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Original Research Article

SPORADIC FORM OF WASSEL TYPE VII PREAXIAL POLYDACTYLY

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Abstract: Polydactyl is a congenital physical anomaly of the fingers and toes. Preaxial polydactyly is rare in this environment and is associated with several syndromes. This is a report of a patient with a rare sporadic form of preaxial polydactyly.

Keywords: Sporadic, preaxial polydactyly, Wassel Type VII.

Introduction: Polydactyl is a congenital physical anomaly consisting of supernumerary fingers or toes.[1] Polydactyly of the hands or feet is a common birth deformity that occurs in many forms, ranging from varying degrees of mere splitting to completely duplicated thumb.[2]Wassel type VII or Triphalangeal thumb is regarded as a subtype of preaxial polydactyly with reported prevalence as 1:25,000.[3]The extra digit is usually a small pieceof soft tissue, but occasionally, it may contain bone without joints, and rarely it may be a complete functioning digit.[1] The extra digit is most common on the ulnar (little finger) side of the hand, less common on the radial(thumb) side, and very rarely within the middle three digits.[1] Polydactyl is classified into three main categories: 1) preaxial polydactyl duplication of the thumb 2) central polydactyl which is duplication of the index, long or ring finger, and 3) postaxial polydactyl which is duplication of the small finger.[1,4]The preaxial polydactyl is the most common duplication pattern in white and Asian populations, occurring in one per 3000 births.[4]Post axial polydactyl is more common In blacks and African Americans.[1,5]In general, polydactyl commonly affects more males than females and involvement of the hand is usually unilateral.[6]This case is been reported due to rarity of this abnormality in our environment.

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Case Report: I.I is a 5-year-old girl who presented at the surgical outpatient department of Jos University Hospital with an extra digit in the distal aspect of the left thumb which was noticed since birth and has been progressively increasing in size. This was not associated with pain. No swelling or deformity in any part of

the body. Pregnancy was uneventful. The mother admitted to taking only the medication prescribed during the antenatal visits at the hospital. No other physical anomalies were seen. She is the 2nd of two children in a monogamous set up. There was no history of similar occurrence in the other sibling.

Musculoskeletal examination revealed an extra digit on the lateral surface of the left thumb. The extra digit was well formed but less developed compared to the other normal digits. It had well developed bones, soft tissue with well-formed nails. The cardiovascular, gastrointestinal and urogenital systems were essentially normal clinically.

A clinical diagnosis of polydactyl of the left thumb was made

Plain radiography of the hand (anterior posterior view) revealed duplication of the left thumb consisting of hypoplastic distal and proximal phalanges seen lateral to the thumb (figure 1). There was an apparent articulation with the 1st metacarpal bone. A radiological diagnosis of preaxial polydactyl type VII was made.

Patient was worked up for surgery. At surgery, duplication of the left thumb with attachment to the parent thumb through a joint was seen. The extradigit (thumb) was amputated and patient is doing well.

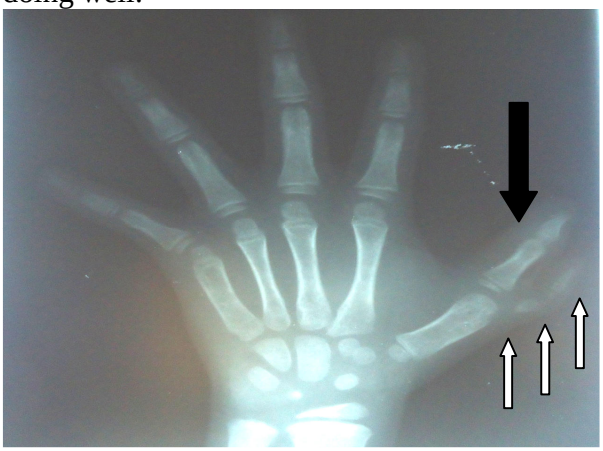

Figure1: Plain radiography of the right hand showing duplication of the right thumb. Note the well-formed phalangeal bones (black arrow) and the hypoplastic phalanges (white arrow).

