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A case of post-axial hexadactyly with a different skeletal anatomy in each of the four limbs

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ABSTRACT

Tetrapolydactyly, that is, polydactyly of all four limbs is a rare congenital anomaly. When six fingers are present in each of the four limbs, the anomaly is called "polydactyly 24". Such conditions may occur sporadically or may be part of some syndromes. We here present a rare case of sporadic polydactyly 24 in a healthy female from Eastern India. She had post-axial polydactyly, type II in one limb and type III in the rest. She denied any functional limitation due to this skeletal anomaly. The relevant literature pertaining to polydactyly has been discussed in details.

INTRODUCTION

Polydactyly is a common congenital skeletal anomaly with varied presentations [1]. The supernumerary digit(s) may be present in one hand or foot or, very exceptionally, may be present in all four limbs [1]. The latter condition is termed tetrapolydactyly [2].

The extra digits may be a cutaneous soft tissue projection, or it may be a fully formed digit with skeletal framework and joints [1]. Such polydactyly may occur sporadically, or it may be part of a congenital syndrome like Down's syndrome, Laurence-Moon-Biedl syndrome or Klippel-Trenaunay Syndrome [1]. In syndromic cases, the polydactyly is usually associated with other skeletal and systemic malformations. Sometimes, the sporadic forms of polydactyly may be familial, associated with specific mutations [3].

The presence of 6 digits in each of the four limbs is termed "polydactyly 24" [4]. This is a very rare malformation and surgical correction of the extra digits is often challenging. We here present such a case of Polydactyly 24. Such report is very rare from India [4].

CASE REPORT

A 15 year old female patient was admitted with tubercular meningitis in the General Medicine department. While examining the patient, we noticed that she had six fingers (hexadactyly) in all the four limbs (Figure 1). All the extra fingers were post-axial, well- formed and had no voluntary movement. X ray study of the four hands and feet revealed that (figure 2):

Right hand: the extra finger had two phalanges, but they were not connected to any metacarpal bone (type II polydactyly)

Left hand: the 5th metacarpal had formed a "T" like head. The medial end of this "T" was connected to the extra finger. This extra finger had two phalanges.

Right foot: the head of the 5th metatarsal was wide and had two facets. The lateral facet was attached to the extra finger.

Left foot: the head of the 5th metatarsal was wide, but not as wide as the right side. Still, it had two small articulating facets, containing the two little fingers. In both feet, the extra finger had two phalanges.

Thus, the remaining three limbs had type III polydactyly.

There was no other bony abnormality or supernumerary structure in the carpal or tarsal bones in X ray. The supernumerary fingers had normal nails. Passive movement was possible for the extra digits in the range 10º-20º.

General examination of the patient did not reveal any other skeletal dysmorphism or internal organ involvement. Her mental development history was normal with normal IQ. There was no family history of similar disorder. Her birth history was

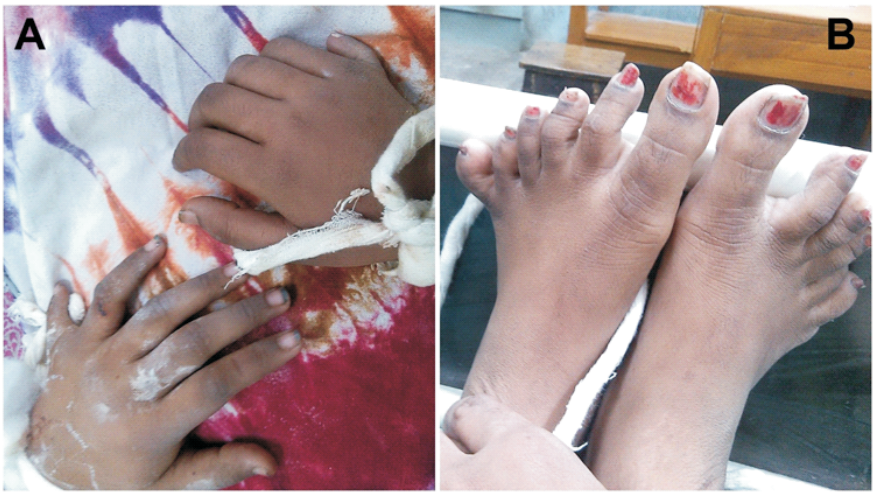

Fig. 1. : Figure showing hexadactyly of hands (A) and feet (B)

