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Familial Ulnar Club Hand

Case Report in 3 Successive Generations

Abstract: We report two 11 and 14 year old girls with bilateral ulnar club hand deformity, who have the familial form of the disease. Their mother and 8 other family members are affected by the same deformity. The congenital form of deformity was first reported by Robert in 1886 and since then, one more case has been reported in the literature. Our report to our knowledge is the first that reports the case in 11 members of 3 generations of a family.

Keywords: club hand, ulnar, familial

Introduction

Ulnar club hand or 'ulnar longitudinal deficiency' is a developmental failure of the ulnar side of the forearm (post axial).

This is one of the rarest anomalies of the upper extremity. According to Brich – Jensen, the incidence is 1 in 100,000 live births. Male to female ratio is 3 to 2; one in four cases is bilateral, and the left-side involvement is slightly more common.[1, 2]

Most cases occur sporadically, although the longitudinal ulnar deficiency may occur as a part of a recognized syndrome.[3, 5]

Robert was the first to report this disease in three members of the same generations: two sisters and one brother. Up to now, no definite teratogenic factor has been found as an etiologic factor.[2, 4]

This anomaly has a wide spectrum of upper extremity defects, ranging from mild hypogenesis of ulnar digits to complete absence of the ulnar side of the forearm and hand. However, radial digit absence or hyperplasia, thumb duplication, and syndactyley are present in 50%.

Normal digits are seen in 11%, absence of one digit in 12% and complete absence in 3%.Carpal abnormality ranges from the absence of one bone to carpal coalition, which is seen in 20% of cases.

Case presentation

Two sisters from Northern Iran (Rasht) with ulnar club hand deformity were referred to Akhtar hospital in Tehran, in winter 2001.

Case 1

A 14-year-old girl presented with bilateral ulnar club hand. and shortened forearms, but no other apparent skeletal anomalies. (Figures 1 and 2)

On clinical examination, there was no evidence of cardiovascular disorders. Her laboratory exams were normal. On radiography (Figure 3) she had a short ulnar bone, and the radiocarpal joint was defective, the radius was bowed and the radiocarpal joint was triangular. She had a dislocation of radial head in the elbow. The second row of carpal bones was normal. According to Swanson classification, she had type I deformity. She was treated surgically with dorsal and volar approaches: A three-level osteotomy of radius along with Z osteotomy in distal ulna was done. The ulna was fixed with a screw, then the flexor carpi ulnaris tendon was lengthened and fixed with an intramedulary pin.

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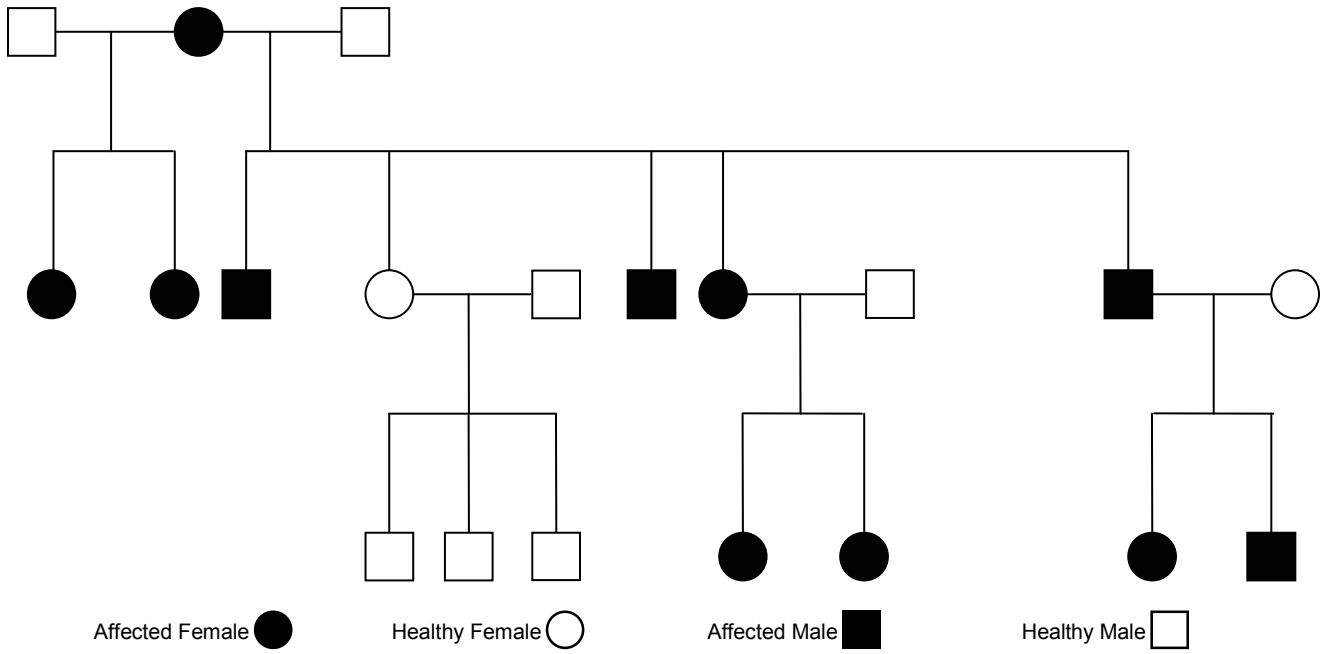
Case 2

An 11-year-old girl (sister of case 1) presented with bilateral club hand deformity, and short upper extremity. Clinical and laboratory findings were normal. She had type I, ulnar club hand deformity. On caryotype, no chromosomal defect could be shown in neither of the cases. The family's pedigree is drawn below.



