Cross Current International Journal of Medical and Biosciences
Abbreviated Key Title: Cross Current Int J Med Biosci
ISSN: 2663-2446 (Print) & Open Access
DOI: 10.36344/ccijmb.2021.v03i01.003

Volume-3 | Issue-1 | January, 2021 |

Case Report

Congenital Right Hand Syndactyly in a child: Plain Radiographic Features and Case Report

Sule MB[1*], Sa'idu SA[1], Ma'aji SM[1], Danfulani M[1], Yunusa GH[1], Umar AU[2], Gele IH[3], Umar FK[3]
[1]Radiology Department, Usmanu Danfodiyo University, Sokoto
[2]Radiology Department, Gombe State University, Gombe
[3]Radiology Department, Usmanu Danfodiyo University Teaching Hospital, Sokoto

***Corresponding author:** Dr. Sule Muhammad Baba

| **Received:** 03.01.2021 | **Accepted:** 16.01.2021| **Published:** 21.01.2021|

Abstract: Syndactyly is a Greek word meaning fused digits, may involve fusion of the soft tissues with or without bony fusion. It is regarded as the most common congenital defect of the hand, detected in 1 out of every 2000 live births, twice as common in males, and more among the Caucasians. This is a 10-month-old male infant that was referred for plain radiograph of the right hand on account of completely fused 2nd to 5th digits and an incompletely fused 1st digit with medial deviation since birth. The plain radiograph showed soft tissue fusion of the 2nd to 4th web spaces completely with a partially fused 1st web space. There is associated fusion of the distal phalanges of the 4th and 5th digit, foreshortening of the 3rd and 4th metacarpals with the middle phalanges of the 2nd and 5th digits. There is flexion of the interphalangeal joint and medial deviation of the 1st digit. There is also haphazard arrangement of the metacarpals and phalanges most especially the 3rd and 4th digits. Prominence of the thenar and hypothenar eminences with mild ulnar deviation of the entire right hand were also demonstrated. The contralateral left hand appears within normal limits, a complementary ultrasound showed normal abdominal organ situs. A diagnosis of congenital syndactyly most likely the complex-complicated form in a 10-month-old infant was established. The surgical repair to establish function and achieve excellent aesthetics was deferred until the child clocks about 24-months of age. We report a case of congenital syndactyly of the right hand due to its radiographic form of presentation in this 10-month-old infant.

Keywords: Web space, Congenital, Digits, Anomalies.

INTRODUCTION

Syndactyly means fusion of adjacent digits, this fusion may either involve the soft tissues or the soft tissues with or without bony fusion. It is regarded as the most common congenital hand malformation mostly found among the Caucasians with an estimated incidence of about 1 in 2000 to 2500 live births [1-5].

Syndactyly is a congenital anomaly of the hand seen more in males than females, and occurs with equal frequency unilaterally or bilaterally and has an autosomal dominant trait with variable expression or reduced penetrance [4-7].

Syndactyly occurs as a congenital anomaly due to failure of separation of fingers at the 6th week of gestation, the fingers are webbed in-utero, these webs are separated by apoptosis, this is followed by skin recession that allow formation of digital interspaces, the complete interdigital spaces are formed at the end of the 6th week of gestation[8].

Anatomically, syndactyly can be classified as either simple or complex, and complete or incomplete. The simple form involves only soft tissues, while the complex form includes side-to-side bony fusion. When the adjacent digits are fused up to the fingertip; this is the complete syndactyly, while it is referred to as incomplete syndactyly when the bony fusion is partial [4].

Syndactyly has also been classified phenotypically, the phenotypical classification happens to be more specific for the affected digits, this has led to syndromic and non-syndromic syndactylies described [4].

The non-syndromic syndactylies only involves the digit and appendage malformation, these has been further classified as nine phenotypes; the syndactyly I to IX, some of these phenotypes are also ascertained by their synonyms and predominantly inherited as an autosomal dominant trait [4, 9, 10, 11].

Quick Response Code

Journal homepage:
https://crosscurrentpublisher.com

Citation: Sule MB *et al* (2021). Congenital Right Hand Syndactyjy in a Child: Plain Radiographic Features and Case Report. *Cross Current Int J Med Biosci*, *3*(1), 9-12.

