Bilateral proximal delta phalanges: an unusual presentation of familial congenital clinodactyly

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Abstract We report a unique case of bilateral proximal delta phalanges with secondary distal accessory ossification centers of the long fingers in a 16-month-old boy. The bony anomalies presented as severe ulnar deviation at the proximal interphalangeal joints bilaterally. This is a unique occurrence of congenital clinodactyly which has not been previously reported. Several family members show similar deformity, although to lesser degrees, suggesting an autosomal dominant mode of inheritance with incomplete penetrance and variable expressivity.

Introduction

Clinodactyly or “bent finger,” refers to angulation deformities affecting a digit in the radioulnar plane. The underlying etiology in congenital cases is commonly a “delta phalanx”, [6] a misshapen triangular (or trapezoidal) bone, which is often caused by a longitudinal bracket epiphysis on the short side of the phalanx. When inherited, they are usually passed down in an autosomal dominant fashion with variable expressivity. This classically involves the middle phalanx of the small fingers bilaterally with radial deviation at the distal interphalangeal (IP) joint. [3] It is important to note, however, that not all presentations are uniform. Here, we present the first case

of familial, congenital clinodactyly affecting the bilateral long finger proximal phalanges of a 16-month-old male, caused by longitudinal bracketed epiphyses with a second distal accessory ossification center.

Case Report

A 16-month-old boy presented to the Hand Clinic at Texas Children’s Hospital for evaluation of bilateral long finger deformities (Fig. 1a). Per the mother, the patient had the deformity at birth, without any significant change since. They had not seen a hand specialist prior to this visit because the patient did not have any functional deficiencies. The patient had previously been an uncomplicated term birth, and up to this point, had met normal developmental and growth parameters. The patient was otherwise healthy, except for a diagnosis of a lingual frenulum.

Physical examination revealed marked ulnar deviation of the bilateral long fingers, originating at the proximal interphalangeal joint, as well as mild ulnar deviation of the bilateral second digits. The patients’ IP joints, however, were stable on exam with full flexion and extension. Additionally, there were no apparent or gross functional deficits in his ability to make a fist, pinch, or grasp objects; however, there was significant overriding or scissoring of the fourth and fifth digits (Fig. 2). Radiographs subsequently demonstrated a complex deformity involving the proximal phalanges of the bilateral long fingers (Fig. 3). The underlying bony anomalies had the appearance of a delta phalanx with a second distal accessory ossification center. Angulation measured 48° on the left and 37° on the right. Mild ulnar deviation to the bilateral second digits was also noted, although no underlying radiographic anomaly was seen.

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Fig. 1 Familial clinodactyly of the long fingers as seen in the 16-month-old male proband (a), brother (b), and maternal grandmother (c)

The patient also had a noted family history for similar hand abnormalities. While the patient's mother was phenotypically normal, she noted that several members in the family also had congenital deformities of the long fingers. Specifically, the patient's brother, uncle, and maternal grandmother all had varying degrees of clinodactyly (Figs. 1b, c and 4). In these cases, deviation was consistently in the ulnar direction. Paternal family history

was unavailable. Multiple attempts were made to have the family return to the Texas Children's Hospital for genetic testing; however, they failed to show up for these scheduled office visits.

Given the lack of functional constraints and the patient's age, the decision was made to observe the patient until 4 or 5 years of age to allow further growth prior to corrective procedures. The long-term plan will be to perform wedge osteotomies of the bilateral delta-type phalanges at that time. If functional limitations or significant finger length discrepancy develop, then earlier surgical intervention may also be indicated.

Fig. 2 Despite obvious scissoring of the digits, the patient did not have significant functional deficits as evidenced here by good use of his left hand

Fig. 3 Anteroposterior view of the patient's left (a) and right (b) hands demonstrate severe ulnar deviation of the bilateral long fingers. The underlying bony abnormality appears to be a longitudinal bracketed epiphysis and secondary ossification center of the long finger proximal phalanges bilaterally

Fig. 4 Family tree demonstrating apparent autosomal dominant with variable expressivity and incomplete penetrance mode of inheritance. Of note, paternal family history was not known

Discussion

This report presents the unique case of bilateral, familial, long finger clinodactyly caused by proximal delta phalanges with a second distal accessory ossification center. Angulation of the digits in the radioulnar plane is highly prevalent in the general population and up to 10° of deviation is considered normal. [3] The majority of these cases are rarely functionally limiting, and most cases can be treated conservatively; however, with more significant deformities, functional, and esthetic consequences may necessitate a surgical intervention. Angulation of $20\text{--}25^\circ$ or greater is generally used as an indication to surgically intervene to prevent functional problems [1, 4, 8].

