Case Report

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Successful surgical reconstruction of atypical variant mirror hand anomaly in a 2-year-old female child: a unique case report

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Introduction: Mirror hand is an extremely rare congenital abnormality characterized by polydactyly and duplication of the ulna, with the absence of the radius and thumb. Atypical presentations of mirror hand were described, including the presence of the radius in a few cases; here the authors report one of the atypical cases of mirror hand that underwent successful management.

Case presentation: A 2-year-old and 7-month-old female child presented with 7 well-developed digits, with an absent thumb; the X-ray imaging of the forearm showed a well-formed ulna and radius with proximal fusion. The patient has good shoulder movement, minor limitations in supination and pronation, and elbow flexion restriction. The patient underwent multiple surgical interventions for pollicization. Follow-up revealed a significant improvement of hand function and appearance.

Clinical discussion: In the literature review, mirror hand is a rare congenital malformation and has many varieties. The management of this deformity is a challenge and differs from case to case; here the authors described a novel variant of this deformity and its successful management.

Conclusion: Mirror hand is a rare congenital abnormality and has a wide spectrum of variants. The management challenge, but with early pollicization with appropriate functional considerations, the outcome is promising.

Keywords: case report, congenital upper limb abnormality, mirror hand anomaly, pollicization, postoperative management

Introduction

Mirror hand is an extremely rare congenital abnormality of the upper extremity characterized by the duplication of the ulna ray, absence of the radius, and polydactyly with symmetry about the midline, generally with seven fingers[1,2]. Mirror hand deformity is classified into two main types according to the bones of the forearm: The classic type (also known as ulnar dimelia) in which the forearm contains two ulnae, and the non-classic type in which the forearm contains a radius and one or two ulnae. The main functional problems that need to be resolved are the stiff elbow and the polydactylous hand, which often requires surgery[3]. Here, we report an atypical presentation of Mirror hand and its successful surgical management, where follow-up revealed significant improvement of hand function and appearance. The manuscript has been reported in line with the SCARE 2023 criteria[4].

HIGHLIGHTS

- Mirror hand is an extremely rare congenital abnormality characterized by polydactyly and duplication of the ulna, with the absence of the radius and thumb.
- The management of Mirror hand is a challenge but with wise and good-planed surgical intervention, the outcome is promising.
- The non-classical type of mirror hand is the presence of ulna and radius might demonstrate normal forearm, wrist, and elbow movement due to its normal forearm structures.

Case presentation

A 2-year and 7-month-old female child presented to the clinic with a congenital abnormality of the left upper limb. The patient had no significant perinatal medical issues, and family history did not reveal any congenital anomalies. The child's growth and development were normal, and there were no other associated anomalies. Physical examination revealed 7 well-developed digits, with an absent thumb [Fig. 1], The child exhibited good shoulder movement, minor limitations in supination and pronation, elbow flexion restriction. Notably, the most radial fingers displayed weak movement, while the remaining six fingers were active. X-ray examination revealed a well-formed ulna and radius in the forearm, with fusion proximally. All seven digits had the usual triphalangeal appearance, and the metacarpals had their growth plates at the distal ends [Figure 2]; the previous features correlate with the atypical variant of mirror hand. The surgical plan included disarticulation of the 2nd and 3rd radial supernumerary fingers at the metacarpophalangeal joints, while the most radial finger was used for thumb reconstruction. The

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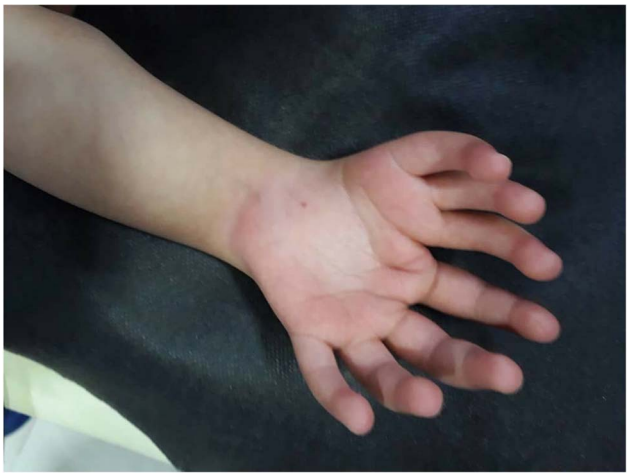