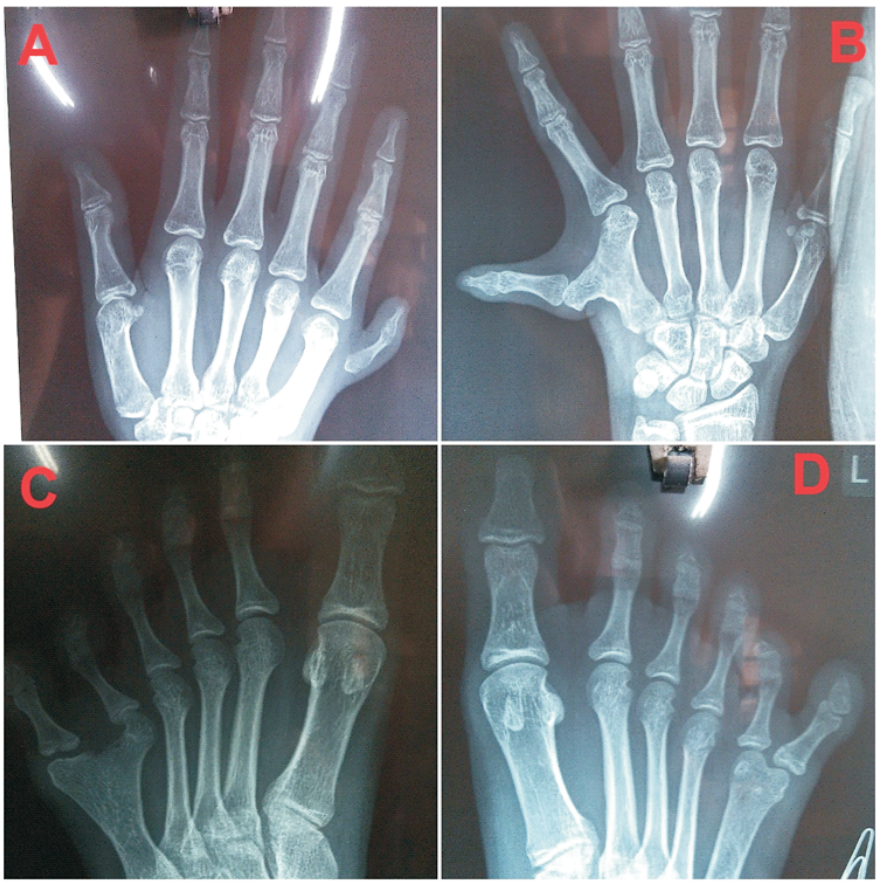

Fig. 2. : X ray of all four limbs A:right hand with isolated extra digit; B: left hand with T shaped 5th metacarpal; C: right foot with wide 5th metatarsal head; D: left foot with wide metatarsal head

uneventful and her mother had not had exposure to any infection or drugs while pregnant.

The patient was advised about surgical options and referred to the plastic surgery department.

DISCUSSION

Polydactyly is anatomically classified into 5 types [5]:

Type I: cutaneous nubbin; type II: pedunculated digit; type III: extra digit articulating with 5th metacarpal/tarsal; type IV: extra digit with extra metacarpal; type V: polysyndactyly. Types I and II are more common [5]. But in our case, the patient had a mixed type with one limb showing type II and the rest showing type III.

