



Case Report

Post-axial polydactyly in four limbs with different bony configurations

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ABSTRACT

Polydactyly is a common congenital digital variation of the hand and foot. It may appear as isolated or in association with other birth defects. Of the different types, post-axial variety is the commonest one. We here report a case of post-axial hexadactyly in all four limbs in a middle-aged female from Eastern India. She was completely asymptomatic and did not reveal any other birth defect. The fingers of only the feet were mobile. Radiography revealed different types of skeletal configurations in the four limbs, knowledge of which is essential for proper planning of the corrective surgeries. © IJAV. 2011; 4: 77–79.

Key words [polydactyly] [post-axial]

Introduction

Polydactyly is a common congenital digital variation of the hand and foot, which is characterized by supernumerary digits. This condition can occur in one limb or can be exceptionally present in all four limbs; a condition called tetrapolydactyly. It may appear as isolated or in association with other birth defects. Polydactyly might be pre-axial, post-axial and central type, the post-axial being the commonest one [1]. Previously, occurrence of polydactyly was explained as the consequence of the failure of programmed cell death during fetal limb development. Early theories for polydactyly concerned disorders were programmed cell death cycle of fetal limb development. But now, specific genetic malfunctions are thought responsible. Hexadactyly of all the four limbs is a very rare skeletal malformation. Wollina et al. described such a case of pure post-axial polydactyly involving all the four limbs but no radiological investigation was possible in that case [2]. Surgical intervention in polydactyly are indicated for various reasons. To have a successful surgical procedure detailed knowledge regarding the bony architecture of the region is necessary. We here present a case of polydactyly involving all the four limbs where bony architecture is grossly different in the different limbs, knowledge of which will help the surgeons to plan the different surgical procedures.

Case Report

A 50-year-old female was admitted in our hospital with fever. She was 163 cm long with normal built and nutrition. Her weight was 60 kg. She was born out of a non-consanguineous marriage. While examining her, it was found that she had six fingers in all the limbs. The extra fingers were post-axial type, on the ulnar and fibular sides in the upper and lower limb, respectively (Figure 1). The dermatoglyphic pattern of limbs was normal. The woman could move the extra fingers of the feet of her own, but not of the hand. All the fingers were well formed. On palpation, there were bony elements in each digit with mobile joints. The number and the pattern of the carpal and tarsal bones were normal. All the other joints of her body were normal. Radiographs of both feet and hands were taken with written permission from the patient explaining the reasons behind (Figures 2, 3). It revealed the following findings:

Right hand: The lateral side of the base of 5th metacarpal was having a facet from which the extra finger with two phalanges had started.

Left hand: The head of the 5th metacarpal was wide and had splitted into two parts and articulated with two phalanges. The lateral one was the proximal phalanx of the extra finger. The extra finger had three phalanges here.

Right foot: The 5th metatarsal head had an extra projection laterally. Top of that projection had articulated with the proximal phalanx of the sixth toe. It was having two phalanges.

Left foot: The 5th metatarsal near its middle had splitted into two. Tip or head of the two splitted parts had articulated with the proximal phalanges of the two lateral toes. The lateral most being the extra toe was also having two phalanges.

She neither had any abnormality of the nails and teeth nor any cleft palate or lip. Echocardiography and abdominal ultrasonography revealed no abnormality. Laboratory tests including hormonal studies and other biochemical tests were normal. There was no mental retardation and her features did not reveal stigmata of any congenital syndrome. None of her first-degree relatives had this sort of a variation.

So this case was finally diagnosed as a case of sporadic post-axial tetrapolydactyly in an adult with well formed phalanges with four different varieties in four limbs. Neither she had any functional limitation nor did she ask for any corrective surgery.

Discussion

Polydactyly is a condition of more than 5 fingers in a limb. It occurs in roughly 1 in 500 live births [3]. The condition is more common in males and related to consanguinity [4]. The mode of transmission can be autosomal dominant or recessive [5]. It is more common in colored people (mostly sporadic) than in Caucasians (usually associated with other congenital anomalies). Mutations in the *GLI3* gene and another locus on chromosome 13 were found to be associated with post-axial polydactyly but the majority



Figure 1. Figure shows the four limbs having an extra finger in the post-axial side in each.

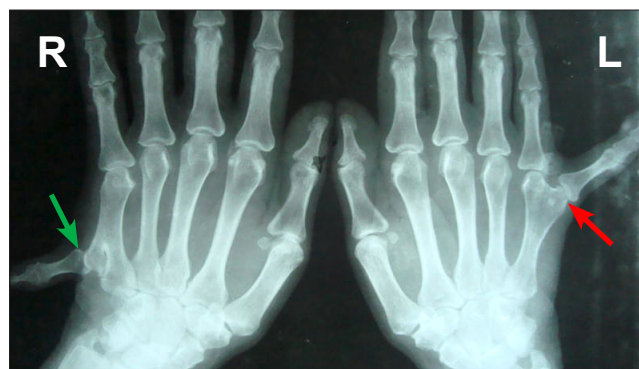


Figure 2. Radiograph of the hands. **Green arrow** shows an extra facet on the lateral side of the base of the right 5th metacarpal from which the 6th finger came out. **Red arrow** indicates wide head of the left 5th metacarpal giving rise to the 6th finger.



Figure 3. Radiograph of the feet. **Red arrow** indicates the wide right 5th metatarsal head and the **green arrow** on the left side shows Y shaped 5th metatarsal.

of cases remain unexplained. Recently another locus on chromosome 19p13.1-13.2 is found to be associated [6]. Different morphological classifications of polydactyly are available in literature. But that by Venn Watson has been widely accepted. He classified post-axial polydactyly into different types, e.g., soft tissue duplication, Y metatarsal, T metatarsal, wide metatarsal head and complete duplication [7]. In our case left hand and the right foot had wide metacarpal and metatarsal head, the left 5th metatarsal is of Y variety but right 5th metacarpal had the phalanges attached at the base (Figures 2, 3). Surgical correction of polydactyly is attempted for cosmetic reasons, to relieve pain if any and to have properly fitted shoes. A pre-surgical radiography, followed by amputation of the accessory digit and excision of the extra bony part is the standard procedure. The type of deformity, type of procedure, and skillfulness of the surgeon play a major role in the success of the surgery [8]. Our case shows the importance of detailed pre-operative study of the radiograph and proper pre-surgical planning since the bony pattern might vary widely even in the same person.

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