Syndactyly is a congenital anomaly, with a strong familial tendency, most often bilateral and commonly seen in males and usually seen in the third web space. The congenital syndactyly can either be primary which occurs as a failure of differentiation or separation, the secondary form of congenital syndactyly occurs as an outcome of antecedent events and produced by refusion abnormality [8].

Association of syndactyly with syndromes is referred to as the syndromic form of syndactyly; some of the syndromes are Poland's, Cleft hand or Aperts [4, 8].

Syndactyly has varying presentation, among which is the severe presentation, this is the complex-complicated syndactyly, this shows skeletal deformity with associated tendon and neurovascular abnormalities, the incidence of the complex-complicated syndactyly increases as the complexity of the syndactyly increases [1, 12].

The diagnosis of syndactyly follows a detailed clinical examination for proper classification of the syndactyly; this is often followed by plain radiography of the affected hand to rule out synostoses, hidden polydactylies and other skeletal deformities [4, 8].

The main treatment of syndactyly is by surgical repair; these include separation of the fused digits, normal web spaces creation with achievement of normal function of the hand, and form an acceptable aesthetical web with minimal complication [8, 13].

The prominence of the thenar and hypothenar eminences has been reported to be idiopathic in most instances, the unilateral cases of such prominence following biopsies showed slight variation from normal cases of increase in muscle fiber size, absence of features of inflammation, pathological blood vessels, fibrosis, or loss of muscle fibers and changes favoring loss of nervous supply were also absent [14-17].

CASE REPORT

This is a 10-month-old male infant that was referred to the radiology department for a plain radiograph of the right hand with fused digits since birth.

The child is the 6th child of a 43-year-old house wife; she admitted to have not adequately attended her antenatal visits and also took a lot of local herbs during the pregnancy of the index case. She also admitted to nonoccurrence of similar deformities among siblings and relations**.**

The patient is conscious and alert, well fed, not dehydrated, not pale, anicteric, not in obvious respiratory and painful distress.

The digits of the right hand appear fused; complete fusion of the medial three web spaces with partial fusion of the 1st web space distally, no any fused digit demonstrated to involve the remaining limbs. Non-tender swelling/prominence demonstrated to involve the region of the thenar and hypothenar eminences.

The patient had a normal blood pressure of 95/60mmHg, normal pulse rate of 100 beats per minute and a normal respiratory cycle of about 16 cycles per minute.

The packed cell volume was normal and about 40%, normal white cell count of 9000/mm^3, and a normal ESR; erythrocyte sedimentation rate of 8mm/hr.

The blood electrolytes were normal; the blood urea and creatine levels were also within normal limits for the patient's age.

The plain radiograph of the right hand done in oblique projection showed soft tissue fusion of the 2nd to 4th web spaces completely with a partially fused 1st web space. There is associated fusion of the distal phalanges of the 4th and 5th digit, foreshortening of the 3rd and 4th metacarpals with the middle phalanges of the 2nd and 5th digits. There is flexion of the interphalangeal joint and medial deviation of the 1st digit. There is also haphazard arrangement of the metacarpals and phalanges most especially the 3rd and 4th digits. Prominence of the thenar and hypothenar eminences with mild ulnar deviation of the entire right hand were also demonstrated (see figure 1). The contralateral left hand appears within normal limits.

A complementary ultrasound showed normal abdominal organs with normal situs. A diagnosis of congenital syndactyly of the right hand most likely the complex-complicated form in a 10-month-old infant was established.

The patient is been prepared for surgical repair of the syndactyly to achieve web space formation, with good restoration of the function of the right hand and excellent aesthetics of the created web spaces and hand when the child reaches 24-months of age.

