Complete Simple Syndactyly

Selukar Mangesh S.[1], Alka K. Bhandare[2], Kulkarni Promod R.[3]

[1]Associate Professor, [3]Professor and HOD, Department of Anatomy, Government Medical College, Latur-413512, Maharashtra, INDIA.

[2]Associate Professor, Rachna Sharir, Dhanwantari Ayurved Medical College, Udgir, Latur, Maharashtra, INDIA.

[1]mangesh70201@rediffmail.com, [3]pramkul54@yahoo.co.in, [2]bakreader_10@rediffmail.com

Case Report

Abstract: Syndactyly is the most common congenital malformation of the limbs, with a large aesthetic and functional significance. Syndactyly is a failure of differentiation in which the fingers fail to separate into individual appendages. This separation usually occurs during the sixth and eighth weeks of embryonic development. The syndactyly word is derived from Greek words *syn,* meaning together, and *dactyly*, meaning fingers or digits. The incidence of syndactyly 1 in 2000-3000 live births.There is currently nine types of phenotypically diverse non syndromic syndactyly, an increase since the original classification by temtamy and McKusick [12]. Non-syndromic syndactyly is inherited as an autosomal dominant trait, although the more severve presenting types and sub types appear to have autosomal recessive and in some cases X-linked hereditary [6]. Gene research has found that these phenotypes appear to not only be one gene specific, although having individual localized loci, but dependent on a wide range of genes and subsequent signaling pathways involved in limb formation. Research into the individual phenotypes appears to complicate classification as new gens are found both linked, and not linked, to each malformation. Consequently anatomical, phenotypical and genotypical classification can be used, but variable in significance. Currently, management is surgical, with a technique unchanged for several decades, although future development will hopefully bring alternatives in both earlier diagnosis and gene manipulation for therapy.

Keywords: Congenital, genetics, limb deformity, complex.

Introduction

Syndactyly is joining or webbing of two or more fingers due to a failure of differentiation of parts in upper limb [1]. Syndactyly may involve fusion of the soft tissue with or without bony fusion. It mainly occurs due to failure of differentiation between adjacent digits caused by the absences of adoptosis in the interdigital mesenchyme [3]. The separation failure occurs between 6 to 8th week of intrauterine life and the condition may recur in affected families. The third, fourth, second and first web spaces are affected in decreasing frequency with around 57% of cases occurring in the third web space [4]. The involvement is bilateral and symmetrical in about 50 % of cases. The male to female ratio is 2:1. Syndactyly of adjacent fingers is extremely common but complete simple Syndactyly of one hand and complete complex Syndactyly of other hand is a rare anomaly [5]. Syndactylism is most common congenital anomaly of the hand with an incidence of 1 in every 2000 – 2500 births. Complete simple in one hand and complete complex in other hand is rare.

Case Report

A 22 year old male presented to surgery clinic with webbing of middle and ring fingers of both hands and distal phalanx of index finger of both hands directed laterally. He was first born child of normal parents, born out of non-consanguineous marriage. Family history was contributory, suggesting that the persons from mother-side were affected by Syndactyly.

On Examination

There is Syndactyly of middle and ring fingers and distal phalanx of index finger is directed laterally. Active movements are present in all digits. Soft tissue nodules were present in Syndactyly fingers. Motor and Sensory system is normal. All other developmental milestones are normal. There is no other congenital anomaly.

X-Ray Findings

Right Hand

3rd metacarpal shows bony synostosis (duplicate). Proximal and distal phalanx of middle and ring finger is united. Distal phalanx of index finger is directed laterally

Left Hand

Proximal and distal phalanx of middle and ring finger is united. Distal phalanx of index finger is directed laterally. On examination and X-Ray findings, suggest that this is a case of Complex Syndactyly.

Discussion

Syndactyly can be classified in several ways. Anatomically the syndactyly is either simple or complex, and complete or incomplete [3]. In **Simple** Syndactyly, fingers are joined by soft tissues. In **Complex** Syndactyly, fingers are united by bone or cartilage and union is usually in a side to side fashion at the distal phalanges. In **Complete** simple Syndactyly, soft tissue union extends up to finger tips. In **Incomplete** simple Syndactyly, soft tissue union is partial and does not extend to the finger tips. In Complicated Syndactyly, fingers are united by bone and include bony abnormalities such as extra, missing or duplicated phalanges. Abnormalities in musculotendinous and

neuro-vascular structure may be present [2]. Most common Syndactyly is between middle & ring finger (57 %), between little & ring finger (27 %) between middle & index finger (14 %) and least frequent between thumb & index finger (03%).10 to 14 % of cases have a family history and remaining cases are sporadic [7]. Syndactyly can be an isolated findings or seen with other anomalies such as acrosyndactyly, synotosis, cleft hand and polydactyly or as a feature in several syndromes including Apert, Poland's, Pfeiffer, Jackson-Weiss and Holt-oram. Temtamy and McKusick concluded in 1978, that there were at least five phenotypically different types of syndactyly involving the hands, with or without foot involvement [12]. The majority of these are thought to be inherited as autosomal dominant traits. Within each pedigree there is uniformity of the type of Syndactyly, allowing for the variation characteristic seen in dominant traits. The non syndromic syndactylies appear to only involve digit and appendage malformation, and have since been expanded to nine phenotypes, named Syndactyly I to IX [6]. The syndromic syndactylies are linked to other abnormalities in the body and these appear to occur alongside the digit anomalies during fetal development.

Management Principles

The current mainstay for the treatment and management of Syndactyly is operative. The indication for operative is functional and aesthetic consideration [8].

Surgical excision of Polydactyly.

