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Case Report

Bilateral Polydactyly with Micropenis- A Case Report

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ABSTRACT

Polydactyly is the presence of more than the usual number of fingers or toes. Ulnar polydactyly is the most common polydactyly of hand. Micropenis is the penile length smaller than 2.5 standard deviations below the mean. Here a case is reported with bilateral hand polydactyly associated with micropenis. There was ulnar polydactyly on the right hand and central polydactyly on left hand. Polydactyly and micropenis usually occur in syndrome like Bardet–Biedl syndrome, Joubert Syndrome etc. Bilateral polydactyly with combination of postaxial and central variety make the case more interesting and rare one.

Keywords: ulnar, central, polydactyly, micropenis.

INTRODUCTION

The term Polydactyly or supernumerary digits refers to the presence of more than the usual number of fingers or toes. Often the extra digit is incompletely formed and lacks normal muscular development. If the hand is affected, the extra digit is most commonly medial (ulnar/postaxial) or lateral (radial/preaxial) rather than central. [1] Polydactyly are usually bilateral. [2]

Lmbr1 gene is required for limb formation and that reciprocal changes in levels of Lmbr1 activity can lead to either increases or decreases in the number of digits in the vertebrate limbs. The loss of digits in mice was observed with reduced Lmbr1 activity in contrast with gain of digits observed in Hx (hemimelic extra toes) mice and human polydactyly patient. [3]

The definition of micropenis was accepted as a penile length smaller than 2.5 standard deviations (SD) below the mean. [4] Micropenis may occur as an independent abnormality by itself or as a clinical finding of many syndromes. [5] It can be deduced that a true micropenis is caused by a hormonal abnormality that occurs after the 12th week of gestation. [6]

Polydactyly and micropenis may occur together as syndrome like Bardet-Biedl syndrome, [7] Coats plus syndrome [8] etc.

Here a case is reported with bilateral hand polydactyly and micropenis.

CASE REPORT

A 3.5 kg, live full-term baby was delivered by normal vaginal delivery to a 30-year-old female who didn't undergo any antenatal ultrasonography. There was no significant family history. Also, history of radiation exposure or intake of drugs was absent. Informed consent was taken from the guardians for examination.

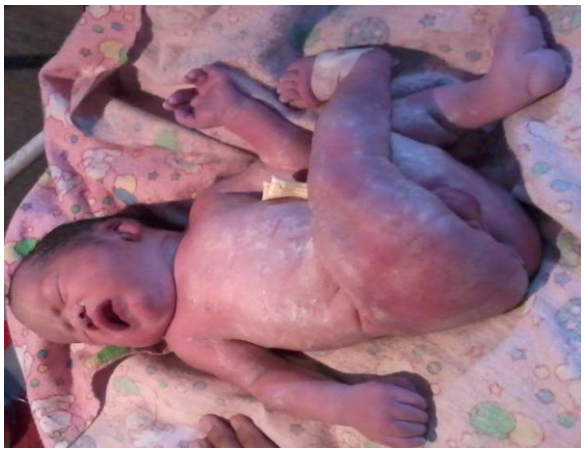

Figure no.1: newborn with bilateral polydactyly. On right hand there is ulnar polydactyly and central polydactyly on left hand.

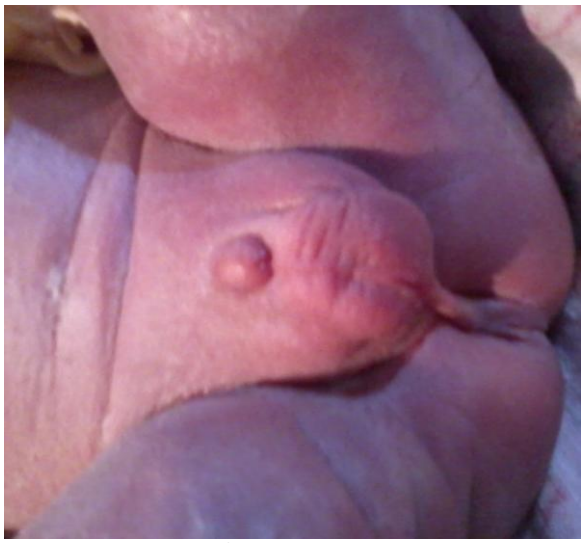

Figure no.2: presence of micropenis with polydactyly.

On external examination, bilateral hand polydactyly was found. There was ulnar or post axial polydactyly on right hand and central polydactyly on the left hand. Central polydactyly was the duplication of the ring finger on left hand (figure no.1, 3, 4).

