

Congenital Ectopic Toenail: A Case Report and Review of the Literature

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Congenital ectopic nail is a rare deformity and usually manifests itself as a small abnormal-appearing nail alongside the normal nail on the phalanx. Fewer than 25 cases have been reported. In most of the cases, abnormal nails have been found on the palmar surface of the fifth finger, although abnormalities in the bone are also occasionally present on the ungual phalanx of the involved digit. The etiology is considered to be either a teratoma or a hamartoma, i.e. the ectopic presence of a stray germ cell. However, both theories have difficulty in fully explaining the features of the congenital ectopic nails. We observed the case of a 10-month-old girl with an ectopic toenail on the medial side of her left second toe. The ectopic nail was removed by punch biopsy. A histological examination revealed the presence of undifferentiated nail tissue, including nail, matrix, and nail epithelium. (*Ann Dermatol* 16(3) 109~112, 2004)

Key Words: Congenital ectopic nail, Deformity of the phalanx

Congenital ectopic nail is a rare malformation and usually manifests itself as a small abnormal-looking nail alongside the normal nail on the phalanx¹. Fewer than 25 cases, mostly in Japan, have been reported. We report a case of an ectopic toenail and review the previously reported cases.

CASE REPORT

A 10-month-old Korean girl was found to have an ectopic nail on the medial side of her left second toe, which had been present since birth (Fig. 1A, B). Apart from the ectopic nail, she had been born without any other anomaly. Her family history was unremarkable. The ectopic nail was clipped whenever necessary, just like a normal nail. Macroscopically, a 2.5 mm-wide and 3 mm-long nail plate was

recognizable, while neither flexion disturbance in the distal interphalangeal joint nor depression of the toe tip was observed. Radiographic examination revealed the presence of hypoplasia of the distal phalanx and thinning of the middle phalanx of the left second toe (Fig. 2A, B). The ectopic toenail was removed by a 4mm punch biopsy. The histological characteristics of ectopic nail are similar to those of normal nails. An ectopic nail consists of a nail fold, a nail plate, and nail bed epithelium with no granular layer in the epidermis. The present example was a typical ectopic nail consisting of non-nucleated, nail-like tissue and nail bed-like tissue with no granular layer (Fig. 3). There has been no relapse during the 10 months since the intervention.

DISCUSSION

The first case of congenital ectopic nail on the palmar surface of the right thumb was reported by Ohya in 1931². Since then, a number of such cases have been reported in the field of dermatology. To the best of our knowledge, 23 patients and 28 lesions, including the one described here, have been reported (Table 1)¹⁻²⁰.

Of the patients examined in the reports, 9 were

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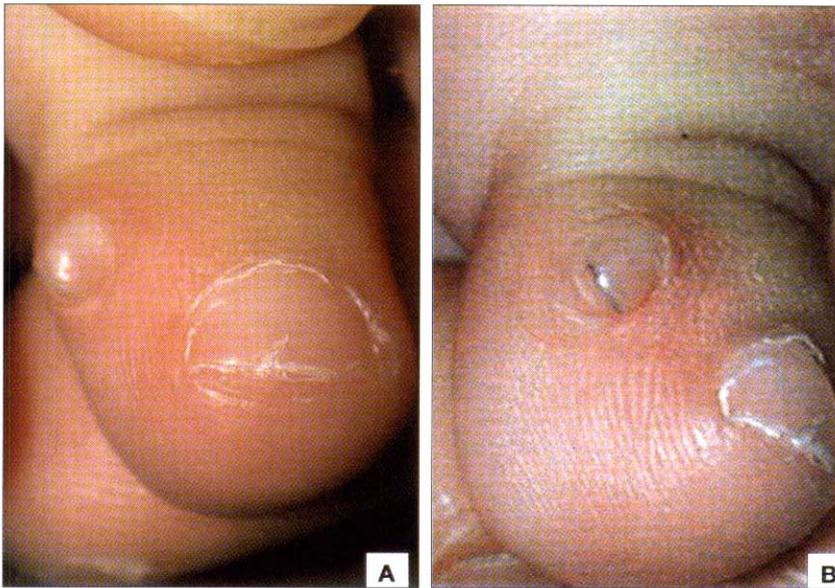


Fig. 1. (A, B) A 2.5 mm-wide and 3 mm-long ectopic nail on the medial side of the patient's left second toe.

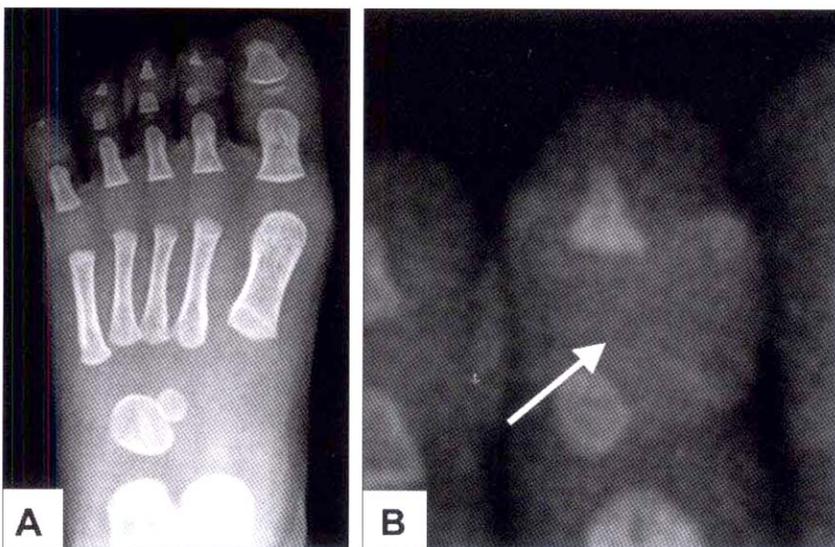


Fig. 2. (A, B) Radiographic examination revealed hypoplasia of the distal phalanx and thinning of the middle phalanx of the left second toe. The arrow indicates an ectopic nail that appeared as a radiodensity-increased soft tissue lesion.