In case of Syndactyly with multiple digit involvement. The Syndactyly involving the thumb, index, ring fingers is felt to benefit from earlier release, usually between the ages of 6 to 12 months and should always be released first in multiple digit involvement [7]. The result in delayed surgery can involve deformity of the digit relating to forced flexion, angulation and/or rotation. Release of Syndactyly should be performed earlier, because early release prevents the malrotation and angulation which is developed from differential growth rates of involved fingers.

In Complex Syndactyly, release of border digits is done at 6 month old age. If more than One Syndactyly is present in the same hand, simultaneous surgical release can be performed [9]. Perform bilateral release whenever feasible to reduce the number of surgeries. Prolong Syndactyly between these digits can cause permanent deformities.

Full thickness skin grafts and split thickness skin grafts are used for soft tissue coverage. Full thickness skin grafts are preferable over split thickness skin grafts because the former are less likely to contract over time and because they usually grow with the patient [10]. The most common full thickness skin graft donor site is the lateral inguinal region close to the anterior superior iliac crest. This area is generally hairless, even in hirsute adult. The skin is taken as an ellipse, and the donor site is closed primarily.

Complications

Re-occurrence of Syndactyly- Distal migration of web can occur, even after successful release. The causes of recurrence are multifactorial. If recurrence is early, it is due to presence of synechiae between the incision lines and poor take of skin graft [11]. Finger Ischemia- It is rare complication but when occurs due to developmental anomaly with the digital vessels or digital vessles are damaged.

Contracture

Contracture develops along the length of incision lines. This can result in a scar contracture and angulation of the finger. To prevent further angulation a Z-plasty or skin graft will be needed to release this contracture [13]. Skin graft complications- Partial skin graft loss results in an open wound that heals by secondary intention.

Conclusion

In conclusion, there are nine types of phenotypically diverse Syndactyly. The non syndromic Syndactyly is inherited as an autosomal dominant trait. New syndromes, genes and causative loci are being found as research continues into congenital hand defects and it appears that each new finding gives as many further investigation as answers in this field. Management is still by surgical correction, and requires optimal planning in terms of patient age and degree of malformation present. This is a non urgent procedure and it must take into account the psychological and social impact on the child. Despite increasing knowledge of the causes of Syndactyly, management has not changed greatly over the years, although the future is likely to see new techniques involving gene manipulation and tissue engineering at least to be created and studied, although current management is low risk. We highlight the rarity of this clinical presentation. Also, we conclude that gene expression being responsible for this malformation.

References

1. Benson MKD, Fixen JA, Macnicol MF, Parsch K. Childern's orthopaedics and fractures. 2nd ed. London: Churchill Livingstone; 2002. Pp. 306-7.
2. Canale ST, Beaty JH, editors. Campbell's operative orthopaedics. 11th ed. Vol 4. Philadelphia Mosby: Elsevier; 2008. Pp. 4403-4.
3. Castilla E.E., Paz J.E.: Syndactyly: Frequency of specific types. Am J Med. Genet, 5: 357-364, 1980
4. De Smet L, Mulier T, Fabryg : Syndactyly of Ring & Small Finger Genet. Counsel 5 : 45-49, 1994.

5. Gillessen – Kaesbach G, Majewski F : Bilateral Complete Polydactyly. Am J. Med. Genet 38, 29-31, 1991.
6. Goldstein DJ, Kambouris M, Ward RE. Familial crossed polysyndactyly. Am J Med Genet. 1994; 50:215-23.
7. Green DP, Hotchkiss RN, Pederson WC, Wolfe SW. Green's operative hand surgery. 5th ed vol 2. Philadelphia PA: Elsevier; 2005. Pp. 1381-2.
8. Hass S.L.: Bilateral Complete Syndactylism of all fingers. Am J. Surg. 50, 363-366, 1940
9. Johnston O, Kirby V.V.: Syndactyly of the ring & little finger. Am J. Hum. Genet. 7, 80-82, 1955.
10. Kettelkamp DB, Flatt AE. An evaluation of syndactylia repair. Surg Gynecol Obstet. 1961; 113:471.
11. Kozin SH. Syndactyly. J Hand Surg Am. 2001; vol.1:1.
12. Netscher DT, Baumholtz MA. Treatment of congenital upper extremity problems. Plast Reconstr Surg. 2007; 119(5):101e-29e.
13. Temtamy SA, McKUsick VA. The genetics of Hand Malformations. New York:: Alan R. Liss New York;1978. Pp.301-22
14. Toledo LC, Ger E. Evaluation of the operative treatment of syndactyly. J Hand Surg Am. 1979;4(6):556-64.

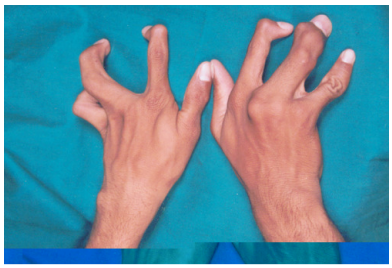

Figure 1: Showing complete simple syndactyly

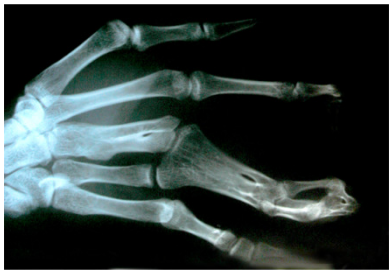

Figure 2: Showing Radiograph of right hand

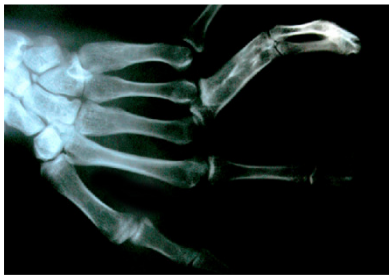

Figure 3: Showing radiograph of left hand