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Case Report

Congenital Ectopic nail of a Thumb Case Report and Review of literature

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Abstract:

Ectopic nail Onychoheterotopia or is a rare condition in which nail tissue is found away from the common nail unit of the digits of the hands or the feet. Onychoheterotopia was first reported by Ohya in 1931. It can be congenital or acquired. The acquired ones are mostly post-traumatic. The congenital ones are very rare. Reported congenital cases often involved the toes. The ones reported in the hand affect the little or ring fingers. Congenital ectopic nails of the thumb are extremely rare. We report a case of congenital onychoheterotopia involving the thumb with varus deformity of the phalanx. Complete surgical excision is important in order to prevent its return. We review the literature and the possible embryological origin of this rare condition and its associated bony deformities and its likely outcome.

Key words: bone deformity; congenital; ectopic nail; onychoheterotopia; thumb; trauma

Introduction

A-9-year old girl of non-consanguineous parents presented with an extra nail on the dorsal side of her right thumb. The deformity was first noticed at birth with no associated other congenital abnormality. The parents sought medical advice. They were reassured and advised on surgical excision at an older age when she grew in size to facilitate safe surgery. Prior to the restart of her school, the parents sought surgical removal for social and psychological reasons as she has no functional disability from it. There was no family history of similar or other congenital abnormality been reported.

Clinical examination revealed the accessory nail on the radial and dorsal aspect of the right thumb. There was a radial deviation of the terminal phalanx. There was a full range of movement of the thumb with no functional loss (Figure.1, 2). No other associated congenital abnormalities were identified.

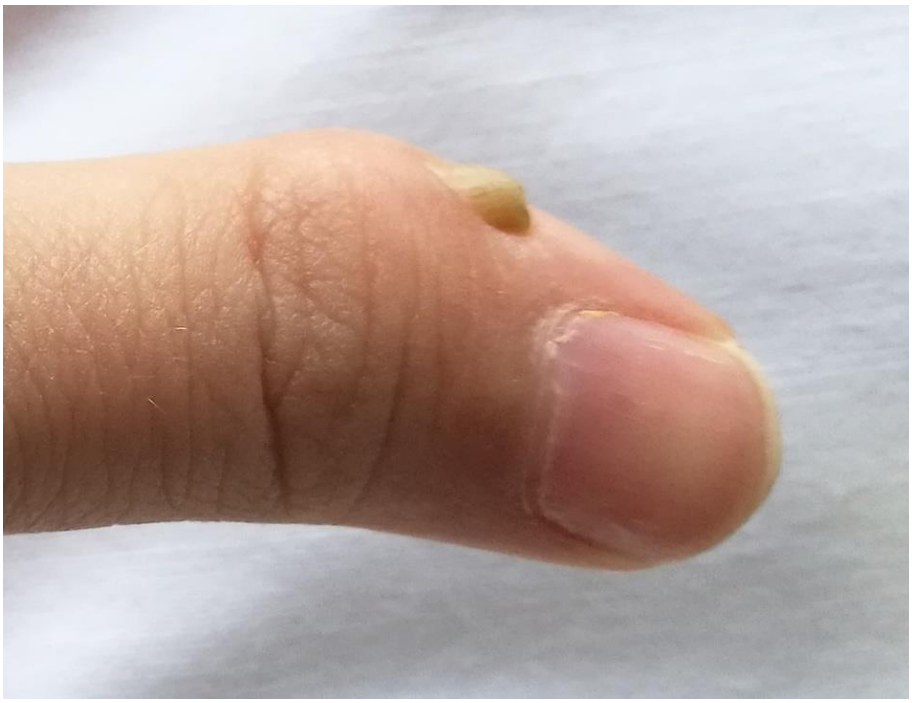

Figure 1: Photo AP Rt. Thumb Showing Congenital Radial Accessory nail

Figure 2: Lateral photo of the thumb.

X-ray of the thumb showed the accessory nail without bony features. There was a 10 degrees of varus deformity of the terminal phalanx (away from the accessory nail) with asymmetrical epiphysis (Figure.3&4).

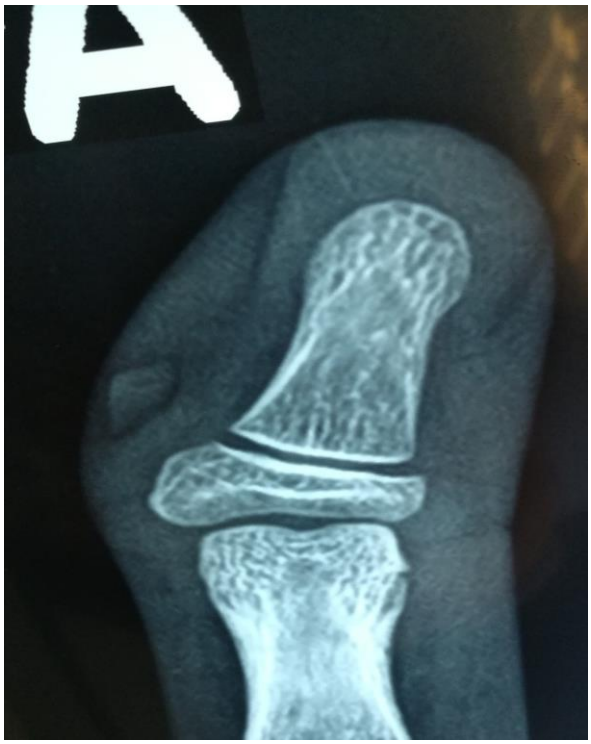

Figure 3: AP x-ray view of the thumb with the ectopic nail without direct bony involvement and 10 degrees of varus deformity away from the ectopic nail and asymmetric epiphysis lager on the ectopic nail side

