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Case Report

Isolated Tetrapolydactyly in a Nigerian Girl

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ABSTRACT

Tetrapolydactyly is a very rare form of polydactyly –common congenital anomaly of the hand or foot. This anomaly is rarest in females. Here, we present an 8 year old girl who presented to the plastic surgery unit of our institution with history of extra digits on the hands and feet since birth. There was no associated pain. Medical attention was not sought at birth, however she presented currently due to cosmetic reason. Tetrapolydactyly was evaluated with plain radiograph and ultrasound.

Keywords: Tetrapolydactyly, female, radiograph, ultrasound.

INTRODUCTION

Polydactyly is defined as a congenital anomaly of the hand or foot with the presence of supernumerary digits. [1] The extra digit is usually a small piece of soft tissue. Occasionally it may contain bones without joints but it may also be a complete functioning digit. [2] Polydactyly can occur as an isolated disorder, in association with other malformations of the hand and feet or as part of a syndrome. It can occur sporadically but it can also be inherited with a mainly autosomal dominant inheritance. [3] It is said to be commoner in males but occurrence in females has been rarely reported. [2]

Polydactyly is a common and very conspicuous congenital anomaly of the hand and feet. It mostly involves only the hand or foot. Polydactyly involving both the hands and feet (tetrapolydactyly) is very rare. [3] We report a rare case of polydactyly involving both hands and feet in a female with review of the relevant literature.

CASE REPORT

We present an 8 year old girl who presented to the plastic surgery unit of Usmanu Danfodiyo University Teaching Hospital Sokoto with history of extra digits on the hands and feet since birth. There was no associated pain. Medical attention was not sought at birth, however she presented currently due to cosmetic reason. The antenatal period was uneventful. She is the fifth child in a polygamous setting. No history of polydactyly in the family. No history of other congenital anomaly.

Examination showed a healthy looking girl, not pale, anicteric. The remarkable finding was on the hands and feet where there was an extra digit on the ulnar side of both hands and on the lateral side of both feet. The extra digits showed motor and sensory functions. Other systemic examinations were essentially normal. A clinical assessment of polydactyly of the hands and feet was made and she was

referred to Radiology department for radiography of the hands and foot.

The radiographs revealed a fully developed sixth digit with metacarpal and phalanges on the ulnar side of the right hand (Figure 1). On the left hand the sixth digit showed only phalanges that were attached to the fifth metacarpal (Figure 1). There were sixth toes on both feet which showed phalanges arising from the lateral branch of bifid fifth metatarsals (Figure 2). A complimentary ultrasound of the abdomen was done to assess for other associated congenital anomaly but showed normal findings. A diagnosis of post axial tetrapolydactyly was made.

She had surgical correction (disarticulation of all the extra digits). The post-operative period was uneventful.

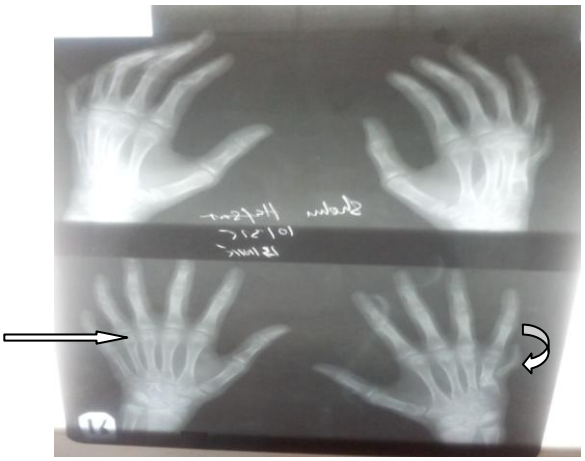
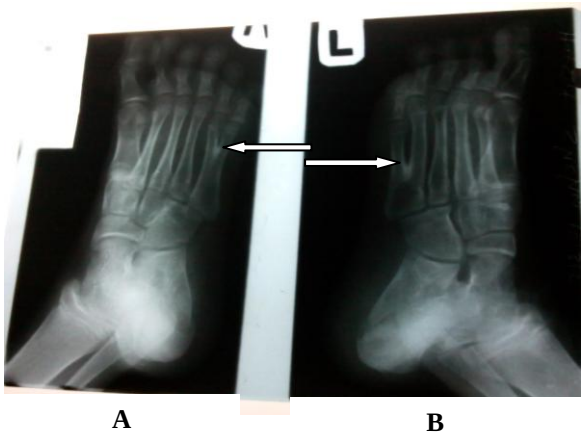


