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Case Report

A Case Report on Atypical Cleft Hand associated with Syndactyly, at St. Luke Catholic Hospital, Woliso

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Abstract

Introduction: Cleft hand is a complex congenital hand malformation presenting as a central deficiency in which the child has missing one or more fingers of the hand. Usually inherited as autosomal dominant with variable penetrance, but sporadic genetic mutation is also a possibility.

Case Presentation: we present a case of unilateral cleft hand with syndactyly and missing three metacarpals and fingers ray, in a 7 weeks old female infant without family history of similar complaints.

Discussion: Cleft hand is a rare congenital hand deformity with longitudinal loss of one or more of the central parts of the hand including index, middle or ring fingers. It is mentioned as abnormal formation or differentiation of hand plate with complex unspecified axis patterning, under classification of congenital anomalies of the hand and upper limb.

Conclusion: Cleft Hand is a rare congenital hand deformity. It can occur in association with central polydactyly and syndactyly. Early diagnosis and treatment is required to improve function and appearance of the hand.

Keywords: Autosomal Dominant; Cleft Hand; Syndactyly; Congenital Anomalies

Introduction

Cleft hand is uncommon congenital anomaly of the hand presenting as a central deficiency of one or more digits of the hand. The incidence is about 1 in 90,000 live birth and 1 in 20 thousand from the general population. Autosomal dominant inheritance with 70% penetrance in majority of the cases, while spontaneous sporadic genetic mutation is also possible in some cases like in our patient.

There are two types of cleft hands exist, which is known as typical and atypical cleft hand [1]. Typical cleft hands are characterized by deep V shaped deformity, positive family history, commonly associated with cleft feet and are bilateral. Nevertheless, the atypical cleft hands have U shaped cleft, usually has unilateral involvement, and has neither cleft feet nor positive family history. The hand malformation is caused by chromosomal errors that result from exposure to teratogens and maternal metabolic disorders during a critical moment of embryonic development.

Case Presentation

A 7 weeks old female infant who was born to gravida VI, para-V, and no abortion mother, presented to orthopaedic department with fused and decreased numbers of left-hand digits. The child was born at term with birth weight of 2.8 Kg and APGAR score of 8. Apart from radial ray deficiency, she had been born without any other anomaly. Her family history was unremarkable.

On physical examination her left hand has three completely fused digits with respective separate nail, wrist in neutral position and no limb length discrepancy identified (Figure 1). No visible abnormality on her back and lower limbs. On Passive range of motion intact interphalangeal, metacarpophalangeal, wrist, unrestricted supination and pronation of forearm, elbow and shoulder joints.

For this we took x-ray and it shows, only two metacarpal bones and loss of entire index, middle and ring ray, with simple syndactyly of thumb and ulnar rays (Figure 2). There is associated duplica-

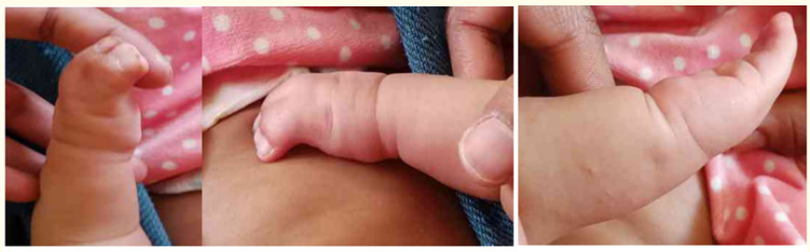

Figure 1: The baby hand pictures; a) raial side, b) dorsal side, and c) ulnar side.

tion of distal phalanx of radial ray with respective nail, and absent all carpal bones. Radius, ulna and humerus looks normal. There is no bony connection between the digits of the two metacarpal bones signifying simple syndactyly. She has no family history, her left hand and both foots are not involved.

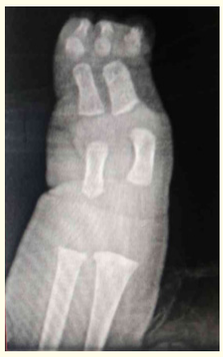

Figure 2: X-ray of the child; showing deficiency of metacarpal and phalanxs.

For this case the following reconstructive surgery plan are needed when the child is beyond 1 year of age.

Surgical release of the syndactyly with zigzag incision on both dorsal and volar surface dividing the fingers and resurfacing the surgical wound with a well-vascularised dorsal trapezoid flap, interdigitating fasciocutaneous flap, and full thickness skin graft to resurface inter digital spaces.

Discussion

Cleft hand is a rare congenital hand deformity with longitudinal loss of one or more of the central parts of the hand including index, middle and ring fingers. Although majority of cases are bilateral and inherited as autosomal dominant with variable penetrance, some of them may arise as a new genetic mutation in a child (de novo mutation) even when the parents are not affected like in our patient.

According to Oberg, Manske, and Tonkin (OMT) [2], classification of congenital anomalies of the hand and upper limb, cleft hand is mentioned as abnormal formation/differentiation of the hand plate with complex unspecified axis patterning and accepted by international federation of society for surgery of the hand (IFSSH) as optimal classification [3].

On most medical journal clinical studies shows that cleft hand is closely related with central polydactyly and syndactyly. Manske (1983:906-8) [4], reported four hands of a pair of identical twins demonstrated manifestation of both cleft hand deformity and central polydactyly. Sateke., *et al.* reported central polydactyly, syndactyly, and cleft hand in five hands of three patients from the same family [5].

Beside these clinical studies, experimental studies are also reported. Ogino et.al has investigated the teratogenic relationship between polydactyly, syndactyly, and cleft hand by inducing the same deformity using myleran in rat foetuses [6].

Phenotypically Various classification system of cleft hand are reported. Barsky [7], defined cleft hand as typical and atypical. The typical cleft hand was characterized by a deep V-shaped defect in the central part of the hand, often bilateral, positive family history, and associated with syndactyl/polydactyly. While atypical type frequently unilateral, spare lower limb, and sporadic (de novo mutation). Our case shows similarity with the atypical cleft hand in terms of being unilateral and having no family history. Manske and Halikis [8], classify based on the first web space and it is the most commonly used classification system for surgical intervention when indicated. They grouped into five entities considering progressive reduction of thumb web space, of which our patient best fit into type V, which is characterised by absent thumb web space, suppressed thumb elements and remain ulnar ray.

Conclusion

Cleft hand is an uncommon congenital anomaly of the hand. It can present in association with central polydactyly and syndactyly. Early diagnosis and intervention help the child to have functionally and aesthetically good hands.

Ethics and Consent

Consent was obtained from the family of the child for publication of this case report with accompanying images and the hospital also approved to publish the case.

Disclosure

There is no conflict of interest among the authors in this work.

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