Circumferential Nail

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Established Facts

- Circumferential nail is a congenital developmental anomaly of the nail.
- Circumferential nail is also known as congenital palmar nail syndrome.
- Dorsal dimelia leads to the formation of circumferential nail.

Novel Insights

- Absence of bone beneath the tubular nail supports the hypothesis that the underlying bone is essential for the normal development of the nail.
- Nail fold capillaroscopy shows similar features on dorsal and palmar proximal nail fold, indicating complete dorsal dimelia.

Keywords

Circumferential nail · Tubular nail · Dorsal dimelia · Nail fold capillaroscopy

Abstract

Circumferential nail is an extremely rare congenital anomaly of nail development. Only a few case reports have been published in the past. We report a 20-year-old male with circumferential nail without any other congenital malformation.

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Introduction

Congenital abnormalities of nail development constitute a heterogeneous group ranging from changes which are hardly noticeable to those with complete anonychia

[1]. Kalikian [2] attempted the first classification of congenital abnormalities of nail development into: absence of nail growth, abnormality of nail augmentation, and abnormality of nail implantation. Circumferential nail (CN) is a very rare anomaly of nail development with only a few cases reported in the past [3–8]. We report an Indian male with CN in the accessory digit.

Case Report

A 20-year-old healthy male presented to us with a tubular nail on the duplicated right thumb since birth (Fig. 1a, b). His concern was the abnormal appearance of the nail. He was born out of a non-consanguineous marriage by normal vaginal delivery. There was no history of drug intake or alcohol use by the mother during pregnancy. There was no similar family history. He had a normal physical and mental development and denied any intervention in the past for his nail and digit. The rate of growth of the affected nail was slower than the other nails.

Fig. 1. **a** Right hand of the patient showing a supernumerary digit arising from the base of the thumb. **b** The extra digit was seen to have a tubular or circumferential nail.

On examination, the duplicated thumb of the right hand showed presence of a tubular nail completely encircling the digit (Fig. 1b). Proximal nail fold and cuticle were present all around the base of the nail. The central portion showed a pinkish, fleshy, slightly hyperkeratotic centre. The mobility of this accessory digit was restricted as compared to the other digits. The rest of the digits and nails were normal; mucocutaneous and systemic examination was also non-contributory. Nail fold capillaroscopy revealed the presence of a normal capillary distribution and morphology on both dorsal and ventral proximal nail folds (Fig. 2a–c). An X-ray of the affected hand revealed a duplicated thumb with triphalangism (Fig. 3a, b). Nail shadow was visible in the film, without any evidence of bone in the centre of the nail (Fig. 3b).

Discussion

An abnormal dorsalisation of the palmar/plantar aspects of hands and feet, respectively, is termed as “dorsal dimelia” [8]. It can present as a full or partially expressed phenotype and occurs due to an abnormal development of the dorsoventral axis of the hand and foot paddle during morphogenesis [8]. CN is an extremely rare congenital anomaly of the nail which occurs in patients with a full expression of the dorsal dimelia phenotype, also known as the “congenital palmar nail syndrome” [8]. In some cases, CN can also occur either as an isolated anomaly or in association with other congenital anomalies [3–9].

CN more commonly involves the ring and little finger nails, rarely being reported on the thumb [7]. Due to a paucity of reports, the exact incidence, gender predisposition, predisposing factors as well as long-term outcomes or pathogenic mechanisms for this condition are not well understood.

Embryologically, the earliest recognisable area of the nail unit (the nail anlage) appears at around the 9th week of gestation. By the 13th week, the nail field is well defined with a matrix primodium underlying the proximal nail fold. By the 14th week, the nail plate starts to emerge and is well established in conjunction with the limb bud. The interaction between various signaling pathways is essential for this normal development.

Upper limb development, including the digits, occurs in three axes: dorsoventral, proximodistal, and anteroposterior [7]. In our case, the duplicated thumb reflected abnormality in all three: dorsal dimelia (dorsoventral axis), preaxial polydactyly (anteroposterior axis) and triphalangism (proximodistal axis). The role of several genes (BMP4, BMP7, BMP-1A, En-1, Wnt7a, Lmx-1) in the development of limb bud has been well described in the literature [7, 10, 11]. Stimulation of BMP (bone morphogenic protein) receptor-1A (BMPR-1A) by BMP4 and BMP7 results in an induction of the transcription factor En-1 in the ventral ectoderm of the limb bud. En-1 is responsible for a normal development of the ventral

Fig. 2. **a, b** Nail fold capillaroscopy showing a normal pattern and distribution of nail fold capillaries on both the dorsal (**a**) and ventral or palmar nail folds (**b**) (Dinolite AM7115MZT, polarised view with linkage fluid, 200×). **c** Dermatoscopy (onychoscopy) of the distal end of the digit shows the hyponychium (Dinolite AM7115MZT, polarised view with linkage fluid, 50×).

structures and also restricts Wnt7a, which in turn induces the transcription factor Lmx-1 (responsible for the development of the dorsal structures). In the absence of En-1, Wnt7a expression extends to both the dorsal and ventral ectoderm, resulting in dorsal dimelia. It has also been argued that any loss-of-function or gain-of-function mutations in these genes should ideally result in expression of the abnormality in all fingers. Al-Qattan and Kfoury [7] proposed that sporadic cases might result from stochastic developmental error.

Baran and Juhlin [12] proposed that nail development is bone dependent, possibly explaining the coexistence of

nail and bone abnormalities in many cases. In our patient, there was an absence of any bony structures in the centre of the CN as seen on X-ray. Previous cases have not commented on this observation. Based on the above hypothesis, it can be argued that presence of bone beneath the nail unit may have prevented the development of CN.

We also attempted nail fold capillaroscopy to see for any changes in proximal nail fold capillary distribution and morphology on the ventral (supposedly palmar) aspect of the extra digit. The normal appearance as well as similarity of nail fold capillaries on both ventral and dor-

Fig. 3. a, b Skiagram of the right hand showing the extra digit arising from the lateral aspect of the first metacarpal. Well-differentiated proximal, intermediate, and distal phalanges can be seen, though the distal phalanx is somewhat shorter (**b**), surrounded by a tubular nail shadow (**a**).

sal aspect of the digit further strengthened the observation of a complete dorsalisation of the palmar aspect of the digit.

CN can be treated by excision of the palmar nail plate and matrix with skin grafting [13]. Reconstructive surgery of the digit may also be undertaken to improve the functioning of the hand. An accessory digit, like in our case, can be totally removed; however, our patient declined any therapeutic and surgical intervention.

Statement of Ethics

Our research paper complies with the guidelines for human studies and animal welfare regulations. The authors state that the subject has given his consent to publish photos and details of the case.

Disclosure Statement

The authors have no conflicts of interest to declare.

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