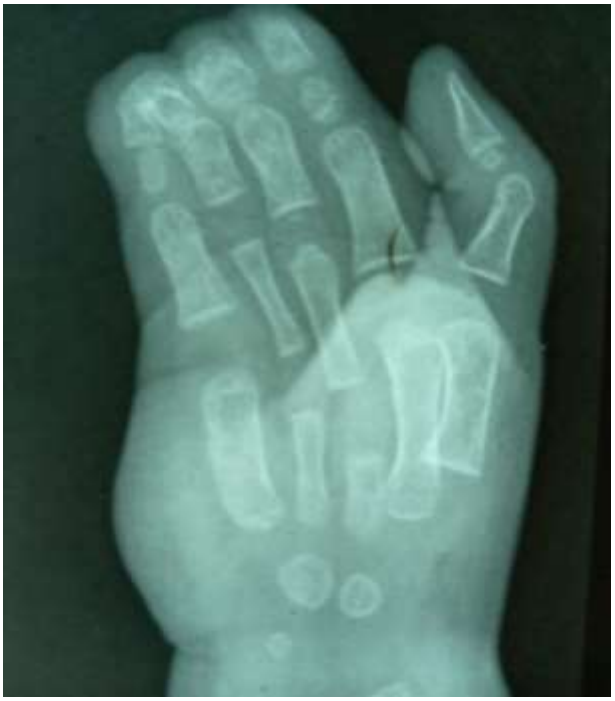

Fig-1: Plain radiograph of the right hand, an oblique view, showing completely fused 2nd to 4th web spaces, with incomplete fusion of the 1st web space with medial deviation of the 1st digit, and flexion of the interphalangeal joint of the 1st digit, fused distal phalanges of the 4th and 5th digits, foreshortened metacarpals (3rd and 4th) and phalanges (middle phalanges of 2nd and 5th digits) with haphazard arrangements of the phalanges especially of the 3rd and 4th digits. Prominence of the thenar and hypothenar eminences with mild ulnar deviation of the entire right hand are also demonstrated

DISCUSSION

Syndactyly is a congenital anomaly of the hand seen more in males than females, and occurs with equal frequency unilaterally or bilaterally and has an autosomal dominant trait with variable expression or reduced penetrance [4-7]. The index case is a male child, born with fused digits only seen on the right hand, only member of the family affected most likely from reduced penetrance, thereby conforming to these literatures.

Syndactyly means fusion of adjacent digits, this fusion may either involve the soft tissues or the soft tissues with or without bony fusion, and mostly found among the Caucasians [1-5]. The case under review has fusion of the web spaces between the 2nd to 6th digits with fusion of the distal phalanges of the 4th and 5th digit, though he is of the African ancestry.

It has been observed that syndactyly of the right finger has predilection for the web space between the middle and ring finger, followed by the index and middle finger web space[8]. The index case had the right hand affected and all the web spaces mentioned above were also involved, affectation of additional web spaces were also noted, thereby conforming to this literature.

Anatomically, syndactyly can be classified as either simple or complex, and complete or incomplete. The simple form involves only soft tissues, while the complex form includes side-to-side bony fusion. When the adjacent digits are fused up to the fingertip; this is the complete syndactyly, while it is referred to as incomplete syndactyly when the bony fusion is partial[4]. The index case had complete fusion of the web spaces; 2nd to 4th, with fusion of the distal phalanges of the 4th and 5th digits; bony fusion, these raise a suspicion of a complex syndactyly in conformity to this literature.

The complex-complicated syndactyly shows skeletal deformity with associated tendon and neurovascular abnormalities, the incidence of the complex-complicated syndactyly increases as the complexity of the syndactyly increases [1, 11]. The case under review shows some complexity in terms of fused web spaces with skeletal affectation of varying forms raising a suspicion of the complex-complicated form of syndactyly, conforming to these literatures.

The fused digits; syndactyly has been classified as syndromic or the non-syndromic syndactylies, the non-syndromic form only involves the digit and appendage malformation, these have been further classified as nine phenotypes; the syndactyly I to IX, some of these phenotypes are also ascertained by their synonyms [4, 9, 10]. The case under review is most probably the non-syndromic form without obvious features to suggest any syndrome, the features and form of fusion resembles the syndactyly class VI; Mitten appearance. There are other radiographic features demonstrated in this patient in addition to the complete fusion of the 2nd-4th web spaces as seen in Mitten form of non-syndromic syndactyly.

The diagnosis of syndactyly follows a detailed clinical examination for proper classification of the syndactyly; this is often followed by plain radiography of the affected hand to rule out synostoses, hidden polydactylies and other skeletal deformities [4, 8]. The case under review had a clinical examination that showed fused 2nd to 5th digits, this was followed by a confirmatory plain -radiography of the right hand, this confirmed the fused web spaces, fusion of the distal phalanx of 4th and 5th digits, foreshortening of the metacarpals (3rd and 4th) and middle phalanges, with haphazard arrangement of the phalanges of the 3rd and 4th digits, thereby conforming to these literatures.