The incidence of polydactyly varies from 1 in 300 in blacks to 1 in 3000 in whites, according to various authors [1]. Tetrapolydactyly is very rare and its exact incidence has not been reported [1]. Of the various types of polydactyly, post-axial

Table 1. : Table showing the syndromes associated with polydactyly

Name of syndrome	Features besides polydactyly	Chromosome involved
Chondroectodermal dysplasia (Ellis-Van-Creveld syndrome) [7]	Hydroitic ectodermal dysplasia; Heart malformations	4p16
Asphyxiating thoracic dystrophy (Jeune Syndrome) [5]	Short ribs, short stature, bell-shaped chest	11q21-q22.1
Familial congenital muscular torticollis[5, 8]	Clubfoot, tibial torsion, shortening of sternocleidomastoid	Variable
Pallister-Hall syndrome	Hypothalamic hamartoma, bifid epiglottis	7p13
Pseudotrisomy 13 [9]	Holoprosencephaly	13q32; 5q35
Acrocallosal syndrome [4]	Absence of corpus callosum; dysplastic ears	12p; variable
Holt-Oram syndrome	Atrial septal defect; thumb abnormalities	12
Down's syndrome	Cardiac malformations; leukemia	21
Fanconi polycythemia	Anemia; leukemia; short stature	16
Meckel syndrome [10]	Renal cystic dysplasia; occipital encephalocele; pulmonary hypoplasia	17
Laurence Moon Biedl syndrome	Retinitis pigmentosa, hypogonadism	19p13
Patau syndrome	Intellectual disability holoprosencephaly	13
KlippelTrenaunay syndrome	Port wine stain; limb hypertrophy	3q26
Al-Awadi syndrome	Mesomelic dwarfism, limb bowing	Unknown
Mohr syndrome [11](as part of oro-facio-digital syndrome)	Polycystic kidney disease; cleft lip and palate	Xp22
VACTERL association	Vertebral anomaly; tracheo-esophageal fistula	Not defined
Fetal valproate syndrome [12]	Facial dysmorphism, spina bifida	None
Carpenter syndrome	Obesity, cranio-facial abnormality	6
McKusick Kauffman Syndrome	Hydrometrocolpos; heart disease	20

hexadactyly is the commonest variant, followed by pre-axial type-I hexadactyly [2]. There are other rare varieties like digit duplication and synpolydactyly [2]. But these rare variants are usually associated with other congenital syndromes.

When the extra digit is on the side of thumb or great toes, it is called pre-axial polydactyly and when the extra finger is towards little finger or little toe, it is called post-axial. Our case was post-axial in all four limbs. These cases are further divided into type A1 to A5, based on chromosomal mutations [6]. In our case, we could not study the genetic mutations due to cost constraints.

The Lmbr1 gene is important for vertebrate limb formation. Altered activity of this gene has been shown to increase or decrease the number of digits in experimental animals [5]. It is also hypothesized that loss of inhibitory pathways during limb development may also be an important factor in giving rise to supernumerary digits [5].

A lot of clinical syndromes may be associated with polydactyly. The table below gives a list of the known associations.

A study from Brazil found that polydactyly was associated with a syndrome in only 14.6% cases [13]. For the post-axial type, this association was even less (around 11%). But, the associated congenital syndromes, if present, may have fatal consequences. Thus, a case of polydactyly needs other investigations to rule out these associated syndromes. A variety of polydactyly, called crossed polydactyly is also reported, where the upper limb has

pre-axial type and lower limb the post axial type, or vice versa [14].

Tetrapolydactyly or polydactyly 24 is a very rare condition. Hence, any specific syndromic association with this particular anomaly has not been studied. Usually, when there is tetrapolydactyly, each of the four limbs has 6 digits. There is a single report of tetrapolydactyly from Romania, of a child with 6 digits in each of upper limbs and 7 toes in each foot [15].

The newest classification system for congenital hand anomalies is the IFSSH classification [16]. This is particularly important from the surgical point of view.

Polydactyly does not always need surgery. In some parts of the world, the extra digit is tied off at birth [1]. But this is a potentially dangerous practice. If needed, cosmetic surgery gives very good results. Such surgery is best done between 1 and 5 years of age [15]. However, the skeletal anatomy is important is deciding on the surgical strategy. A case like ours, where each limb has a different surgical anatomy, needs a different plan for each limb.

CONCLUSION

Tetrapolydactyly is a very rare congenital skeletal anomaly. It may be associated with clinical syndromes. In cases like the present one, where the skeletal anatomy varies in different limbs, surgical correction may be challenging.

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