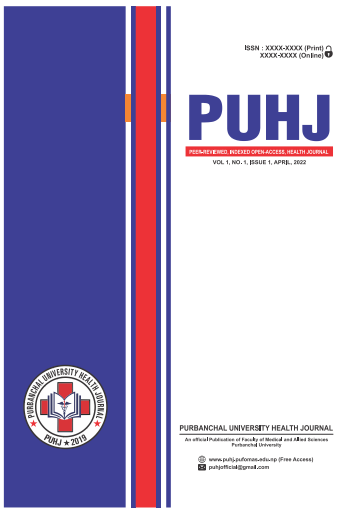

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Non-Syndromic Bilateral Congenital Mitten Hand with Bilateral Polyonychia with Left Radial Polydactyly: A Rare Case Report

Robin Shrestha[1*]

Abstract

Congenital hand conditions are common and many of these conditions are relatively minor and do not affect function. Congenital hand anomalies may occur sporadically or may be the result of various inherited genetic anomalies. A spoon shaped or mitten hand is usually associated with Apert syndrome. Here I am reporting a rare case of non-syndromic bilateral mitten hand with bilateral polyonychia with left pre-axial polydactyly which has never been reported.

Keywords: Apert syndrome; Congenital hand anomalies; Polydactyly

Introduction

The functional area of hand located at the distal end of the upper limb includes five fingers, palm and wrist.[1] The five fingers are normally separated at the metacarpophalangeal joint level. When there is presence of extra digit, it is called as polydactyly while webbing of digits is called as syndactyly. Polydactyly is the condition where there is presence of extra digits. The thumb responsible for 40% of hand function must be able to oppose the other digits with a stable pinch. Radial polydactyly encumbers this motion when the duplicated digits deviate from normal alignment.[2] Webbing of the digits or syndactyly is the most common abnormality of the newborn hand. It happens as an isolated anomaly or as part of a syndrome.[3] When all the five fingers are webbed together, the condition is known as mitten hand and is usually associated with a syndrome.

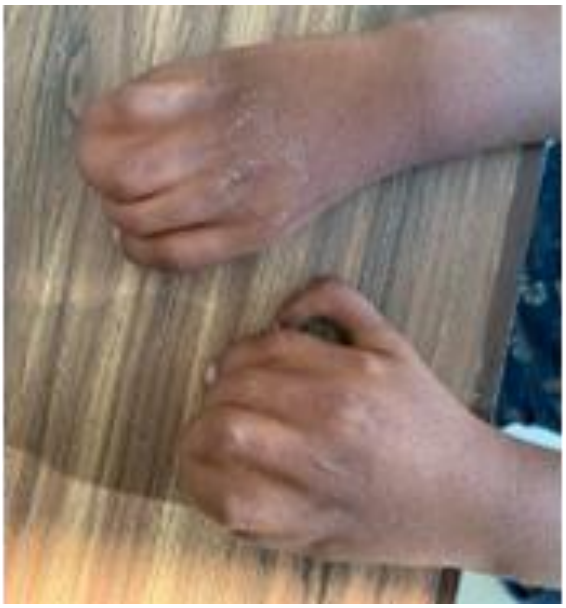
Congenital mitten hand has been most commonly described as part of acrosyndactyly seen in Apert syndrome. The type I Apert hand or "spade" hand is characterized by complex syndactyly of 4 fingers with sparing of the thumb. Type II Apert hand or "mitten" hand demonstrated additional complete simple syndactyly of the first web space too while type III Apert hand or "rosebud" hand is characterized by osseous fusion of all digits resulting in a concave palm and a complete syndactyly of the first web.[4]

A case of mitten hand is rare and that too without any associated syndrome is even rarer. This 4 years old male child not only had bilateral mitten hand, but also had bilateral polyonychia and left Wassel Type VI preaxial polydactyly. Thus, as a rarest of rare case, I present to you this rare variant of non-syndromic bilateral mitten hand.

Case Presentation

A 4 years old healthy male child was brought to the out-patient department with the fused fingers. At the time of the birth, the fused fingers were noted with no other associated abnormality. The developmental milestone was normal except the function of the hand which was halted due to the fused fingers. He is the 5th child in the family with 4 elder sisters. All the siblings are normal and there is no known familial history in both paternal and maternal lineage.

On evaluation, his systemic findings were within normal limit. And on local examination, the hands were cupped with flexion noted at the proximal and distal interphalangeal joints. The thumb and all 4 fingers of the right hand were fused together with presence of 7 nails at the distal end. On the left hand, along with the fused thumb and fingers with 6 nails at the distal end, there was presence of extra digit on the radial side. He could flex the joints to some degree but couldn't extend it. The X-ray of the hand revealed fused distal phalanx of long and ring finger at the distal end on right hand while fused distal phalanx of long, ring and small fingers at the distal end with extra digit radially with its own metacarpal.



