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Case Report
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ULNAR DIMELIA, A CASE REPORT

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Ulnar Dimelia which is also known as mirror hand, is a very rare congenital disorder of the upper limb and represent extreme forms of polydactyly. It is considered a duplication of ulna and fingers.

Case Report

A seven years old healthy boy presented with left unilateral mirror hand deformity with no family history of any congenital disorders.

On physical examination there were 7 fingers with no thumb which was replaced by 3 separated fingers radially (fig.1), and were well formed but somewhat smaller with slight flexion at the proximal interphalangeal joints with obvious absence of the thenar muscles (fig.2), the most ulnar digit was the most functioning with acceptable extension and flexion.

Figure 1.

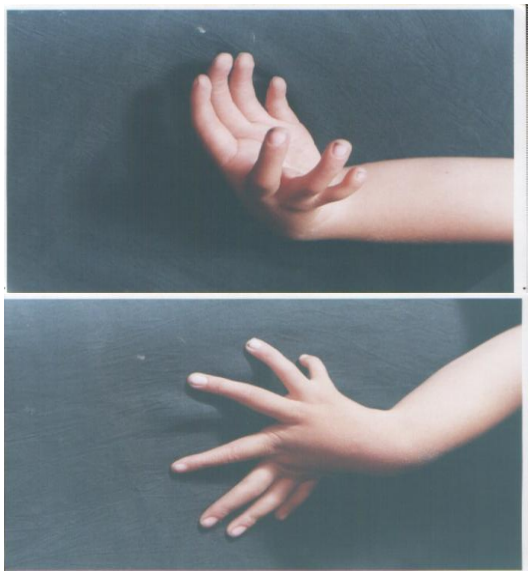
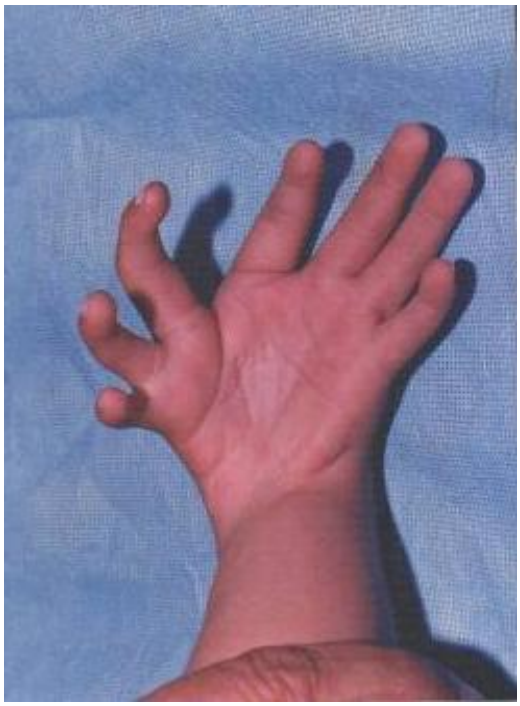


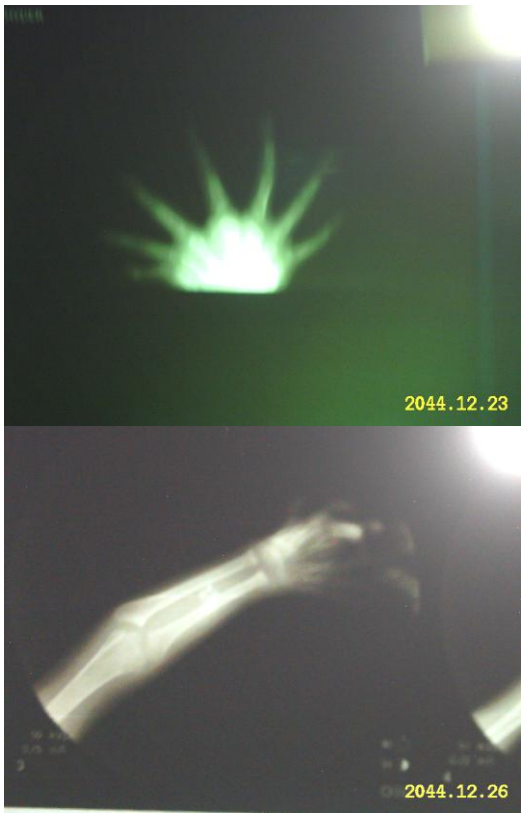
Figure 2.



The elbow joint was in extension posture with active flexion of only 30 degrees, the forearm rotation was limited, the wrist was in 20 degrees flexion due to tight tendons.

On radiographic evaluation the ulna was duplicated with absent radius and the lower humerus articulating with 2 olecranon processes. There was no proximal carpal row at the wrist and in the hand there were 3 well formed extra-metacarpal (MCP) bones radially

articulating with separated triphalengeal digits (fig. 3).



The operation was conducted under general anesthesia with application of tourniquet. The first step was to obtain maximum flexion of the elbow. A longitudinal incision was done radially over the pre-axial ulnar bone. We excise about 1 inch from the proximal extra ulnar bone followed by reconstruction of the collateral ligament and soft tissues to the proximal part of the post-axial ulna. The next step was excision of the most 2 radial fingers with preservation of the skin flap from the middle with releasing of the first web. Shortening osteotomy at the level of the metacarpal neck of the remained digit was done and the metacarpal head rotated for about 80 degree of apposition and fixed by 2 k wires. A cortical bone graft from the amputated MCP bone was pressed between the 2nd MCP bone and that of the pollicised digit to fix it in abduction, and the skin flap was transposed to first web. The last step was correction of the wrist flexion; tenotomy of the tight flexor carpi ulnaries and palmaris longus tendons was performed with capsulotomy. A long arm posterior splint was applied with elbow in flexion. Wrist was extended and the new thumb was abducted. Post operatively the K-wires and the splint were removed after 4 weeks and another splint to the thumb was applied to keep it in abduction and opposition for additional 4 weeks.

Discussion

Mirror hand is a very rare complicated upper limb congenital anomaly, a few cases were reported. The largest series describe this anomaly include only 3 patients, the cause and management remain a controversy[1-3].

In this patient, there were 3 radial separated extra digits, all were triphalengeal (9 phalanges) articulating with 3 separated MCP bones with absence of thenar muscles. Really this clinical condition differs from another rare congenital condition called triplicated thumb. The most ulnar extra digit looks well developed and have the best function was preserved and pollicised and the reconstructive principles was the same with other forms of the extra digits. Our aim from surgery was to restore elbow flexion, forearm rotation and appropriate thumb from the functional point[4,5].

It is concluded that although it is very uncommon complicated condition and we are not familiar to deal with such entity, after operation we end with elbow flexion up to 85 degrees and a new thumb with acceptable function, in this young boy just like true triplication of the thumb, all of the principles of thumb reconstruction must be used.

References

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