Figure 1. Photographs preoperatively, demonstrating the asymmetrical mirror hand with seven digits.

technique used for thumb reconstruction involved shortening and resection of its metacarpal shaft to simulate the trapezium. The proximal phalanx acted as the new metacarpal, which was rotated and angulated to properly position the new thumb. The metacarpophalangeal joint served as the carpometacarpal joint of the new thumb. The K-wires were used in the first carpometacarpal joint to stabilize the position of the thumb and guarantee its proper alignment with the rest of the hand [Figure 3]. The first dorsal interosseous muscle was converted to an abductor pollicis brevis muscle, and the first volar interosseous muscle was converted to an adductor pollicis muscle to facilitate thumb function. The skin flaps were carefully trimmed to create the palm side and ensure the new thumb had enough tension for grasping, and the skin was closed using vicryl sutures [Figure 4]. A pressure dressing of wet cotton and a plaster cast were applied after surgery, and the surgery was deemed successful. After one month, the cast and K-wires were removed, and the patient commenced a motion therapy regimen. Subsequent improvement was observed, enabling the patient to grasp and pinch. However, after one year, growth and lengthening of the new trapezium (previous metacarpal) were identified, prompting a decision for a second surgery to further shorten the trapezium, which also concluded successfully. The patient's recovery was uneventful, and a 16-month follow-up revealed significant improvement of hand function, with the child now able to use their hand for grasping and other activities, effectively using the thumb for opposition and flexion, and could actively extend and flex the wrist. In addition, the hand had an improved appearance [Figure 5].

Discussion

This case presents an atypical variant of mirror hand, in the presence of a well-formed ulna and radius in the forearm. The fusion between the ulna and radius proximally is a new anatomical abnormality and did not report before. mirror hand or ulnar dimelia is an extremely rare congenital malformation of the upper extremities; it is characterized by symmetrical polydactyly, absence of the thumb, and in the typical some cases, duplication of the ulna. Term "Mirror hand" come from the symmetric fingers position about its midline in the arrangement 1–2–3–4–3–2–1; only about seventy cases have been reported in the literature[5,6]. Al-Qattan *et al.*[2] classified this malformation as a spectrum of 5 types and many subtypes. The two main categories of mirror hand deformity based on the forearm bones: are the classic type, often referred to as ulnar dimelia, in which the forearm has two ulnae with the absence of a radius, and the non-classic type, in which the forearm has a radius and one or two ulnae[3]. Many Skeletal malformations may occur, such as the absence of the radial artery, the duplication of the ulnar artery, the shortness of the radial nerve, or the duplication of the ulnar nerve (occasionally with collaterals to the median nerve)[3]. In this

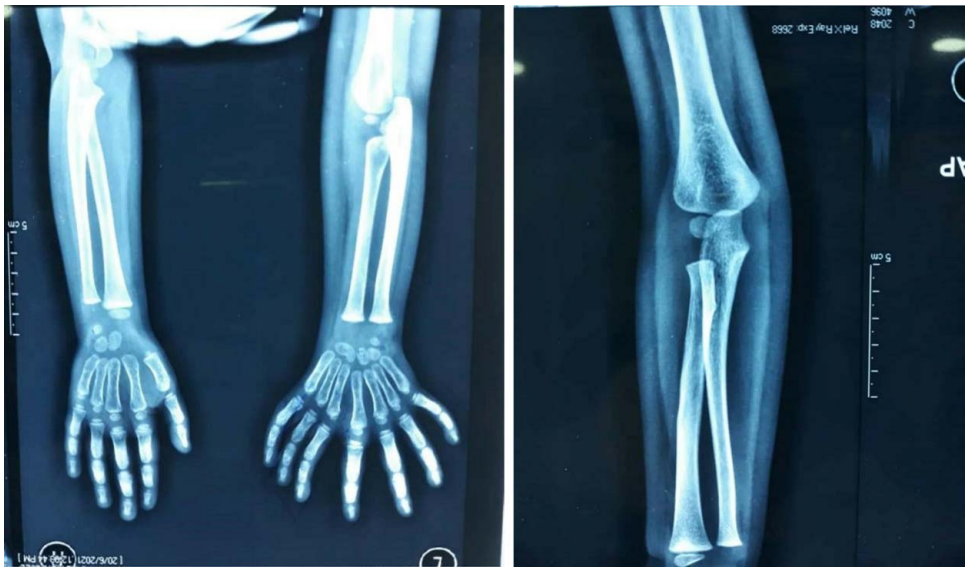

Figure 2. Anteroposterior radiograph of the patient's arms, showing fully developed digits with usual triphalangeal appearance, no thumb, well fully forearm formed from ulna and radius with proximal fusion.