Clinically significant clinodactyly can occur as a congenital (familial, syndromic, or sporadic) or traumatic deformity. The majority of familial cases of clinodactyly are inherited as an autosomal dominant trait with variable expressivity. This case, however, likely represents a separate clinical entity from the traditional familial form, as the pattern of involved family members (Figs. 1 and 4) and range of severity imply that this is an autosomal dominant trait with incomplete penetrance and variable expressivity. Interestingly, bilateral proximal delta phalanges of the great toes have also been reported in a family with only isolated hallux varus congenitus deformity [7].

Cases of clinodactyly can also be classified based on whether the underlying cause is either a soft-tissue or bony abnormality. Soft tissue restraints include abnormal fibrous bands as reported by Wood, as well as syndactyly. [9] Bony clinodactyly, on the other hand is usually caused by a delta phalanx in which an abnormal physal plate causes restricted longitudinal growth on the short side of the phalanx. The deformity classically involves the middle phalanx of the small fingers bilaterally with radial deviation. The most commonly involved digit in clinodactyly is the small finger, followed by the thumb and then the ring finger, whereas index and long finger involvement are relatively rare. [2–4] The hereditary component, involve-

ment of the long, and index finger bilaterally, as well as the presence of a secondary ossification center, suggest underlying genetic abnormalities. Given that this involves the central two digits, it is also possible that the underlying gene responsible may be similar to the gene involved with central synpolydactyly. [5] Without genetic testing, however, as was not possible in this situation, this cannot be confirmed.

The majority of cases of delta phalanges usually involve the middle phalanx, whereas documented cases of proximal delta phalanx are rare. These tend to present earlier with more significant angulation due to the proximal location of the lesion and are more likely to cause functional disability. In this case, the patient presented at 16 months of age with a $\sim 35\text{--}50^\circ$ angulation deformity bilaterally. Additionally, the index and long fingers were somewhat shortened, rotated, and had abnormal flexion and extension at the interphalangeal joints. Impressively, the patient did not present with any gross functional disabilities, which highlights the remarkable ability of children to adapt to congenital hand deformities. The significant scissoring demonstrated upon grip formation; however, would undoubtedly lead to functional issues in the future. For these reasons, it was deemed appropriate to observe this patient for interval growth to determine the optimal timing of future correction. The patients' deformity also radiographically demonstrated the presence of a third ossicle. While this could potentially be a manifestation of a separate ossicle articulating with the proximal phalanx or a separate ossicle forming a coalition with the bracketed epiphysis, its close proximity to the proximal phalanx makes it more likely to be a secondary ossification center.

Several methods have been described to treat these deformities. These generally involve performing a physiolsis or osteotomy (opening, closing, or reverse wedge) with soft-tissue lengthening as needed. Physiolsis may facilitate remaining growth potential with a reported overall correction of 11° , [4] but as expected, it yields better results when performed before 6 years of age, with more severe preoperative deformity ($>40^\circ$ angulation), and when managing trapezoidal phalanges. Attempting repeat physiolsis is not recommended as an initial procedure given the minimal benefit and risk of premature physal fusion. However, delayed wedge osteotomy can safely follow physiolsis to gain further correction. The patient in this case will require and likely receive an opening or closing-wedge osteotomy of the proximal phalanx with soft-tissue lengthening procedure for the shorter ulnar side to manage his deformity. If concern exists regarding the length of the digit at the time of surgery, the digit could also be lengthened at the time of osteotomy or distracted if the discrepancy proved severe. The patient may

also require a surgical procedure to correct the underlying ulnar deviation of the index fingers, and would be managed using similar surgical techniques.

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