The prominence of the thenar and hypothenar eminences is usually congenital and idiopathic and show some changes following biopsy [14-17]. The index case had the prominence since birth and obvious cause of the prominence of this group of muscles was not identified; however, biopsy was not performed in this case.

The main treatment of syndactyly is by surgical repair, these include separation of the fused digits and restoration of normal web spaces to achieve normal function of the hand, and form an acceptable aesthetical web with minimal complication, these repair most often done at 18 to 24 months [4, 8, 11]. The index case is been planned for surgical repair for the syndactyly at the age of two-years in conformity to these literatures.

CONCLUSION

Syndactyly is often diagnosed by clinical examination and plain radiography; the plain radiography confirms the diagnosis and helps in ruling out bony synostoses and other associated anomalies.

REFERENCES

1. Jordan, D., Hindocha, S., & Dhital, M. (2012). The Epidemiology, Genetics and future Management of Syndactyly, The Open Orthop. *J*, *6*, 14-27.
2. Burke, F.D., McGrouther, D.A., Smith, P.J. (1989). Principles of hand surgery; 15:256.
3. Eaton, C.J., Lister, G.D. (1990). Syndactyly. Hand Clin; 6:555.
4. Dao, K.D., Shin, A.Y., Billings, A., Oberg, K.C., Wood, V.E. (2004). Surgical Treatment of Congenital Syndactyly of the Hand. *J Am Acad Orthop Surg*, 12:39-48.
5. MacCollum, D.W. (1940). Webbed fingers. *Surg Gynecol Obstet*, 71:782-789.
6. Kettlekamp, D.B., Flatt, A.E. (1961). An evaluation of syndactylia repair. *Surg Gynecol Obstet*, 113:471-478.
7. Kelikian, H. (1974). (ed): Syndactyly, in Congenital Deformities of the Hand and Forearm. Philadelphia, PA: WB Saunders, 331-407.
8. Nangineedi, N., Harish, G.P., Rafi, M. (2019). Management of syndactyly: a clinical study. *Int Surg J*, 6:2806-2812.
9. Goldstein, D.J., Kambouris, M., Ward, R.E. (1994). Familial crossed polysyndactyly. *Am J Med*, 50:215-223.
10. Malik, S., Percin, F.E., Ahmad, W. (2005). Autosomal recessive mesoaxial synostotic syndactyly with phalangeal reduction maps to chromosome 17p 13.3. Am J Med Genet A, 134:404-408.
11. Temtamy, S.A., McKusick, V.A. (1978). The Genetics of Hand Malformations. New York: Alan R. *Liss New York*, 301-322.
12. Canale, S.T., Beaty, J.H., Eds. (2008). Campbell's operative orthopaedics. 11th ed. Philadelphia: Mosby Elsevier, 4403-4404
13. Mei, H., Zhu, G., He, R., Liu, K., Wu, J., Tang, J. (2015). The preliminary outcome of syndactyly management in children with a new external separation device. *J Pediatr Orthop B*, 24:56-62.
14. Kalay, T., Gilhuis, H.J., Kraan, G., van Alfen, N. (2016). Congenital Bimelic Hypertrophy of the Hands. *Case Rep Neurol*. 2016; 8:34-38.
15. Mirastschjski, U., Damert, H.G., Mawrin, C., Schneider, W. (2009). Myopathic changes in bilateral hypertrophy of the first interosseus muscle of the hand. *J Neurol*, 256:1551-1554.
16. Gilhuis, H. J., Zöphel, O. T., Lammens, M., & Zwarts, M. J. (2009). Congenital monomelic muscular hypertrophy of the upper extremity. *Neuromuscular Disorders*, *19*(10), 714-717.
17. Takka, S., Doi, K., Hattori, Y., Kitajima, I., & Sano, K. (2005). Proposal of new category for congenital unilateral upper limb muscular hypertrophy. *Annals of plastic surgery*, *54*(1), 97-102.