress as patients become more active, as infants and babies who are not yet walking have minimal trauma to their feet and nails.

The stress caused by the repetitive microtraumas can cause complications that can make the condition more obvious and recognizable. However, other possible triggers exist, including drugs that can induce nail changes, such as paclitaxel, as discussed in case 3 above. There have been cases published on this condition being diagnosed in teenage patients,<sup>3</sup> as this is rare, however, the above cases are unique in that these patients were diagnosed much later in the 5<sup>th</sup>, 8<sup>th</sup>, and 7<sup>th</sup> decades of life.

Features of congenital malalignment of the great toenail include transverse ridging often seen in the nail, as well as discoloration of the affected nail(s), which may be exacerbated by microtraumas to the affected areas. Infections of the nail plate can also occur, which may also contribute to the discoloration, and the malalignment of the nail can cause the nail to grow in such a way that it becomes imbedded in the lateral nail fold, causing redness, inflammation, and tenderness (ingrown nails),<sup>2</sup> which was a complication reported by two of the above described patients. It is suggested that about half of patients affected by this condition and diagnosed in infancy or early childhood will have spontaneous realignment of the nails before the age of 10 years.<sup>1,6</sup> However, it is important to note that this exact number is difficult to elucidate, as there are very little long-term follow-up data available.<sup>1</sup> It is also important to note that this condition may have a genetic component, as it seems to have autosomal dominant inheritance with variable expressivity.<sup>6</sup> Thus, it may be useful for clinicians to consider taking a family history to identify relatives that may have symptoms suggestive of this condition when they are considering this condition on the differential for a given patient.

The exact pathogenesis for congenital malalignment of the great toenails is



**Figure 3.** Bilateral malalignment of the great toenails induced by paclitaxel.

unknown, although several theories having been suggested. Several of these theories include genetic causes, lateral rotation of the nail matrix, and an abnormality in the extensor tendon of the hallux, which is the ligament that connects the matrix to the periosteum of the distal phalanx. Environmental causes may also play a role, such as intrauterine or vascular insults during fetal growth. Another hypothesis involves desynchronization in growth between the nail and the distal phalanx of the hallux, which forces larger nail plates to grow laterally in order to fit the underlying bony space.<sup>1,2,7</sup>

The acquired form of congenital malalignment of the great toenails may be caused by surgical or mechanical trauma. In fact, a history of prior trauma has been reported in 51.7% of cases.<sup>2</sup> In cases 1 and 2 presented here, both patients reported histories consistent with acquired malalignment precipitated by prior trauma to the nail.

An accurate diagnosis is important to prevent unnecessary treatments that can have adverse effects, such as the oral antifungal therapy that the described patient in case 1 received, which did not successfully treat the condition. The differential diagnosis should include, but is not limited to, onychomycosis, lichen planus, nail apparatus tumors, ectodermal dysplasia syndromes, connective tissue disorders, and other dermatoses.

In order to differentiate conditions on the differential diagnosis, certain clues and diagnostic modalities can be used. Onychomycosis would show that the culture of a nail biopsy is positive for fungal organisms, commonly *Trichophyton rubrum* and *Trichophyton mentagrophytes*, which congenital malalignment of the great toenail would not show unless a superimposed fungal infection was present. Onychomycosis

often requires treatment with oral antifungals, such as terbinafine, fluconazole, and itraconazole, as topical antifungals are often ineffective at completely eradicating the infection.<sup>8</sup> Lichen planus of the nails often presents with alterations of the nail plate that cause thinning of the nail plate and longitudinal ridging, as opposed to transverse ridging that is seen in congenital malalignment of the great toenail. Lichen planus of the nail can also result in a loss of transparency of the nail, resulting in a gray discoloration.<sup>9</sup> Various tumors may also affect the nail apparatus and should be on the differential, however, these may only affect a portion of the nail and can often be diagnosed with a biopsy, which can guide treatment for the lesion.<sup>10</sup> Ectodermal dysplasias are heritable conditions that affect ectodermal structures and their appendages, including hair, nails, teeth, and sweat glands. In the nails, these conditions can result in dystrophy, hypertrophy, and abnormal keratinization. These conditions can be diagnosed and their effects elucidated with modern molecular genetics techniques.<sup>11</sup> Nail changes in connective tissue diseases may include erythema of the proximal nailfold, splinter hemorrhages, capillary loops in the proximal nailfold, periungual erythema, thin nail plates, increased longitudinal curvature, transverse curvature, and a white discoloration in the nails. In these conditions, the proximal nailfold is the most affected area of the nail, which may help to distinguish congenital malalignment of the great toenail, which affects the growth of the entire nail.<sup>12</sup>

There are a wide variety of treatment options based on the severity of the condition, the degree of deviation, and the extent to which the patient wishes to address the condition. Treatment options for the acquired form of this condition remain the same as those for congenital cases, though spontaneous resolution is unlikely to occur for the acquired

cases. As mentioned earlier, approximately half of congenital cases will improve or resolve on their own, so conservative treatments are often initiated first in these cases. As this condition can be exacerbated by repeated trauma,<sup>13</sup> no matter what age in life it presents at, patients should be counseled on wearing properly fitting, comfortable footwear and protecting the feet.<sup>2</sup> Certain activities can exacerbate the condition as well, including dancing and active sports that cause impact to the feet, so this should be discussed with patients as well.<sup>1</sup> Definitive treatment for both congenital and acquired malalignment of the great toenails is surgical correction, which may be performed in patients ideally 2 years of age or younger.<sup>1</sup> One surgery involves creating a wedge-shaped excision, dissecting the nail unit away from the bone, and rotating the whole nail unit before suturing it into place, allowing the nail plate to grow in parallel to the distal phalanx.<sup>1,2</sup> Another surgical option involves complete removal of the nail plate and matrix, termed “nail matrixectomy,” will result in resolution of symptoms, however, this may not be cosmetically acceptable to some patients. Finally, surgical elongation of the extensor tendon of the affected hallux has also been suggested as a potential surgical treatment modality.<sup>1</sup> In acquired cases that are diagnosed in older patients, spontaneous correction is uncommon, so surgical correction is usually the most viable treatment option to optimize corrective results.<sup>3</sup> Patients should be counseled about this, however, they may opt not to undergo an invasive treatment for a benign condition, which is demonstrated by one of the patients described above who declined surgical intervention.

## CONCLUSION

Congenital malalignment of the great toenail is a dystrophic nail disorder of unknown origin that is inherited in an autosomal dominant fashion with variable expressivity. This condition usually presents in infancy or early childhood, making late onset, often referred to as “acquired,” as described in the cases above, uncommon presentations. Complications of this conditions include infection, discoloration of the affected nail(s), and ingrown nails that result in erythema, inflammation, and tenderness. The described cases are unique because this condition was diagnosed in much older patients than the condition is typically first recognized in. Thus, even though it is rare, it is important for clinicians to be aware of this diagnosis in patients of all ages that fit the criteria, but to effectively rule out other possible diagnoses on the differential. It is important that the astute clinician maintains a high index of suspicion for this condition when patients present with lateral deviation of one of more of their toenails so that an accurate diagnosis can be made in order to efficiently initiate appropriate treatment and avoid unnecessary therapeutic modalities and their side effects.

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