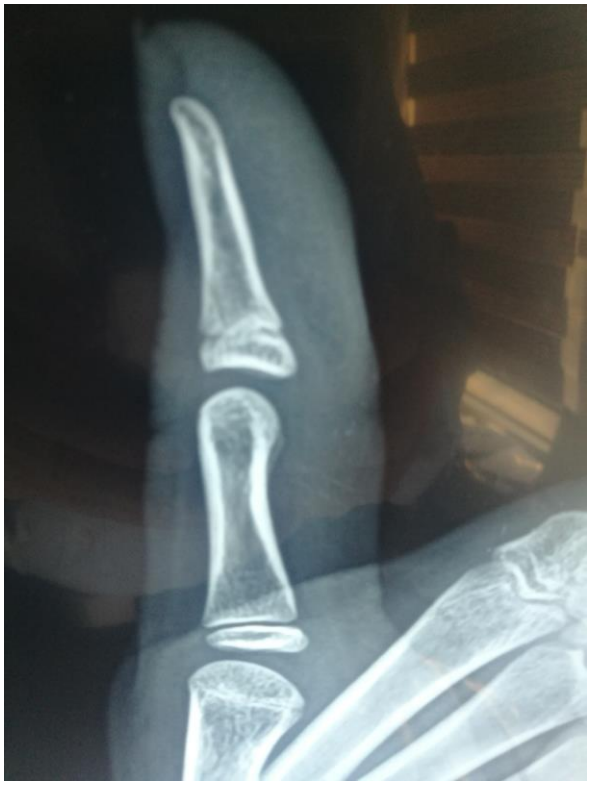

Figure 4: Lateral X-ray Rt. Thumb with no bony deformity

The nail was excised by an elliptical incision with clear margins down to the joint capsule under general anaesthetic. The wound subsequently healed fully.

Histological examination of the accessory nail and surrounding tissue showed increased vascularity at the base of the removed accessory nail which may be the cause of the increased asymmetrical growth of the epiphysis.

At one year follow up, the wound healed fully with full return of the use of the thumb.

Discussion

Onychoheterotopia or ectopic nail is a rare condition in which nail tissue is found away from the common nail unit of the digits of the hands and the feet. It was first described by Ohya in 1931 and he named it *polyoonychie congenita sine ploydactylie*, however, the term ectopic nail was used by Kikuchi in 1978[2]. The feet are the commonest site with the 5th digital been most affected. In the hands, they are rare with the fifth digit of the hand been most affected. Acquired or post-traumatic ectopic nails are the commonest. [3,4]. Congenital ectopic nails are rare and often seen affecting the toes or the little fingers. They can be either single or multiple [5,6,7,8,9,10]. Ectopic nails affecting the thumb are extremely rare hence our report[7,11].

Ectopic nails can be either single or multiple. They arise spontaneously or after a trauma, they grow more slowly than normal nails and they can recur after incomplete removal [9,12].

The double toenail variety has been reported as a familial inherited type as double little toenails, or inherited accessory nail of the fifth toe. They are onychodysplasia rather than onychoheterotopia with variable genetic expression and different from true ectopic nails [9,12].

The Dorsal dimelia or congenital palmar polyonychia variety is the presence of an ectopic palmar nail at the tip of the finger with evidence of dorsalization of the palmar skin[14].

X-ray radiographs are important to exclude associated bony abnormalities. Several bony abnormalities associated with ectopic nail have been reported. They include bony defect of the terminal phalanx of M or Y variety and fusion of the distal terminal phalanx. The varus deformity with asymmetrical epiphysis in our case has not been previously reported. Natural correction of the deformity after excision of the ectopic nail has not been seen with other bony deformities. [11,15]

The pathogenesis of ectopic nails remains unclear. The presence of ectopic germ cells or nail tissue representing a type of hamartoma or teratoma has been suggested. The possibility of a vestigial nail of occult polydactyl type has also been proposed. Other proposals dyskeratotic skin growing into a nail and as part of inherited syndromes such as Congenital Palmer nail syndrome or Pierre-robin syndrome [4,12,16,17]. However, there is no explanation in the literature to the very rare occurrence in the thumb in comparison to the commoner occurrence in the lesser fingers and toes. Embryological development may provide a future explanation to this difference.

Conclusion

We report a rare case of isolated congenital onychoheterotopia of the thumb. The extremely rare occurrence in the thumb in comparison to the lesser digits may have an embryological reason. The associated mild varus deformity of the distal phalanx could have resulted from the increased vascularity at the base of the ectopic nail causing an increase in the growth of the growth plate and bone tissue on the affected side. There is no evidence that excision of the ectopic nail improves the associated bone deformity.

Declarations

1. Statement of human and animal rights -All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008 (5). Informed consent was obtained from all patients for being included in the study). Approval to carry out this study was gained from Institutional Review Board (IRB)& Research Committee at the Al-Balqa Applied University.

2. Statement of informed consent-Informed consent was obtained from the patient's parents for being included in this study.

3. **Conflict of interest statement,** 'The Authors declare that there is no conflict of interest'.

4. Statement of funding- No funding has been received by any of the authors in relation to this article from anybody.

Declaration of conflicting interests: The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Ethical approval: Approval to carry out this study was gained from Institutional Review Board (IRB)& Research Committee at the Al-Balqa Applied University.

Informed consent: Written informed consent was obtained from the parents of the child for anonymized patient information to be published in this article.

Contributiorship details: Samir Sakka wrote the first draft of the manuscript. All authors reviewed and edited the manuscript and approved the final version of the manuscript

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