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Cleft foot and hand syndrome: A case report

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Abstract

Cleft foot and hand (Split or Lobster foot and hand) or Partial Adactyly is a rare inherited anomaly in which a single cleft extends proximally into the foot or hand. There is failure of development of the central digital rays of hand or foot to a variable extent. It is characterised by hypoplasia/ aplasia of the phalanges, toes, metacarpals and metatarsals. The presentation may be an isolated anomaly or associated with a syndrome. It usually occurs in conjunction with clawing of the hand. We report a case of bilateral cleft feet and cleft hands with triphalangeal thumbs, as such cases are rare to come across.

Keywords: Cleft foot, cleft hand, split foot and hand, lobster foot and hand

Introduction

Split-Hand Split-Foot Malformation (SHFM) also known as Ectrodactyly or cleft hand/foot, is rare congenital disorder. Birch-Jensen estimated the incidence of cleft hand deformity at 1 in 90,000 births. More recent studies record the incidence of central ray anomalies to be closer to 1 in 10,000 (approximately 0.7 per 10,000 live births) [9]. There is a median cleft in the hands or feet, often called as "lobster claw". It may be unilateral or bilateral, occurring as an isolated anomaly or may be associated with a syndrome. Majority of the cases live a normal life with very little effect on limb function [8].

Isolated variety (SHFM-1) is a type I failure of formation - longitudinal arrest with transverse terminal deficiency affecting the central digital rays. There are no proximal deficiencies of the bones, muscles, tendons, vessels or nerves. It has autosomal dominant inheritance. It is the only form associated with sensorineural hearing loss [8].

We report a case of bilateral cleft feet and cleft hands with triphalangeal thumbs, along with a brief review of literature.

Case Report

A 2 and half year old male child was brought with a history of malformed hands and feet since birth. He was first in birth order, born out of a non-consanguineous marriage. There was no family history suggestive of any bony deformity. There was no history of drug intake or exposure to radiation during antenatal period.

Examination revealed presence of four digits in both hands. A deep V-shaped cleft was present due to absence of central digital ray, separating bony fusion or syndactyly of first and second metacarpals from syndactyly of ring finger and little finger (Fig. 2). Feet revealed a deep cleft extending proximally, involving distal one third of feet with normal great toes and syndactyly of third, fourth and fifth toes on both sides. Second toe on both sides was missing (Fig. 1). No other skeletal and systemic abnormality was detected on detailed examination. There were no other midline defects like cleft lip or cleft palate.

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Fig 1: Both feet show a deep cleft in place of an absent 2nd toe and metatarsal. There is syndactyly of third, fourth and fifth toes. No abnormality or absence of tibia seen.



Fig 2: Both hands show a deep V-shaped median cleft separating syndactyly of 1st and 2nd fingers from that of 4th and 5th fingers. There is absence of 3rd finger and metacarpal.



Fig 3: X rays of both feet showing normal great toes, absence of 2nd metatarsal and phalanges and absence of phalanges of 3rd toe. There is a deep cleft separating the great toe from the fused 3rd, 4th and 5th toes. Tarsals, tibia and fibula appear normal.



Fig 4: X rays of both hands showing absence of metacarpals and phalanges of middle finger, replaced by a deep cleft separating the fusion of first 2 fingers from that of the last 2. Both hands have triphalangeal thumbs.

Discussion

Split hand foot syndrome (SHFM) or Ectrodactyly is a syndrome of limb malformation which affects central rays of hand and foot. It may be autosomal dominant, autosomal recessive or X-linked inheritance, depending on chromosomal mutation.

The presentation of SHFM can vary from syndactyly, aplasia or hypoplasia of the phalanges, metacarpals and metatarsals, and median clefts of the hands and feet. Severity differs not only between patients, but also between limbs of an individual.

SHFM is of two types: Syndromic and Non-syndromic. The syndromic type may be associated with tibial aplasia, mental retardation, ectodermal and craniofacial defects, cleft lip or palate and deafness, while the Non-syndromic form presents only with limb defects.

Barsky [1] described two types of cleft hand - Typical and Atypical. Typical cleft hand has a deep palmar cleft which separates the two central metacarpals. One or more rays are absent, and the existing digits tend to be confluent and of unequal length. In the Atypical cleft hand, the central rays are absent and only short radial and ulnar digits remain with a shallow cleft.

In cleft feet, generally one or more toes and parts of their metatarsals are absent. Often, the tarsals are abnormal. Although the deformity varies in degree and type, the first and fifth toes are usually present. If a metatarsal is partially or completely absent, its respective toe is always absent.

Other associated abnormalities include cleft lip and palate, reduction in number and size of the phalanges, syndactyly, polydactyly, triphalangeal thumb (as in our case). Scalp defect, genitourinary anomalies with atresia of the nasolacrimal duct and buphthalmos [3-6]. Some syndromes associated with this condition are [9].

- EEC (Ectrodactyly–Ectodermal dysplasia–Cleft) syndrome
- Cornelia de Lange syndrome
- Acrorenal syndrome
- Focal Dermal Hypoplasia
- Ectrodactyly and Cleft Palate syndrome
- Ectrodactyly/Mandibulofacial dysostosis
- Ectrodactyly and Macular Dystrophy

The diagnosis of SHFM is based on clinical examination of limbs at birth. X-rays are required to identify the skeletal anomalies. Ultrasonography in antenatal period aids early recognition. Diagnosis may be difficult in neonates with a central ray defect. In such cases, nails and dental X-rays may help in identifying involvement of ectodermal structures. Hair distribution and birth weight should be noted. Electron microscopic examination of the hair shaft may be helpful. These children must be reviewed during the first year of life, to confirm normal development of ectodermal structures. A renal ultrasound scan may help make the diagnosis of EEC or acrorenal syndrome^[9].

In older children and adults with ectrodactyly, minor associated ectodermal anomalies are seen, especially of the teeth. Language milestones should be checked.

Treatment is surgical correction for improving function and appearance. Prosthetics are useful to achieve normal functioning. Reconstructive surgery for deformed limb is done when necessary. Other treatment is symptomatic and supportive. Genetic counseling of parents should be done.

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