Radial Polydactyly-A Case Report.

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Abstract:-
Polydactyly is a congenital physical anomaly with a wide range of manifestations, ranging from varying degrees of mere splitting to complete duplication of digit. When duplication occurs alone, it is usually unilateral and sporadic. Polydactyly can be classified as ulnar, radial and central Polydactyly. Isolated Polydactyly is autosomal dominant whereas syndromic Polydactyly is autosomal recessive. This is a case of radial polydactyly of right hand - A 38 year old carpenter presented with right shoulder pain and other occupational related somatic problems as his daily work of carpentry involves the usage of thumb mostly. Physical examination revealed type 2 radial polydactyly and it is not moving independently. Normal sensations are seen over the extra digit. Polydactyly is a malformation with a prevalence rate is 5 - 19% per 1000 live births.

I. INTRODUCTION.
Polydactyly is a congenital physical deformity of human having supernumerary fingers or toes1. The extra digit is usually a small piece of soft tissue that can be removed. Occasionally it contains bone without joint. Rarely it may be a complete functioning digit with joint. The extra digit is more common on ulnar side of the hand, less common on the radial side, or very rarely within the middle three digits. These are respectively known as post axial, pre axial and central polydactyly. Post axial and pre axial are to be called as Ulnar and Radial polydactyly respectively. The extra digit present commonly as an abnormal fork in an existing digit or it may rarely originate at the wrist as a normal digit does2. A 3 ½ year old Indian boy had 7 digits on each hand and 10 toes on each foot sets world record for most digits2. The incidence of congenital deformities in newborn is approximately 2%. 10% of these deformities involve upper extremities4,5.The congenital hand committee a sub group of International Federation of societies for Surgery of the hand decided in 1995 to discontinue the words preaxial and postaxial polydactyly6. Recently a gene responsible for pre axial polydactyly type II and type III is located on chromosome 7q 36.

WASSELS classification7 for polydactyly is being widely used in clinical fields.

II.Types and Classification
1. Ulnar Polydactyly
2. Radial Polydactyly
3. Central Polydactyly

ULNAR POLYDACTYLY: The classification of ulnar polydactyly exists of either two or three types. The two-stage classification involves type A and B. In type A there is an extra little finger at the metacarpophalangeal joint, or more proximal including the Carpometacarpal joint. The little finger can be hypoplastic or fully developed. Type B varies from a ribbon to an extra, non-functional little finger part on a pedicle.
RADIAL POLYDACTYLY: Wassel classification is the most widely used classification of radial polydactyly. It is based upon the most proximal level of skeletal duplication:

Type 1: Distal phalanx  
Type 2: Inter phalangeal joint  
Type 3: Proximal phalanx  
Type 4: Metacarpo phalanjeal joint  
Type 5: Metacarpal phalanx  
Type 6: Carpo metacarpal joint  
Type 7: Triphalangeal thumb

The most common type is Wassel 4 (about 50% of such duplications) followed by Wassel 2 (20%) and Wassel 6 (12%).

CENTRAL POLYDACTYLY: The classification of central polydactyly is based on the extent of duplication and involves the following three types: Type I: a central duplication, not attached to the adjacent finger by osseous or ligamentous attachments. It frequently does not consist of bones, joints, cartilage or tendons. Type IIA: a non syndactylosus duplication of a digit or part of a digit with normal components and articulates with a broad or bifid metacarpal or phalanx. Type IIB: a syndactylosus duplication of a digit or part of a digit with normal components and articulates with a broad or bifid metacarpal or phalanx. Type III: a complete digital duplication, which has a well-formed duplicated metacarpal.

III. CASE REPORT

A 38 year old male of Indian origin presented with radial polydactyly of right hand with no other significant complaints. He is having hypoplastic thumb with normal sensations. Being a carpenter his occupational skills are hampered and his fine skilled movements are restricted by the extra digit.

IV. DISCUSSION.

In this case the person is suffering from simple radial polydactyly without any major health complaint. Isolated polydactyly is often autosomal dominant, while syndromic polydactyly is autosomal recessive. A large proportion of polydactyly is isolated. Although they can be associated with an immense amount of anomalies which can include aneuploidic syndrome. Trisomy 13 tends to give postaxial polydactyly. Limb development begins with the deactivation of a group of mesenchymal cells of lateral plate mesoderm. Homebox containing (HOX) genes regulates patterning in vertebrate limb development. The upper limb buds are visible by day 26 or 27 and the lower limb bud appears a day or later. By the end of 6th week mesenchymal tissue in the hand plants condenses to form digital rays. These mesenchymal condensations outline the formation of digits. The interval between digital rays are occupied by loose mesenchymal tissue. Later intervening mesenchymal tissue breaks down and form notches between digital rays. As the breakdown of mesenchymal tissue progresses separate digits are formed by the end of 8th week of intrauterine life. By apoptosis limb development occurs from 24 – 36 day after fertilization. This is the critical period of limb development. Any deviation in this period results in limb anomalies.

Minor
limb anomalies are relatively common and they can be usually corrected surgically. This case of Radial Polydactyly belongs to Type 2 of most widely used WASSELS classification. The most common type of WASSELS classification is Type 4(50%). Our case is the second common type of Radial Polydactyly. Surgical treatment of Polydactyly depends upon type of polydactyly. Because neither of two thumb component is normal, a decision should be taken on combining which element to create the best possible thumb. Instead of amputating the most hypoplastic thumb preservation of skin, nail, collateral ligament and tendons is needed to augment the residual thumb. In this case some hand surgeon recommend BILHAUT-CLOQUET procedure, the technique contains a composite wedge resection of the central bone and tissue. But most commonly used technique for all kinds of Polydactyly is ablation with collateral ligament reconstruction. This is recommended in all cases of thumb duplications with a hypoplastic and less functional thumb. The preferable thumb to preserve is Ulnar thumb. In this procedure by detaching the radial collateral ligament from distal to proximal, a periosteal sleeve is preserved. This way, the radial collateral ligament band of radial digit will functions as the absent radial collateral ligament of the preserved ulnar thumb. On Top Plasty Procedure is indicated in Type 4,5,6 by some surgeons. In our case most preferable technique is ablation with collateral ligament reconstruction because the advantages of this technique is reconstructed joints tend to remain flexible and also, it preserves the nail bed.

V. CONCLUSION

Polydactyly is the most common congenital digital anomaly of the hand and foot. It can occur as an isolated disorder in association with other hand/foot deformity as a part of any syndrome. Surgery is necessary to create a single functioning thumb and is indicated to improve cosmesis. Skin, nail, bone, ligament and musculoskeletal elements must be combined to construct an optimal digit. Therefore, treatment is wholly dependent on the anatomic components present, the degree of syndactyly, and the function of the duplicated finger.

REFERENCES:
2. “One year old Indian boy breaks world record after being born with thirty four fingers and toes”. Daily Mail (LONDON) ; 25 July 2011.