Unilateral Cleft Hand with Cleft Foot

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Abstract:
Congenital anomalies of the hand form an important class of congenital malformations. They have a huge functional importance because of the part played by the hand in the daily activities of a person. The deformities also have significant cosmetic significance and may also be associated with other anomalies. Amongst the congenital anomalies, central deficiency or cleft hand is relatively rare. The association of cleft foot with cleft hand is an even more rare occurance. We present a case report of a 6 year old child, born of a non-consanginous marriage, having congenital central deficiency of ipsilateral hand and foot.

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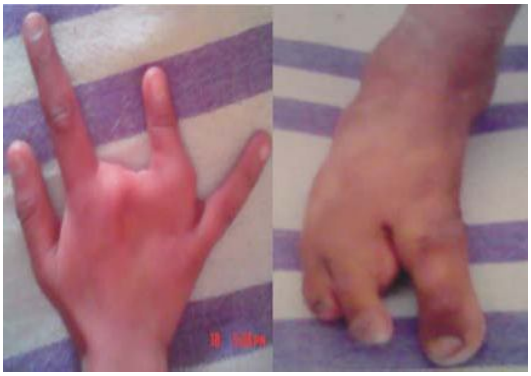
Introduction

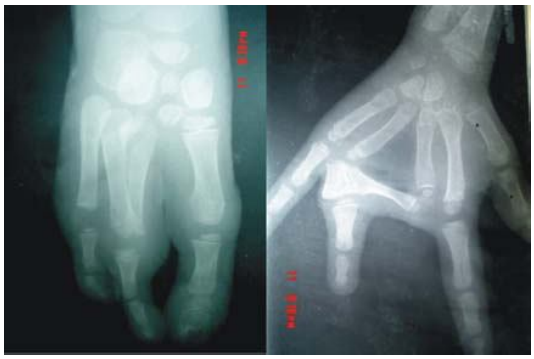
Case Scenario

A 6-year old male child, born of a non-consanguineous marriage, was referred from the Department of Pediatrics & Neonatology, SKIMS to the Department of Orthopedics, SKIMS Medical College with a deformity of the right hand and right foot since birth. The main complaint of the patient was inability to hold things properly in the hand and difficulty in wearing slippers. The patient was born full-term by normal vaginal delivery. The antenatal and the peri-natal history did not reveal any significant problem. The general and systemic examination of the child was normal, except for the abnormalities in the hand and the foot. The child had not received any sort of treatment for this condition.

The right hand of the child showed the absence of middle finger. The bases of the index and the ring fingers were connected by a bony bridge. The index finger was longer than its counterpart in the normal hand. The thumb web was mildly narrowed. The hand was of the same size as the opposite hand. The thenar and hypothenar muscles showed wasting. The wrist and the forearm were normal. The wrist and elbow joints had normal range of motion. The movement of the metacarpo-phalangeal joints of index and ring fingers was restricted. The distal neurovascular status was normal. The right foot showed a deep cleft with absence of second and third toes. The ankle and hind foot was normal in appearance and movements. The patient had normal hair and normal dentition. Ophthalmological examination was normal. The child did not have cleft palate or lip.



The radiograph of the right hand revealed absence of all the phalanges of the middle finger. The gap between the third and fourth metacarpals was increased with the latter being smaller in size. A transverse bone was seen lying in the base of the cleft. The proximal phalynx of the fourth metacarpal was short and thick and formed a pseudo-joint with the cross-over bone. Radiographs of the right foot showed absence of the second metatarsal and all the phalanges of the second and third toes. Further, the distal phalynx of the great toe was short and broad. Skin biopsy of the patient did not show any evidence suggestive of ectodermal dysplasia.



Considering the clinical and radiological picture a diagnosis of unilateral cleft hand with cleft foot was made. The parents were offered surgical treatment but they were unwilling to go for surgery.

Discussion

Cleft hand refers to a group of congenital hand abnormalities in which the central bony elements are missing. It is the fifth most common congenital anomaly of the hand [1]. The condition is known by different names like ectrodactyly, pincer split hand, lobster claw hand, crab claw hand and split hand complex. However, cleft hand is now the most widely accepted name for this condition [(2, 3)].