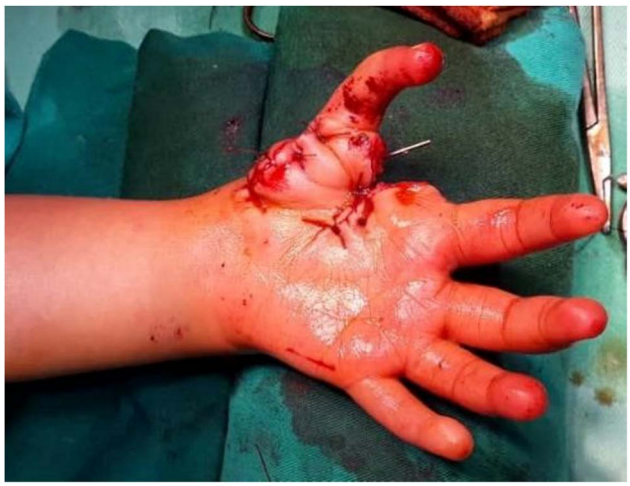

Figure 3. Photograph of the hand after surgery immediate.

case, the patient has seven fully developed digits, no thumb, well-formed forearm formed from ulna and radius with proximal, we did not investigate the inner skeletal malformation as the forearm function was normal, and it did not need any therapeutic intervention. The distal end of the limb bud in the embryo is characterized by the presence of an apical ectodermal ridge (AER) that covers the progress zone, a region of proliferating mesenchyme. The zone of polarizing activity (ZPA) marks the posterior border of this zone. Fibroblast growth factor 4 (FGF4) regulates the AER, controlling limb growth from proximal to distal. The sonic hedgehog (Shh) gene regulates the ZPA, responsible for limb patterning along the anterior to the posterior axis. FGF4 and Shh interact through feedback loops to initiate the expression of secondary signalling molecules, such as homeobox genes and bone morphogenic proteins, coordinating limb growth integration[2]. The pathogenesis of mirror hand is linked to upper limb embryology, where the Zone of Polarizing Activity (ZPA) governs the development of the radio-ulnar dimension of the limb. Mirror hand's aetiology is associated with the duplication of the ZPA signalling centre situated in the posterior margin of the limb bud. This duplicated signalling centre, driven by the sonic hedgehog protein, influences the development of the radio-ulnar axis[3,7]. It seems less likely that any synthetic medication taken during pregnancy is responsible for it[8]. The clinical manifestation of classical mirror hand includes polydactyly, absence of the thumb, hyperflexed wrist and stiff elbow in extension. Underdeveloped muscles and instability can both affect the shoulder, Forearm rotation is extremely restricted. Elbow and wrist movements are frequently also restricted. The non-classical type, the presence of ulna and radius, might demonstrate normal forearm, wrist, and elbow movement due to its normal forearm structures[6,8]. In our case, the patient demonstrated good shoulder movement, little restrictions on supination and pronation, and elbow flexion restriction due to their existing normal radio-ulnar joints with fusion proximal. The diagnosis can be performed simply due to the hand's remarkable and unique appearance[8]. Mirror hand can manifest in a variety forms; the treatment must be adjusted to the characteristics of the affected hand. Using the patient's cerebral plasticity to improve limb usage and function. The appropriate age for surgery is uncertain. Even while early surgery takes advantage of brain plasticity and ease of integration into activities, we must contend with the neurovascular and musculocutaneous systems' small size, and surgical management should be done before the opposite function of the thumb begins, for these reasons, around the second year is a good age[5,9]. The surgery for correction of this anomaly includes excision of supernumerary digits, after preserving 1 digit for thumb reconstruction[7]. Various surgical procedures have been employed to repair the thumb, ranging from doing nothing to formal pollicization[10], which may necessitate numerous complicated and repetitive surgeries. Because mirror hand defects are dynamic in nature, functional and anatomical alterations may occur even after a complete restoration[3]. In the surgical approach to the mirror hand, it is recommended to perform graded surgery and retain the limb in a long arm splint with the elbow flexed, wrist extended,

