Ectopic Nail
- Report of a Case and Review of the Literature -

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Ectopic nail is a rare disorder of nail organs, which can be divided into congenital and acquired type. The cases of acquired type are more rare than that of congenital type and frequently caused by inoculation of nail matrix by injury or trauma. The structure of the ectopic nail is similar to the ordinary nail, but it usually grows vertically. There are conflicting opinions concerning why the normal nail grows outward instead of upward. Herein we reported a 67-year-old man with a vertically growing ectopic nail at the plantar aspect of the right big toe without prior definite injury to explain inoculation of a nail matrix into the regional skin. Histologically, the ectopic nail inserted almost vertical to the surface epidermis at root. Roentgenographically, there were no abnormal findings of the distal phalanx of the affected toe. (Dermatol Sinica 22 : 183-186, 2004)

Key words: Ectopic nail, Nail matrix, Bone deformity

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Accepted for publication: December 30, 2003
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INTRODUCTION

An ectopic nail is an extra nail in an abnormal site and is a very rare deformity.\(^1\)\(^-\)\(^4\) It can be divided into congenital and acquired type, and the cases of acquired type are more rare than that of congenital type. The structure of the ectopic nail is similar to the ordinary nail, but it usually grows vertically. Conflicting opinions concerning why the ordinary nails grow out instead of up have been discussed by Kligman and Baran.\(^5\)\(^-\)\(^6\) We reported another case of acquired vertically growing ectopic nail and reviewed the literature, and why the nail grows outward instead of upward will be discussed.

CASE REPORT

A 67-year-old male had a protruding hard keratotic, asymptomatic horn on the plantar aspect of right big toe for 7 years. The lesion had repeatedly grown forward after clipping, as with ordinary nails. He did not remember any injury or trauma to the regional skin. His family history was noncontributory. Reviewing his medical history, he suffered from one episode of cerebral vascular attack one year ago. Physical examination revealed a 0.8 x 0.3 cm hard nail bend forward distally and obliquely inserted into the pulp of the right big toe. (Fig. 1) In the X-ray film, there was no abnormal finding such as Y-shaped bifurcation at the distal phalanx of right big toe. Under the impression of accessory rudimentary nail, the lesion including the hard structure and underlying dermis was excised en bloc.

Histological examination revealed faintly eosinophilic, regularly stratified, thick, keratinous layers (an ectopic nail plate) in the center of a cul-de-sac composed of squamous epithelium. (Fig. 2) The ectopic nail bent toward distally and inserted almost vertical to the surface epidermis at root, precisely speaking, the angle was 80°. Under the root of the ectopic nail plate, there were stratified squamous layers without definite granular layers. It invaginated downward into the dermal tissue. The features of these stratified squamous layers were character-

Fig. 1
A hard, keratotic, asymptomatic horn on the plantar aspect of right big toe.

Fig. 2
An ectopic nail plate showed faintly eosinophilic, regularly stratified, thick, keratinous layers. The cul-de-sac, composed of the epidermis, was directed vertically to the skin.

Fig. 3
The turning point between the nail matrix and proximal nail fold-like epidermis is identified.
istic of a nail matrix. The squamous epithelium on the lateral wall of a cul-de-sac was composed of deeply eosinophilic, thick, keratinous layers, analogous to the cuticle of a proximal nail fold. This epithelium displayed several distinct granular layers, such that the turning points between the nail matrix and proximal nail fold-like epidermis could be identified. (Fig. 3) After surgical resection, there was no recurrence of the ectopic nail during 6 months' follow-up period.

DISCUSSION

Ectopic nail (onychoheterotopia) is a relatively rare disorder, which can be divided into congenital and acquired type.

There have been fewer than 20 cases of congenital ectopic nail described in the English literature. All the congenital ectopic nails are present since birth and they can occur in any position of the distal phalanx of the fingers or toes.1,2 Some of these cases may share a common triad of a ectopic nail, absent flexion and X-ray abnormalities of the distal phalanx and are described as "congenital palmar nail syndrome";4,7 some may associate with either absent flexion1 or radiological abnormalities;9 however, most cases show normal function and X-ray appearance of the underlying bones. It is dependent on whether or not the nail matrix contact with the ungual phalanx periosteum, which can impede intramembranous ossification and deform the bone shape.10

Histologically, both light and electron microscopy show that the congenital ectopic nail matrix bears structural similarity to the normal nail matrix.2 However, a study discussed about the content of the congenital ectopic nail demonstrated that the component of amino acids in the ectopic nail were different from those in the normal nail.11 This suggested that a congenital ectopic nail is not merely a smaller version of a normal nail.

With regard to pathogenesis, although one author considered the congenital ectopic nail might be a kind of rudimentary polydactyly,4 most authors suggested that it developed from stray germ cells (i.e. a teratoma or hamartoma), because there did not show a Meissner body and nerve bundle among the ectopic nails.1,3,12

In contrast to the congenital type, the cases of acquired ectopic nail were more rare. After review the English literature, only two reports discussed about cases of acquired ectopic nail.13,14 The clinical appearance and histological findings of the acquired ectopic nails were similar to those of congenital type, but they presented at adult life rather than at birth.13-15 Though the exact pathogenesis was unknown, most authors considered the acquired ectopic nail was caused by inoculation of nail matrix due to injury or trauma.13 In our case, the patient did not remember any obvious trauma to the nail matrix, but he was a farmer and usually walked barefooted. It is possible to inoculate a nail matrix after unnoticed injury.

Interestingly, the direction of the growth of the acquired and most congenital ectopic nails is vertical, which is different to that of normal nails.10,14,15 The whole subject of direction of normal nail growth remains questionable and worthy of further consideration. On this subject, studies of ectopic nail may provide helpful information. According to Kligman,5 the cul-de-sac in which the nail is formed determines that the nail should grow outward instead of upward. He autografted a 5-mm punch specimen from the thumbnail matrix onto the forearm, and the transplanted nail matrix produced a vertical column of nail because there was no cul-de-sac to direct it. This view was questioned by Baran,6 who found that any change in the cul-de-sac might result in change of the nail plate, so he insisted that the nails on abnormal phalanges are abnormal. Kikuchi et al.15 reported a congenital vertically growing ectopic nail and agreed with Kligman's view. Kato14 presented a case of acquired vertically growing nail, whose nail had proximal nail fold but no proper nail bed. Since he considered it may be that the absence of a nail bed, rather than the absence of a proximal nail fold (or cul-de-sac), promoted upward growth of a nail plate instead of outward growth. In our case, the ectopic nail protruded.
vertically at root, and bend caudally on the distal part. Histologically, this entire nail structure was composed of a cup-shaped cul-de-sac, a nail plate, and a circumferentially located proximal nail fold-like epidermis; however, no nail bed structure was identified. So we agreed that the nail bed was critical for the nail to grow outward instead of upward.

In conclusion, we reported the second case of acquired vertically growing ectopic nail. Roentgenographically, there were no abnormal findings of the distal phalanx of the affected finger. After surgery, there was no recurrence noted during follow up.

REFERENCES