Cleft hand has many variations, but the absence of the central portion of the hand is the main feature of the condition. Deficiency varies from absent long phalanges to absent individual finger, to monodactyly or even the absence of all digits [(4, 5, 6)]. The depth of the palmar cleft depends upon the remaining metacarpal bones. If present, the cleft is shallow and when absent, it is deep. Wider clefts are associated with more adducted and deficient thumb [(7)].

Lange has divided cleft hand into two types – typical and atypical. The incidence of typical cleft hand is 1:90,000 births and 1:120,000 population. Atypical cleft hand occurs in 1:150,000 births and 1:200,000 population [8]. Blauth and Falliner collected 303 cases from literature, including their 35. Bilateral cleft hand was seen in 163 (56%) and unilateral in 140 (44%). The association of cleft foot is even less, with less than one-third of having this deformity.

Flatt has described cleft hand as a functional triumph and a social disaster [9]. The typical cleft hand has a fissure in the palm and is almost normal in size. Typical cleft hand arises due to a failure in the normal process of formation of interdigital spaces which occurs at Carneige stage 22 (50 days post-ovulation). Chromosome defects associated with cleft hand have been mapped to two different loci, viz. 7q21.2-q21.3 and 10q25 [10, 11]. The fingers adjacent to cleft are longer and thicker. Cross-bone is often seen lying in base of the cleft. Angiography shows digital vessels are normally developed but variable in number. Typical cleft hand can be unilateral or bilateral. The former is usually sporadic. Bilateral cleft hand has a familial tendency and 50% are hereditary with dominant inheritance.

Atypical cleft hand is now better known by the name symbrachydactyly [12]. Atypical cleft hand arises due to a failure in the formation of bones in the digit. The condition is usually unilateral and the feet are not involved. Generally more than two central digits are involved in the condition. Rudiments of missing finger are present with some active movement. The condition does not show any inheritance pattern and is not associated with any other associated anomaly.

Cleft hand is often associated with syndactyly or polydactyly in the same hand (in unilateral cases) with syndactyly of thumb-index being common. Bony fusion may be present. Anencephaly, cleft lip, cleft palate, clinodactyly, scoliosis, imperforate anus, anonychia, cataracts and deafness may be associated. Other musculoskeletal anomalies including hypoplasia or pseudoarthosis of clavicle, absent pectoralis major muscle, short humerus, synostosis of elbow, short forearm, absent ulna, radioulnar synostosis, bilateral absence of tibia, bilateral dislocation of hip, short femur, hypoplastic patella, clubfoot, calcaneovalgus, deviated nasal septum and congenital ptosis may also be seen [13, 14]. Genitourinary system anomalies are also sometimes seen. A number of syndromes may be associated with cleft hand [15]. Some patients may present as ectrodactyly, ectrodermal dysplasia- clefting (EEC) syndrome which is a combination of typical cleft hand, cleft lip and palate and an atypical form of ectrodermal dysplasia. EEC may also be associated with abnormalities of dentition, lacrimal duct abnormalities and hair changes [16].

The management of cleft hand is surgical, with prosthesis no longer being used, except for cosmetic purposes. The principles of surgery are good pinch and grasp, followed by acceptable cosmesis. Surgical reconstruction involves cleft closure, syndactyly release, thumb adduction correction and removal of transverse or deforming bones. Syndactyly needs to be released first, with border digits by 6 months and central digits by 18 months. After six months, cleft is closed, either alone or combined with thumb adduction deformity. Snow and Litttler [17] described a technique of simultaneous correction. Miura and Komada [18] technique is a technically simple method of correction of both cleft and thumb adduction.

In the atypical type pinch is difficult due to hypoplasia of thumb or little finger. The management involves deepening of the web by Z-plasty in the first stage with removal of redundant bone segments or rudimentary digits. Later a metatarsal osteotomy needs to be done to improve grasp. Tendon transfers, when needed, should be delayed until the patient reaches 3 years of age.