Discussion: Polydactyly is the most common congenital hand malformation.[2] Thumb polydactyly is believed to arise from excessive cell proliferation and disturbed cell necrosis of pre-axial ectodermal and mesodermal tissues before the eighth week of embryonic life.[7] Polydactyly of the thumb occurs in many forms, ranging from varying degrees of splitting to completely duplicated thumb. Occasionally it consists of fleshy nubbins on the radial border.[8]Preaxial polydactyly commonly affects males more than females and involvement of the hand is usually unilateral and commonly seen in whites compared to blacks.[4,6]The index patient is a black female and involvement of the hand was unilateral. Wassel has classified polydactyly of the thumb into Types 1-VII,[8] and they are as follows.[8]

Type I: Bifid distal phalanx
Type II: Duplicated distal phalanx
Type III: Bifid proximal phalanx
Type IV: Duplicated proximal phalanx
Type V: Bifid metacarpal
Type VI: Duplicated metacarpal
Type VII: Triphalangia

Type IV is the most common with duplication of the proximal phalanx which rest on a broad metacarpal while Type II is the second most common.[5,8]The index case presented with Type VII preaxial poydactyly (Triphalangeal thumb), a rare form of polydactyl associated with several syndromes.[5]Triphalangeal thumb is a rare malformation of uncertain pathogenesis, it represents persistence of a middle phalanx owing to failure of fusion of the middle and distal phalanges of the normal first digit.[9]Triphalangeal thumb may occur as an isolated defect, or as a feature of a syndrome of congenital anomalies.[1,9]Isolated thumb polydactyly is often autosomal dominant, sporadic in occurrence and rarely autosomal recessive while syndromic polydactyly is commonly autosomal recessive.[9]These syndromes include Holt-Oram syndrome, Blackfan-Diamond anemia, Townes-Brockssyndrome, Greig cephalopolysyndactyly,

imperforate anus, cleft palate, tibial defects.[1,5]There may also be associated radial hypoplasia, bone marrow dysfunction, congenital heart disease, lung hypoplasia or agenesis, sensor neural hearing loss, onychodystrophy, mental retardation.[9]None of these syndromes were seen in this index patient. However, genetic analysis of multiple Dutch pedigrees with triphalangeal thumbs has localized this genetically transmitted malformation to the long arm of chromosome 7.[6]

The isolated triphalangeal thumb usually presents as a long fingerlike thumb with deviation in the same plane as the fingers.[6] Most of the triphalangeal polydactyly have their level of duplication at or proximal to the metarcarpophalangeal joint.[6] In this index patient it was also located proximal to the metarcarpophanlangeal joint. In triphlangeal thumbs, apart from the morphological forms, the involved ligaments, muscles, tendons and joints of the duplicated thumb, distal interphalangeal joint and the radiocarpal joint may be hypoplastic, malformed, or absent with varying degrees of stiffness or instability.[6]The radiocarpal joint of this patient was normal andno other deformities were noted in the other hand.

Radiographs of the affected limb are very important to show whether the rudimentary digit contains skeletal elements. The degree of deviation of the digit and the size of the articulating metacarpal or metatarsal also may be helpful in surgical planning.[10] Surgery is the main stay of treatment. However, there is no standard surgical procedure for correcting polydactyly of triphalangeal thumb.[6]Surgery is recommended in the first year or life, generally between 9 and 15 months of age before the development of pinch and fine motor function.[1] However the index patient had her surgery when she was 5 years old as this was when she presented. The aim of the treatment is to achieve a single, functioning thumb with acceptable appearance. Usually the thumb that is retained is the one with anatomic features that will give better stability, strength and mobility,[11] This is usually, but not invariably, the ulnar thumb. The patient had surgery and she is doing well

Conclusion: Sporadic occurrences of radial polydactyl as in this case do occur. Radiological evaluation is crucial to pre-operative evaluation for appropriate surgical planning.

Conflict of Interest: No conflict of interest

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