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Case Report

Familial congenital ulnar drift/windblown hands: *Case report in 4 successive generations*

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Key words: Congenital ulnar drift, windblown hands, ulnar drift

Introduction

Congenital ulnar drift (CUD) of fingers is a rare congenital anomaly consisting of multiple hand deformities that progressively worsens affecting the normal function of hands and causing cosmetically unsatisfactory appearance. This was first described in 1897 by Boix[i] (Boix, 1897).

Although mostly acquired (Rheumatoid arthritis/Jaccourds arthropathy), possible familial type CUD of autosomal dominant inheritance is infrequently reported in literature (Chowdary, 2009).

Here we report on five individuals of four successive generations of a family, with CUD, which to our knowledge is the first such report in the country.

Case presentation

15 days old, term infant presented to the neonatal clinic with B/L symmetrical ulnar drift (UD) of fingers at MCP joints and developing flexion contractures which were tender on palpation. On clinical examination there was no other evidence of skeletal or cardiovascular abnormalities.

X-ray showed B/L ulnar deviation of wrist joints and B/L MCP joints without fractures or subluxation of joints. She was referred for early physiotherapy.

The rest of the affected family members have UD with flexion contractures but function with minimum disturbance to activities of daily living.

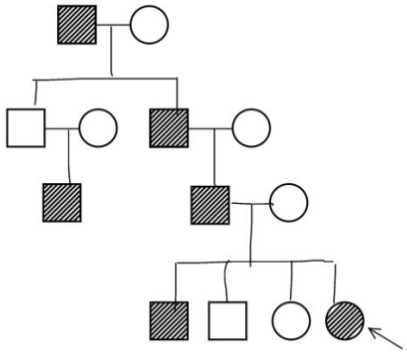

Figure 1: Family's pedigree

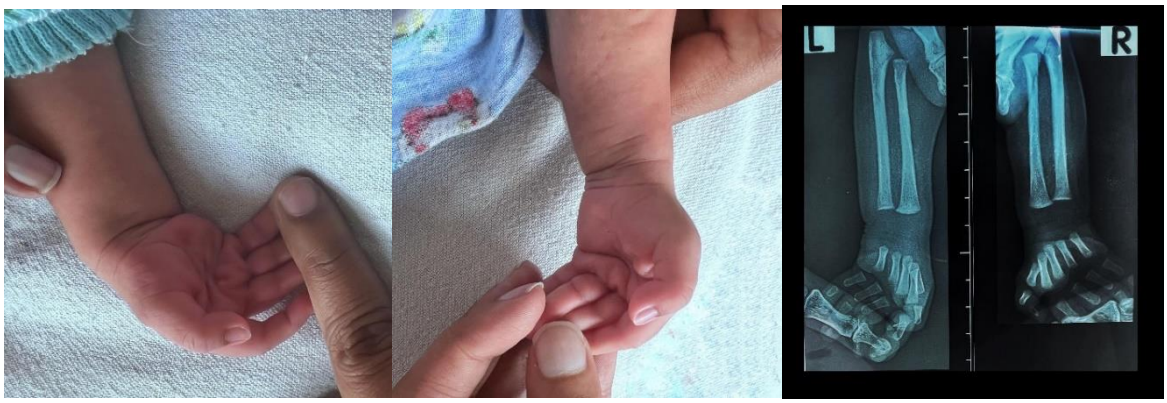

Figure 2: Appearance of the hand of the baby and Radiographic findings

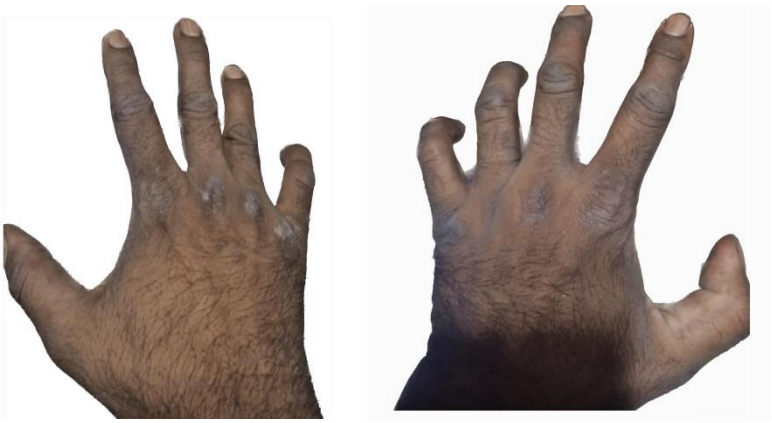

Figure 3: Appearance of the hands of baby's father

Discussion

CUD unlike UD is a rare condition associated with various syndromes; Arthrogryposis, Marfan and other hyperlaxity syndromes, freeman-Sheldon syndrome and Escobar syndrome. Associated craniofacial, foot and other anomalies will be observed in these unlike in our case.

CUD comprises of 3 components; ulnar deviation of digits, MCP joints and PIP joint flexion contractures with clasped thumb deformity.

Insufficiency of aponeurosis (Boix, 1897), hypoplasia / absence of extensor tendons (J.R. Fisk, Jan 1 1974) and malformation of the retinaculum cutis - mid palmar fascia/ natatory ligament (E Zancolli, 1985 Aug) are some pathophysiology suggested for CUD. Severity of CUD vary from mild fasciocutaneous to bony deformities (E Zancolli, 1985 Aug).

Management of CUD could be done non-surgically with splinting or surgically, out of which the second is preferred and benefited if attended before 2-3 years of age. (Virchel E. Wood, 1990) (Kalliainen, 2003)

Even though surgical correction creates near normal anatomical appearance and function, it does not exclude the chances of recurrence, especially when done very early in life. (Chowdary, 2009)

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