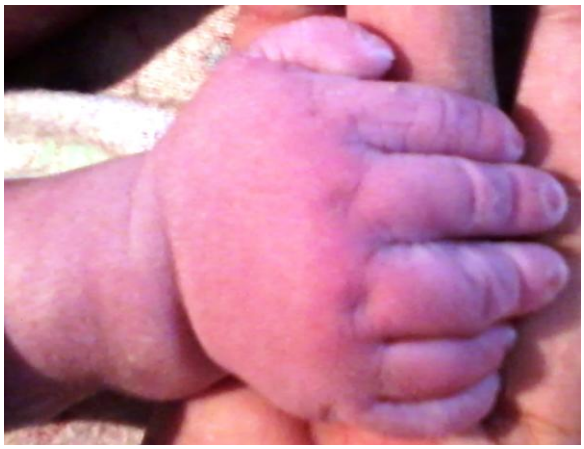

Figure no.3: ulnar polydactyly on right hand.

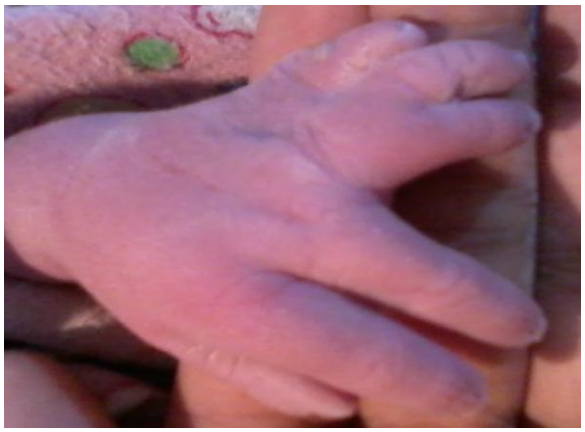

Figure no.4: central polydactyly on left hand with syndactylous duplication of ring finger.

External examination revealed that the ulnar polydactyly on right hand was type A according to two-stage classification of ulnar polydactyly by Temtamy and McKusick [9] (figure no.3). On the other hand, the central polydactyly was type IIB according to classification of central polydactyly based on the extent of duplication [10] (figure no.4). There was absence of polydactyly on foot.

On examination of the external genitalia, micropenis was found (figure

no.2). Further investigations could not be possible as patient and attendants were abandoned and could not be followed up thereafter.

Table no.1: classification of central polydactyly is based on the extent of duplication. [10]

Type of Central Polydactyly		Extent of Duplication
Type I		A central duplication, not attached to the adjacent finger by osseous or ligamentous attachments.
Type II	A	A nonsyndactylous duplication of a digit or part of a digit with normal components and articulates with a broad or bifid metacarpal or phalanx.
	B	A syndactylous 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.

DISCUSSION

Polydactyly can present alone or as part of many syndrome due to genetic disorder. The defects may be due to autosomal recessive or autosomal dominant disease. [11]

The two-stage classification of ulnar polydactyly according to Temtamy and McKusick 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 nubbin to an extra, non-functional little finger part on a pedicle. [9] Thus, in present case, type A ulnar polydactyly is present on the right hand (Figure no.3).

Thus, in the present case, type IIB central polydactyly is present on the left hand (figure no.4).

The syndromes which occur with ulnar polydactyly are: Greig cephalopolysyndactyly syndrome, Meckel syndrome, Ellis–van Creveld syndrome, McKusick–Kaufman syndrome, Down syndrome, Bardet–Biedl syndrome, Smith–Lemli–Opitz syndrome. [12]

The syndromes associated with central polydactyly are: Bardet–Biedl syndrome, [7] Meckel syndrome, [13] Legius syndrome. [14]

Causes of true micropenis can be examined under three headings: hypogonadotropic hypogonadism due to pituitary/hypothalamic insufficiency, hypergonadotropic hypogonadism due to primary testicular insufficiency, and idiopathic. [15- 18]

Polydactyly and micropenis are together found in syndromes like Bardet–Biedl syndrome, [7] Coats plus syndrome, [8] Joubert Syndrome [19] etc. In these syndromes polydactyly were either preaxial or postaxial polydactyly. But in the present case there is combination of ulnar and central polydactyly (figure no.3 and4).

CONCLUSION

The case is a unique one as with micropenis there was bilateral hand polydactyly which was too postaxial in one hand and central in other hand.

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