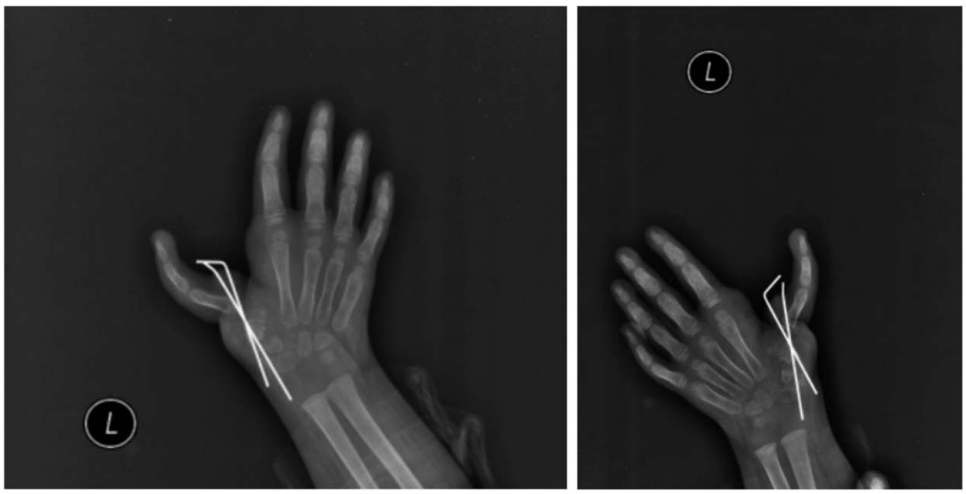

Figure 4. Simple hand X-ray (postoperative): demonstrating the effective correction of mirror hand anomaly and the creation of a new thumb, with temporary stabilization achieved using two K-wires.

