Postaxial Polydactyly

Aggarwal AN,[1] Goyal MK,[1] Gupta Rishi N[1]
[1]Department of Orthopaedics, University College of Medical Sciencesand Guru Teg Bahadur Hospital, Delshi, India.

ABSTRACT

We present a case of postaxial polydactyly with well formed six digits on left hand and seven digits on right hand. Both conditions are rare and combination of these two conditions even rarer. The patient also had supernumerary sixth right toe and cleft lip. Very few cases of postaxial polydactyly are reported previously.

Key Words: *cleft lip, polydactyly, postaxial, supranumerary toe, syndactyly*

INTRODUCTION

Polydactyly is the most common congenital digital anomaly of the hand and foot. It may appear in isolation or in association with other birth defects. Postaxial polydactyly involves the fifth digit or ray. Postaxial polydactyly can occur by itself, or more commonly, as one feature of a syndrome of congenital anomalies.

Postaxial polydactyly is approximately 10 times more frequent in blacks than in whites and is more frequent in male children. In contrast, postaxial polydactyly seen in white children is usually syndromic and associated with an autosomal recessive transmission.

Postaxial polydactyly in both hands is very rare and along with one foot is even more rare. The combination of all these features along with cleft lip were present in our case.

CASE REPORT

An 18 month, male child presented with bilateral post axial polydactyly of hand (Ulnar side) and polydactyly of right foot. He had six well formed digits in left hand and right foot. Right hand had seven digits with syndactyly between 6th and 7th digits. The child also had cleft lip which was operated upon by a paediatric surgeon at age of six months.

There was no history of similar congenital anomaly in his parents or two siblings and there was no consanguinity. Roentgenograms of the left hand showed 6 digits, each having normal metacarpals and phalanges (Figure 1). Right hand showed 7 digits. Fifth digit had normal phalanges and only head of metacarpal. 6th and 7th digits had a common bifid metacarpal and normal phalanges. There was ulnar deviation of the 6th and 7th digits. Right foot showed six digits with broad fifth metatarsal. Sixth toe had normal phalanges (Figure 2).

The accessory digits of right hand were excised. The bifid metacarpal was shaved off and ulnar collateral ligament reconstruction of the 5th metacarpophalangeal joint was also performed. Accessory digit of the left hand was excised and hypothenar muscles were repaired. Accessory digit of the foot was also excised to enable the child to wear a shoe.

DISCUSSION

Literature on post axial polydactyly is scant. The low observational value, low medical interest, management of mild forms of polydactyly by paediatrician and

Correspondence:
Dr. Manoj Kumar Goyal
Department of Orthopaedics
University College of Medical Sciences
Dilshad Garden, Delhi-110095, India.
Email: drmanojgoyal@yahoo.com
Phone: 11-22134595

obstetricians without proper documentation and, it not being considered a congenital anomaly by some, are a few reasons.

Finley et al in a 2 year prospective study in USA determined incidence of polydactyly (all types) to be 2.3/1000 in white male population, 0.6/1000 in white female population; 13.5/1000 in black male population and 11.1/1000 in black female population. Incidence in Sweden was 1/1000 with equal distribution for males and females[1].

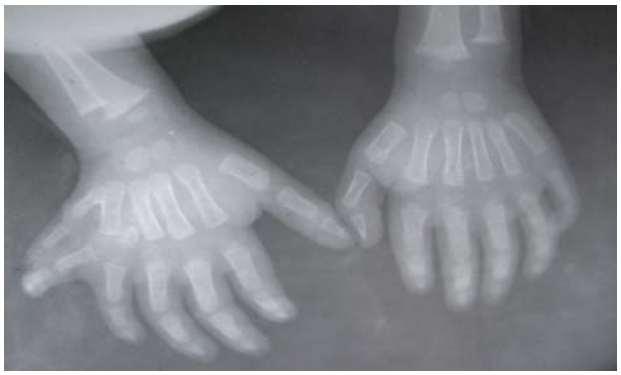

Figure 1. Radiograph of both hands showing 7 digits in right hand with well formed bifid 6th metacarpal. The left hand has well formed 6th metacarpal and phalanges

