Isolated hexadactyly of foot with central polydactyly

INTRODUCTION

Presence of supernumerary digits i. e. more than five digits per limb is known as polydactyly.[1] This is a congenital duplication type of anomaly of fingers or toes leading to development of more than five digits or toes. Polydactyly is relatively common condition affecting the African American population with an incidence of 13. 9 per 1000 live birth and 0. 3 in 1000 live birth in Caucasian population.[2] Despite extensive search of the literature we were not able to find the recent incidence of the polydactyly of foot. Polydactyly has been classified as preaxial, central, post axial depending on the location of duplication.[3]80% cases of foot polydactyly are postaxial, 15% of the polydactyly cases are preaxial and the least common being the central polydactyly of foot.[4, 5] Males and females are almost equally affected and unilateral involvement is twice as common as bilateral involvement. Polydactyly of foot is less frequent and has different genetic basis than hand polydactyly.[6] The polydactyly may present as isolated anomaly or associated with established genetic syndromes.[7]30% of non-syndromic cases have positive family history and mostly expressed as autosomal dominant with variable penetrance. [8]Environmental and genetic factors have also been reported in the aetiology of polydactyly. Mutation of GLI3 gene and other loci on chromosome 7q21-q34 and 19p13. 2 have been identified for this anomaly. [9, 10] Ellis -van crevaldsyndrome, trisomy 13, tibial hemimelia and trisomy 21 are some of the syndromes associated with polydactyly.[11, 12]

We report a case of isolated hexadactyly of foot with central polydactyly. In this case there is metatarsal bifurcation without any intermetatarsal

widening since the extra toe though with a angular deviation of 45^{o}is not in the transverse plane of foot. Patient’s parents seek surgery due to inability to wear as shoe, repeated injury due to trauma to the extra toe and for cosmetic reason.

CASE PRESENTATION

A 4 year male child consulted to our department with complains of poor cosmesis and difficulty in wearing footwear due to extra toe of Right foot (Figure 1). On examination, there was a single supernumerary toe over the second metatarsal bone. The extra toe was not in the plane of the foot adjacent to the normal digit but inclined at 45^{o} to the transverse arch of the foot. It was well developed with a rudimentary nail and having normal sensation. There was no associated syndactyly. On roentgenogram, the bony structure of the supernumerary digit was well developed with T shaped bifurcation of 2 nd metatarsal bone proximal to head of metatarsal (Figure 2). Interestingly, the T shaped bifurcation of the metatarsal was angled at 45^{o} to the transverse arch of the foot. Family history was negative for similar or other congenital deformities. Maternal history was uneventful. We describe this malformation as rare case because there was no intermetatarsal widening despite complete duplication of phalange as T- bifurcation of metatarsal was in a plane vertical to the longitudinal axis of toes. Both longitudinal and transverse arch of the foot maintained. Patient had no other congenital malformation and growth and development was normal. The treatment involved was surgical removal of extra toe with excision of T shaped bifurcation of metatarsal and repair of periosteum. After surgical

removal of extra toe the child was able to wear shoes and walk normally (Figure 3).

DISCUSSION

Polydactyly is most common congenital anomaly of foot, accounting to 45% of congenital foot deformities. [13]There are different methods of classification for foot polydactyly. Classification system is usually based on morphological presentation.

Venn-Watson proposed a morphological classification based upon the anatomical configuration of metatarsals & the duplicated bony parts. The metatarsal variations are T shaped, Y shaped, widened fifth metatarsal head, complete fifth metatarsal duplication, short block first metatarsal head.[14] Blauth and Olasonclassified each individual malformation longitudinally according to the level of bifurcation such as distal phalangeal, middle phalangeal, proximal phalangeal, metatarsal or tarsal. All the toes are given roman number I-V starting from Great Toe to little Toe.[15] Watanabe et al. defined morphological classification upon ray involvement and level of duplication . They Classified foot polydactyly into three major groups of medial, central and lateral and each group was subdivided into tarsal, metatarsal, proximal phalangeal and distal phalangeal sub groups. [16]

Recently, Seok HH introduced new classification (Table 1). Theoretically 27 permutations or classes are possible. Each group has its own significance; Sreflects the need for syndactyly release, need for SSG, Areflects the axis deviation and reflects the need for wedge osteotomy during surgery, M reflects the metatarsal duplication status, and after excision maintenance of

transverse arch of the foot which plays important role to support the body. [12]

Our case cannot be classified as per the SAM classification because the classification does not identifies the deviation of the duplicated metatarsal in vertical plane.