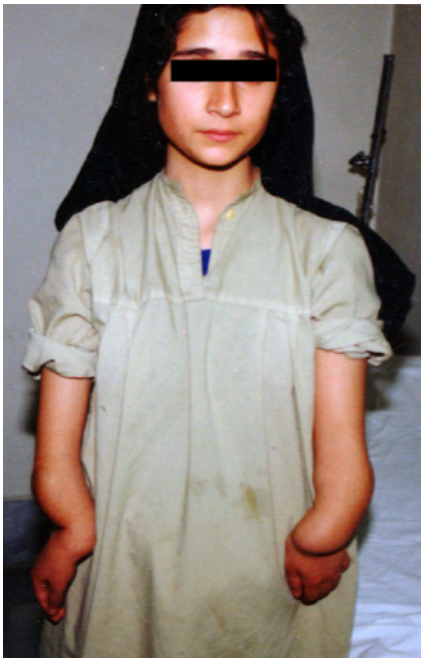
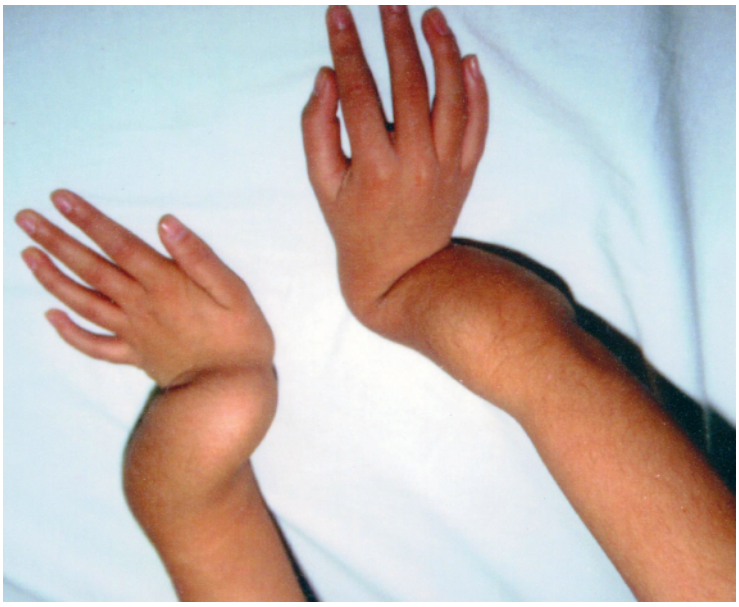


Figure 1: General appearance of case 1.

Figure 2: General appearance of case 1's mother and sister (case 2)



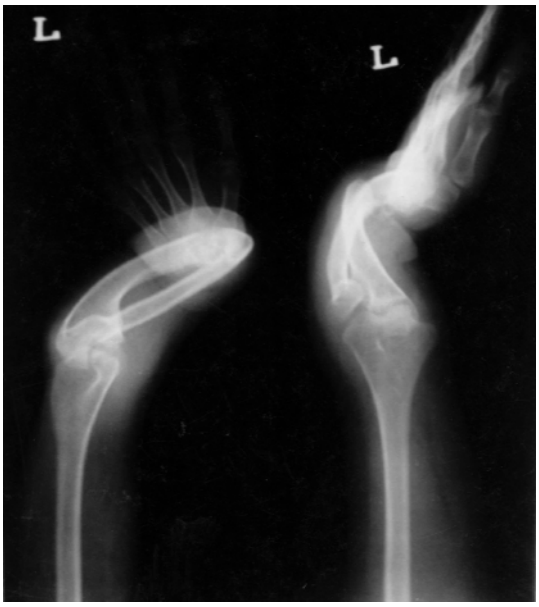


Figure 3: Forearm appearance and Radiographic findings in case 1, AP & AT.

Discussion

Ulnar club hand is the second rarest upper extremity anomaly. No teratogenic factor has been found responsible, yet. Some authors have shown a limb defect gene as a factor.[7, 8]

All reported cases have been sporadic ones, except a report by Roberts in 1886, which presented a familial type in 3 patients of one generation.

In our report, the mother of the patients had the same anomaly. Three of their brothers (one of them was dead) had the same deformity, with one of their sisters and 3 nieces (5, 7 and 10-year-old) and a 9 – year-old nephew.

The familial form of ulnar club hand, passing from one generation to another has not been reported previously.

In this family, 11 members suffer from the same anomaly, all of who were examined clinically and proved to be otherwise normal.

According to Swanson's classification 4 types are recognized:

- Hypoplasia or partial ulnar deficiency
- Total ulnar defect
- Total or partial ulnar defect with humeroradial synostosis
- Total or partial ulnar defect associated with congenital amputation at wrist.

The radiological differential diagnosis includes:

- Short ulna
- Dyschondrosis
- Madelung deformity
- Nievergelt syndrome.

Treatment of this syndrome is both surgical and non-surgical. In infants, serial casting could be used and should be considered until correction is achieved, and then removable splints may be used to maintain the correction. Indication for surgical intervention are syndactyly, radial bowing, presence of ulnar anlage ,dislocation of the radial head with limited elbow extension, forearm pronation and supination as well as internal rotation deformity of humerus.

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