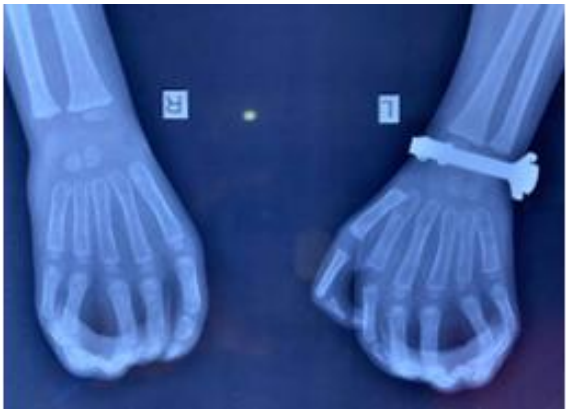


Figure 1: Pre-operative bilateral mitten hand with bilateral polyonychia with left pre-axial polydactyly

The patient parents were counselled regarding the treatment plans and the requirement of at least 2 settings of operation followed by proper physiotherapy. The pre- and post-care along with the probable complications were well counselled. The child was evaluated and underwent pre-anesthetic check-up with the plan for the first setting of operation on 11th September 2022.

Surgical Procedure

The patient was positioned in supine with both arms abducted after giving the general anesthesia. The tourniquet was placed at the right arm level. Right hand 1st webspace creation was planned with triangular flap and straight-line incision marking on dorsum and volar for separation of the thumb and the index finger while a starfish flap was planned for creation of 3rd web space along with straight line incision marking on dorsum and buck gramcko flap marking for pulp creation after separation of the fused distal segment of the distal phalanx along with straight line incision marking for separation of long and ring finger. The incisions were made and meticulous hemostasis was achieved. The cut ends were sutured and dressing was done. There was no requirement of skin graft placement. Then the left-hand marking was done in similar fashion after placing the torniquet on the arm level. After exsanguination, the torniquet was inflated and the surgical procedure was repeated in similar fashion.

He was followed up closely for the next 6 months and on 27th April 2023, the second setting of operation was planned. The 2nd and 4th webspace creation was planned with the starfish flap marking and the separation of the index finger from long finger and the ring finger from the small finger was planned with a straight-line incision marking on dorsum and volar surface. The incisions were made and meticulous hemostasis was achieved. The cut ends were sutured with polygalactic acid 5-0 sutures and dressing was done. This time also there was no requirement of skin graft. The plan for separation of the index and long finger was planned with a starfish flap for creation of the 2nd webspace and a straight-line incision on the dorsum and palmar surface. The 4th webspace creation was planned with a starfish flap creation along with straight line incision marking on dorsum and palmar surface and a buck gramcko flap marking for pulp creation after separation of the fused distal segment of the distal phalanx for separation of the ring and the small finger was done.

Figure 2: After first surgical release

The post operative period was uneventful. He is currently doing the physiotherapy. He now can eat by himself and have started to write and play games using the hand.

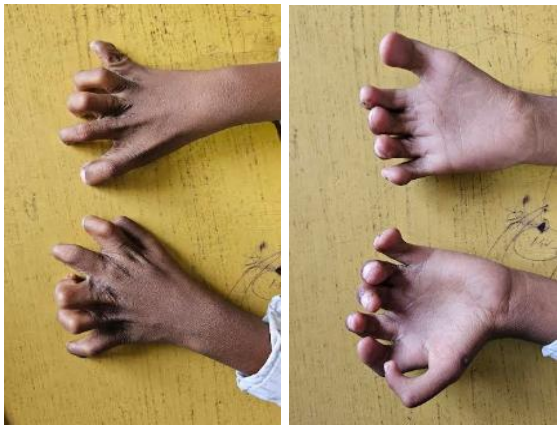


Figure 3: 12 months post-operative condition

Discussion

Congenital hand conditions are common and estimated to occur in at least 2.3 per 1000 of total births. Many of these conditions are relatively minor and do not affect function. Congenital hand anomalies may occur sporadically or may be the result of various inherited genetic anomalies. Environmental factors, diet, infections and other causes are much less common. The hand develops at between four to eight weeks of intrauterine life during embryogenesis and most anomalies have already developed by the time the pregnancy is diagnosed.[5]

There was no known family history, the developmental milestones of the child were normal as per the age and no other abnormalities affecting other part of the body thus the cause in this case is sporadic. Along with the findings of the mitten hand, the left hand had a well-developed extra digit on the radial side. As per the Wassel classification, the thumb duplication in our case is Type VI which accounts for 3 to 4% and as per the Temtamy and McKusick classification, its Preaxial Type 4.[2] Mitten hand is mostly associated with the Apert syndrome but in our case, the developmental milestone was normal.

As the priority of the patient was to separate the fingers, we separated all five in right and all 6 fingers in the left. The functionless hand is now of some function. Before the child grows further, we plan to reposition the thumb in its proper place so that he could further benefit in his functional status.

The release of syndactyly is successful and now he is on physiotherapy. Once the repositioning of the thumb and excision of the polydactyly could be achieved, the treatment will be complete and he will gain more of the confidence and hand function.

Conclusion

Congenital bilateral mitten hand of a 4 years old child which remained functionless has gained the proper digitization and he uses it to feed himself. A rare case but with simple maneuver of syndactyly release, the child has not just gained the confidence but also is achieving the functions of a normal hand.

Acknowledgements

The well-informed written consent was taken from the parents for the publication purpose without disclosing his identity. I would like to thank Dr. Niran Maharjan and Dr. Dhiraj Singh for their invaluable support along with the entire operation theatre team.

Conflict of interest

None

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