FIGURE 1: Radiograph of both hands oblique (above) and antero-posterior (below) views showing fully developed sixth digit on the ulnar side of the right hand (straight arrow) and an extra digit attached to the fifth metacarpal on the left hand (curved arrow).



A **B**

FIGURE 2: Radiographs of both foot oblique views (A and B) of the same patient showing extra toe on the lateral side of the foot bilaterally. Note the bifid fifth metatarsals as well (arrows).

DISCUSSION

Polydactyly is one of the most common congenital anomalies of the hands or feet consisting of supernumerary fingers and toes. Various incidences have been reported but on average, incidence in blacks is 1 in 3000. [2,4] This condition can occur in one limb or can be exceptionally present in the four limbs a condition called tetrapolydactyly as in the case here presented. The incidence of tetrapolydactyly has not been reported as the condition is reported to be rare. [2,3]

The extra digit in polydactyly is most common on the ulnar side of the hand (post axial ray), less common on the radial side (pre axial ray), and very rare within the middle three digits (middle or central ray). [2] This case had extra digits on the ulnar sides (post axial ray) bilaterally.

Five types of polydactyly are usually encountered in the hands: Type I- cutaneous nubbin, Type II- pedunculated digit, Type III- articulating digit with fifth metacarpal, Type IV- fully developed digit with sixth metacarpal and Type V- polysyndactyly. Type I and II ulnar polydactyly are more prevalent. [5] Our case showed a mixture of type III and IV polydactyly.

Polydactyly has been said to be commoner in males and most of the polydactyly reported in the literature confirms this. [2,6,7] Post axial ray poydactyly is the common presentation in blacks, usually in males as an isolated disorder. The occurrence of either polydactyly or tetrapolydactyly in females has been rarely reported. [2] Polydactyly can occur as an isolated disorder, in association with other malformations of the hand and feet, or as part of a syndrome. Some of the syndromes include Holt-Oram syndrome (heart-hand syndrome), Down's syndrome, Fanconi polycythemia and Klippel Trenaunal syndrome. There may be associated cleft palate, hearing difficulties, renal anomalies, limb and vertebral anomalies. [2,6] Ibrahim [6] reported a case of heart – hand syndrome in a 3 year old boy in Kano. [6] There was no other associated

congenital anomaly in the case here presented. In polydactyly, the extra digit may be functional as demonstrated in this case.

The imaging methods used in evaluating polydactyly are plain radiography, magnetic resonance imaging (MRI) and ultrasonography. The aim of plain radiograph of the hand and feet is to identify if the extra digit has a bony component and also to define the extent of attachment to the normal bone. MRI shows the soft tissue component of the extra digit including muscular and tendinous attachments. Ultrasonography is used to evaluate associated anomalies such as renal or cardiovascular anomalies. Pre natal ultrasound can also diagnose polydactyly in utero. [4]

The clinical management of polydactyly is usually surgical correction. This is almost always indicated not only for cosmetic improvement but also for better function of the hand or foot. Surgical reconstruction generally is performed between 18 months to 5 years. [3] Also majority of cases of polydactyly without bony component usually have the extra digits tied off at birth. [2] This case presented late at 8 years and she had surgical correction of the tetrapolydactyly.

CONCLUSION

Polydactyly is a common congenital anomaly of the hand or foot, but tetrapolydactyly affecting all the hands and feet are very rare especially in females. A rare case of tetrapolydactyly in a girl evaluated with plain radiograph and ultrasound is presented.

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