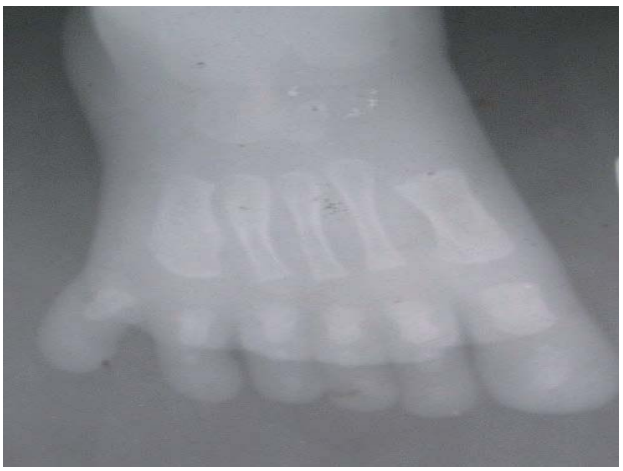

Figure 2. Radiograph of right foot showing postaxial polydactyly with broad 5th metatarsal

Turek and Stelling classified postaxial polydactyly as rudimentary masses devoid of other tissue elements (Type-I); subtotal duplication with some normal elements that typically articulate with a bifid or broad metacarpal (Type-II) or duplication of the entire osteoarticular column including the metacarpal (Type-III). Type III postaxial polydactyly occurs rarely[2].

Temtamy and Mckusik classified postaxial polydactyly as fully developed (Type-A) or rudimentary skin tag (Type-B). Type A polydactyly is rare and an extra digit with separate metacarpal is even rarer[3].

Castilla et al found prevalence of postaxial polydactyly of hand and or foot to be 14.3/10,000 (n = 1,582,289 births). Postaxial polydactyly of hand was 11.0/10,000. Combined prevalence of hand and foot polydactyly was only 1.2/10,000. Only a negligible proportion of isolated 5th digit hexadactyly was expected to belong to type-A; i.e. a proximal (carpal or metacarpal) implanted and functional extra digit[4].

Well formed postaxial polydactyly is inherited as autosomal dominant trait without any associated anomalies in African American infants. However it is associated with many congenital anomalies and syndromes in Caucasians. Castilla et al reported 23.4% cases of associated anomalies[4]. Our case also had cleft lip as an association. Other association can be chromosomal abnormalities (13 trisomy), eye abnormalities (Laurence Moon Bardet Biedl syndrome) and bony dysplasias etc[5].

Seven digits as noted in the right hand is a rare presentation. Castilla et al compared two large studies i.e. Latin American collaboration study of Congenital Malformations (ECLAMC) (n = 3128957 live births from 1963 to 1993), and the Spanish Collaborative Study of Congenital Malformations (ECEMC) (n = 1,093,865 live births from 1976 to 1993 and 7,271 stillbirths from 1980 to 1993). Prevalence of isolated seven or more digits was 1.1/100,00 in ECLAMC study and 2.0/100,000 in ECEMC study. Only one case of seven or more digits in association with other anomalies was noted in ECLAMC study while ECEMC study had 8 cases[6].

REFERENCES

1. Finley WH, Gustavson KH, Hall TM, et al. Birth defect surveillance: Jefferson county, Alabama, and Uppsal County, Sweden. South Med J. 1994;87:440-5.
2. Stelling F. The upper extremity. In: Ferguson AB, editor. Orthopaedic Surgery in Infancy and Childhood. Baltimore: Williams and Wilkins; 1963. p. 304-9.
3. Temtamy S, McKusik V. Polydactyly. Birth defects 1978;14:364.
4. Castilla EE, Dutra MDG, Fonseca RL, et al. Hand and foot postaxial polydactyly: two ssdifferent traits. Am J Med Genet. 1997;73:48-54.
5. Bayne LG, Costas BL, Lourie GM. The upper limb. Morrissy RT, Weinsteen SL, editors. Lovell and Winters Pediatric Orthopaedics. 4th ed. Philadelphia: Lippincott Raven; 1996. p. 781-848.
6. Castilla EE, Fonseca RL, Dutra MDG, Bermejo E, Cuevas L, Frias MLM. Epidemiological analysis of rare polydactylies. Am J Med Genet. 1996;65:295-303.