Indication for surgical management in cases of polydactyly is difficulty in wearing footwear, repeated trauma to the extra digit due to its abnormal position, cosmetic and psychological concerns. Though the surgical management of syndactyly is straight forward; to achieve satisfactory surgical results the surgical plans should be individualised. The M1and M2 variety (SAM classification) with metatarsal extension cases should be treated with care to prevent the residual bone deformity. Widened metatarsal arch may require repair of the transverse metatarsal ligament.

In our case the polydactyly was central involving the second ray with T shaped bifurcation of 2 nd metatarsal proximal to the metatarsal head. The extra toe was not in the plane of the normal toes. The bifurcated metatarsal and the extra digit was angulated at 45^{o} to the transverse arch. In this case the polydactyly toe was dorsal in position without any intermetatarsal widening thus bony excision with repair of the periosteum was done with good post -op outcome.

After surgical removal of the extra toe there was no residual deformity of the metatarsal and intermetatarsal widening with satisfactory cosmetic and functional outcome.

CONCLUSION

The central polydactyly is rare type of polydactyly of foot. Central polydactyly with metatarsal extension needs particular attention for surgical correction as there is post-op residual intermetatarsal widening. Our case was a rare variety of central polydactyly because the extra toe was dorsally oriented with T bifurcation 45^{0} angulated vertically to the transverse arc of foot. So there is no metatarsal widening despite central polydactyly. This type of polydactyly does not fit into any of classification described previously by the authors. The previous classifications need to be modified to include central polydactyly with vertical oriented T bifurcation of metatarsal bone without intermetatarsal widening.

REFERENCES

1. Ahmed H, Akbari H, Emami A, Akbari MR. Genetic Overview of Syndactyly and Polydactyly. Plast Reconstr Surg Glob Open. 2017; 5 (11).
2. Castilla E Paz J, Mutchinick O, Mounz E, Giorgiutti E, Gelman Z. Polydactyly: a genetic study in south America. Am J Hum Genet. 1973; 25(4); 405-412.
3. Temtamy S, McKusick V A. The genetics of hand malformations with particular emphasis on genetic factors. Birth defects, 1969; 14: 364-423.
4. Bethur MV, Linton JL, Barnes DA. The spectrum of pre axial polydactyly of foot. J Pediatric Orthop 2011; 31(435-47)

5. Phleps DA, GorganDP, Polydactyly of the foot. J Pediatric Orthop. 1985; 5(4) : 446-51.
6. Castile E. E da Grace Dutta M, Lugarinho de Fonseca R, Paz JE. Hand and foot postaxial polydactyly; two different traits. Am J Med Genet 1997; 73(1); 48-54.
7. Morrison BE, D Mello SR, Polydactyly in mice lackingHDACq/HDRP. Exp. Biol Med.(Maywood)2008; 233(8): 980-8.
8. Zguricas J, Baker WF, Heus H, Lindhout D, Heutnik P, Hovius S E, Genetics of limb development and congenital hand malformations. Plast Reconstructive Surg. 1998; 10194: 1126-35.
9. Radhakrishna U, Bornholdt D, Scott HS, Patel UC, Rossier C, Engel H. The phenotypic spectrum of GLI3 morphopathies includes autosomal dominant preaxial polydactyly type-IV and post axial polydactyly type A/3; No phenotypic prediction from GLI3 mutations. Am J Hum Genet. 1999; 65(3): 645-55.
10. Galjaard RJ, Smits AP, Tuerlings Jh, Bias AG, Bertoli Avella AM, Breedveld G. A new locus for postaxial polydactyly typeA/B on chromosome 7q21-q34. Eur J Hum Genet 2003; 11(5): 409-15.
11. Enkins S, Morrelt DS, Ellis-Van Creveldsyndrome : Case report and review of literature, cutis 2009; 303-5.
12. Synd KL, Roelker S, Ramchandran V, Durbin L, Bensin D W. Analysis of Ellis Van Creveld syndrome gene products: implications for cardiovascular development and disease. Hum Mol Genet 2009; 18(10: 1813-24.

13. Seok HH, Park JU, Kwon ST. New classification of polydactyly of the foot on the basis of syndactylism, axis deviation, and metatarsal extent of extra digit . Arch Plast Surg. 2013; 40(3): 232-7.

14. Venn-Watson EA. Problems in polydactyly of the foot. Orthop Clin North Am. 1976; 7(4): 909–27.

15. Blauth W, Olason AT. Classification of polydactyly of the hands and feet. Arch Orthop Trauma Surg. 1988; 107(6): 334–44.

16. Watanabe H, Fujita S, Oka I. Polydactyly of the foot: an analysis of 265 cases and a morphological classification. Plast Reconstr Surg. 1992; 89(5): 856–77.