men and 14 were women. Of them, 20 patients with a total of 25 lesions represented ectopic nails on the fingers. The lesions were located on the little finger in 18 instances, on the middle finger in three, and on the thumb in one. It was thus concluded that the most frequent site of occurrence was the little finger. In five of the patients, the lesions occurred bilaterally. In the remaining cases, ectopic nails were observed on the toes of two patients and on the heel of one.

Radiographic examination revealed depressions or other deformities in the distal phalanx in 12 cases.

Thinning of the middle phalanx, as well as hypoplasia of the distal phalanx, of the affected toe were observed in our patient. Undifferentiated nail structures were observed in all of the cases in which histopathological examination was carried out. In most cases, the lesions were treated via simple resection, but flap transpositions were occasionally performed; no instance of recurrence was reported.

Various theories have been proposed regarding the etiology of ectopic nails. However, no consensus has emerged to date³. Ohya² put forth the embryonic aberration theory, the polydactylism theory, and the

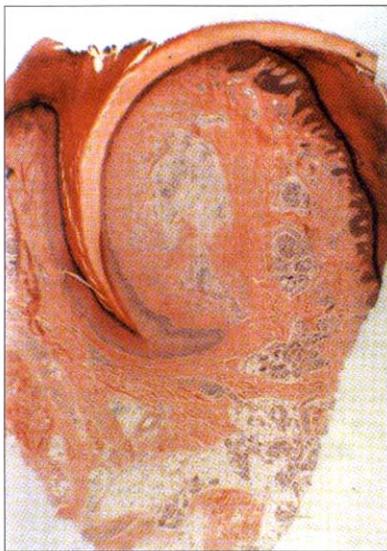


Fig. 3. An ectopic nail consists of a nail fold, and non-nucleated nail plate and nail bed epithelium with no granular layer in the epidermis (H&E, original magnification $\times 10$).

theory of abnormal keratinization leading to changes in the nail. Kikuchi et al⁴. and Higashi et al⁵. argued that congenital ectopic nails are a form of polydactyly on the grounds that polydactyly occurs predominantly on the thumb or little finger, that ectopic nails always occur on the distal phalanx, that ectopic nails often occur on more than one finger or toe, and that they are sometimes observed in members of the same family. On the other hand, Isago et al⁶. supported the embryonic aberration theory, since ectopic nails are usually located on the ventral surface of the finger tip, while polydactylism occurs more frequently on the thumb.

Although some patients exhibited no skeletal abnormality, a number of patients, including our case, exhibited with thinning or hypoplasia of the distal phalanges^{1,3,6-14}. Such aberrations were gradually resolved, after resection of the ectopic nails, suggesting that the deformities were secondary to the presence of the nails³. It is suggested that the

Table 1. Review of the Previously Reported Cases of Ectopic Nail

No.	Author	Year	Sex/Age	Location	Bone deformity
1	Ohya	1931	F/21	Rt. 1st finger, palmar side	not checked
2	Kihuchi	1978	M/40	Rt. 5th finger, palmar side	-
3	Kikuchi	1978	F/55	Rt. 3rd finger dorsal side	-
4	Miura	1978	F/7	Lt. 4th fingers palmar side	+
5	Miura	1978	M/4	Lt. 4th finger palmar side	+
6	Kikuchi	1984	F/55	Lt. 3rd finger ulnar side	-
7	Yamasaki	1984	F/5	Lt. 5th finger palmar side	+
8	Katayama	1984	M/9	Lt. 5th finger palmar side	-
9	Markinson	1988	F/31	Rt. 3rd toe dorsal side	-
10	Aoki	1991	F/34	Bilat. 5th fingers palmar side	not checked
11	Kato	1992	F/64	Lt. 3rd finger palmar side	-
12	Rider	1992	M/18m	Bilat. 5th finger palmar side	+
13	Rider	1992	M/9m	Rt. 5th finger palmar side	-
14	Kopera	1996	M/25	Rt. heel	-
15	Kuniyuki	1996	F/26	Rt. 5th finger, Lt 4th finger palmar side	+
16	Muraoka	1996	M/4	Rt. 5th finger palmar side	+
17	Tomita	1997	F/23	Rt. 5th finger tip	+
18	Kimura	1997	M/25D	Bilat. 5th fingers dorsal side	+
19	Al-Qattan	1997	F/5	Rt. 5th finger palmar side	+
20	Iida	1997	F/43	Bilat. 5th finger palmar side	+
21	Kamibayashi	1998	F/6	Lt. 5th finger palmar side	+
22	Kamibayashi	1998	M/4	Rt. 5th finger palmar side	+
23	Our case	2003	F/10m	Lt. 2nd toe medial side	+

operation should be performed as early as possible, before marked deformity of the involved phalanx sets in.

We describe a case of congenital ectopic toenail and suggest that the operation should be performed as early as possible, before deformity of the phalanx becomes marked.

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