Management of cleft foot depends upon the deformity. Abraham's classification [19] is widely used for managing cleft feet. Type I having a central ray or deficiency (usually 2nd or 3rd ray) extending upto mid-metatarsal level being managed with soft tissue syndactylism and partial hallux valgus correction, if needed. Type II has a deep cleft up to the tarsal bones with forefoot splaying, for which soft tissue syndactylism with first-ray osteotomy is needed. Type III is a complete absence of the first through third or fourth rays, for which surgery is not required.

Whatever the type of cleft hand, the management of this condition must start at an early age to achieve good functional results. The obstetricians and the pediatricians must be aware of this condition and early referral to orthopaedician may help in achieving a good cosmetic and functional result.

References

1. Tsuge K, Ischii S, Lieba Y. Report of the Japanese Society for Surgery of the Hand, Committee on Congenital Malformations of the Hand. Orthop Surg (Jpn) 1980; 31:1959-1963.
2. Swanson AB. A classification for congenital hand malformations. J Hand Surg 1976; 1: 8-22.
3. Wood V. The cleft-hand (Central deficiencies) In: Green DP, ed. Operative Hand Surgery, Churchill-Livingstone; New York, 1982: p233-243.
4. Maisels DO. Theory of Pathogenesis of Lobster Claw deformities. Hand 1970; 2:79.
5. Kato S, Ischii S, Ogio T, Sciono H. Anamolous hands with cleft formation between 4th and 5th digits. J Hand Surg 1983; 8: 903-913.
6. Miorra T. Cleft hand involving only the ring and small finger. J Hand Surg 1988; 13A: 530-535.
7. Manske PR, Halikis MN. Surgical classification of central deficiency according to the thumb web. J Hand Surg 1995; 20A: 687.
8. Tertarry SA, McKuisk VA. Absence deformities as isolated malformations. In Bergsma D, Mudge JR, Paul NW, Greene Sc, (Eds). The Genetics of Hand Malformations. New York; Alan R Liss, Inc, 1978F: 36.
9. Flatt AE. Cleft hand and central defects. In Flatt AE (Ed). The care of congenital hand anomalies. St Louis, MO: Quality Medical Publishing 1994; 337.
10. Buss PW. Cleft Hand/Foot: Clinical and developmental aspects. J Med Genet 1994; 31-72.
11. Rass-Rothchild A, Manouvrier S, Gonzales M, et al. Refined mapping of a gene for Split Hand- Split Foot malformation (SHFM3) on chromosome 10q25. J Med Gent 1996; 33: 996.
12. Manske PR. Symbrachydactyly instead of Atypical cleft hand (letter). Plas Reconst Surg 1993; 91: 196.
13. Miura T, Nakamura R, Suzuki M, Watanabe K. Cleft hand, Syndactyly and Hypoplastic thumb. J Hand Surg 1992; 17B: 365.
14. Ogino T. Teratogenic relationship between polydactyly, syndactyly and cleft hand. J Hand Surg 1990; 15B: 201.
15. Tertarry SA, McKuisk VA. Absence deformities as isolated malformations. In Bergsma D, Mudge JR, Paul NW, Greene Sc, Eds. The Genetics of Hand Malformations, New York: Alan R Liss, Inc 1978:73.
16. Thakkar S, Marfatia Y. EEC syndrome sans clefting: Variable clinical presentations in a family. Indian J Dermatol Venereol Leprol 2007; 73: 46-48.
17. Snow JW, Littler JW: Surgical treatment of cleft hand. Transactions of the International Society for Plastic Reconstructive Surgery, Fourth Congress, Rome. 1967, Excerpta Medical Foundation.
18. Miura T, Komada T: Simple method for reconstruction of the cleft hand with an adducted thumb. Plast Reconstr Surg 1979; 64: 65.
19. Abraham E, Waxman B, Shirali S, Durkin M: Congenital cleft-foot deformity treatment. J Pediatr Orthop 1999; 19: 410.