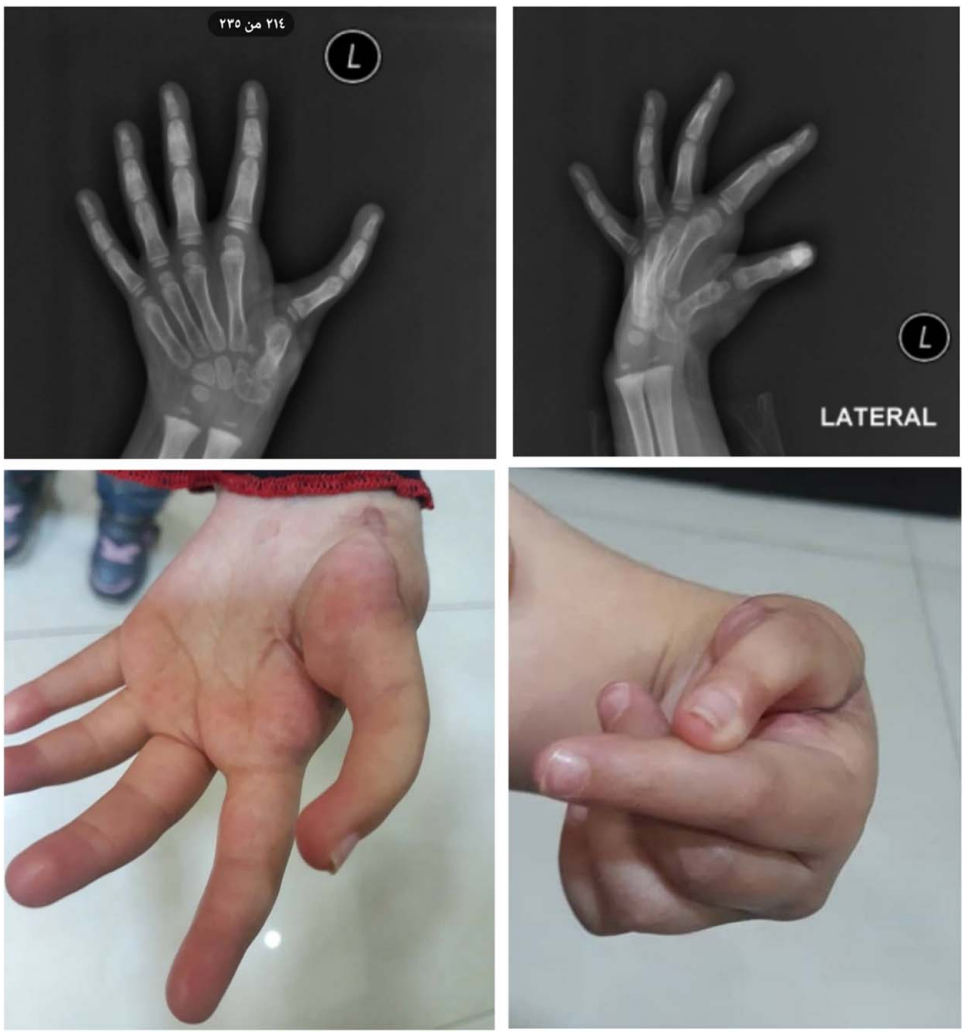

Figure 5. Photographs and radiograph during follow-up show satisfactory results and aesthetic appearance.

and thumb abducted for 4-6 weeks. As a result, it is recommended to do long-term follow-up because surgical repairs are effective at any age, yet the first surgery must be done as soon as possible[11]. However, because there is no double opposing ulna, elbow function and forearm rotation do not have to be addressed. Unpredictable anatomic variations must be considered prior to surgery, and a complete inventory of hand function at the outset can help assess anatomic abnormalities[1].

Conclusion

Mirror hand is an extremely rare congenital abnormality and has a wide spectrum of variants. Managing the mirror hand is a challenge but with early pollicization with appropriate functional considerations, the outcome is promising. However, surgical management of mirror hand has multiple methods, as we presented one of these methods and its outcomes; further analytical studies are needed to identify the best surgical intervention to treat similar future cases.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient's parents for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Not applicable.

Author contribution

H.A. collected the patient's data and drafted the manuscript. M.A. drafted the manuscript, revised the manuscript. M.S. drafted the manuscript. Y.A. collected the patient's data and drafted

the manuscript. A.A. collected the patient's data, performed the procedure, revised the manuscript, and supervised the study.

Conflicts of interest disclosure

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References

[1] Yang SS, Jackson L, Green DW, *et al*. A rare variant of mirror hand: a case report. J Hand Surg Am 1996;21:1048–51.
[2] Al-Qattan MM, Al-Thunayan A, De Cordier M, *et al*. Classification of the mirror hand-multiple hand spectrum. J Hand Surg Br 1998;23:534–6.
[3] Alwatari M, Araji Z, Khalaf A, *et al*. A rare variant of mirror hand deformity. J Surg Case Rep 2023;2023:rjad501; Published 2023 Sep 8.
[4] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg Lond Engl 2023;109:1136.
[5] González-Pola R, Hernández-Álvarez MA, Domínguez-Castro CR, *et al*. Bilateral mirror hand: 2-year follow-up and literature review. Mano en espejo bilateral: seguimiento de 2 años y revisión de la literatura. Acta Ortop Mex 2020;34:261–6.
[6] Winge MI, Bye K. Mirror hand deformity: a report of two cases. JBJS Case Connect 2013;3(4 Suppl 6):e127.
[7] Gaba S, John N, Bhogesha S, *et al*. Mirror hand: an uncommon neglected case managed with pollicisation. World J Plast Surg 2017;6:263–5.
[8] Barton NJ, Buck-Gramcko D, Evans DM. Soft-tissue anatomy of mirror hand. J Hand Surg Br 1986;11:307–19.
[9] Mohamadreza G, Kaveh B, Arash S. Exellent result of a mirror hand anomaly treatment. Iran J Pediatr 2013;23:118–9.
[10] Al-Qattan MM, Al-Kahtani AR, Al-Sharif EM, *et al*. Thumb reconstruction without formal pollicization in mirror hand deformity: a series of four cases. J Hand Surg Eur Vol 2013;38:940–7.
[11] Harpf C, Hussl H. A case of mirror hand deformity with a 17-year postoperative follow up. Case report. Scand J Plast Reconstr Surg Hand Surg 1999;33:329–33.