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Ectrodactyly-Non-Syndromic Type Split Hand-Foot Malformation (SHFM): A Rare Congenital Longitudinal Limb Deficiency Disorder in a Newborn from Eastern Zone of Uttar Pradesh

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Clinical Image

A 4 days old male preterm baby born to 22 yrs. old primigravida mother by non-consanguineous marriage and delivered by caesarean section at 34 weeks by gestation with birth weight of 2.2 kg was admitted to our NICU with congenital absence of middle three fingers in right hand (central digit rays), middle finger in left hand and Syndactyly,

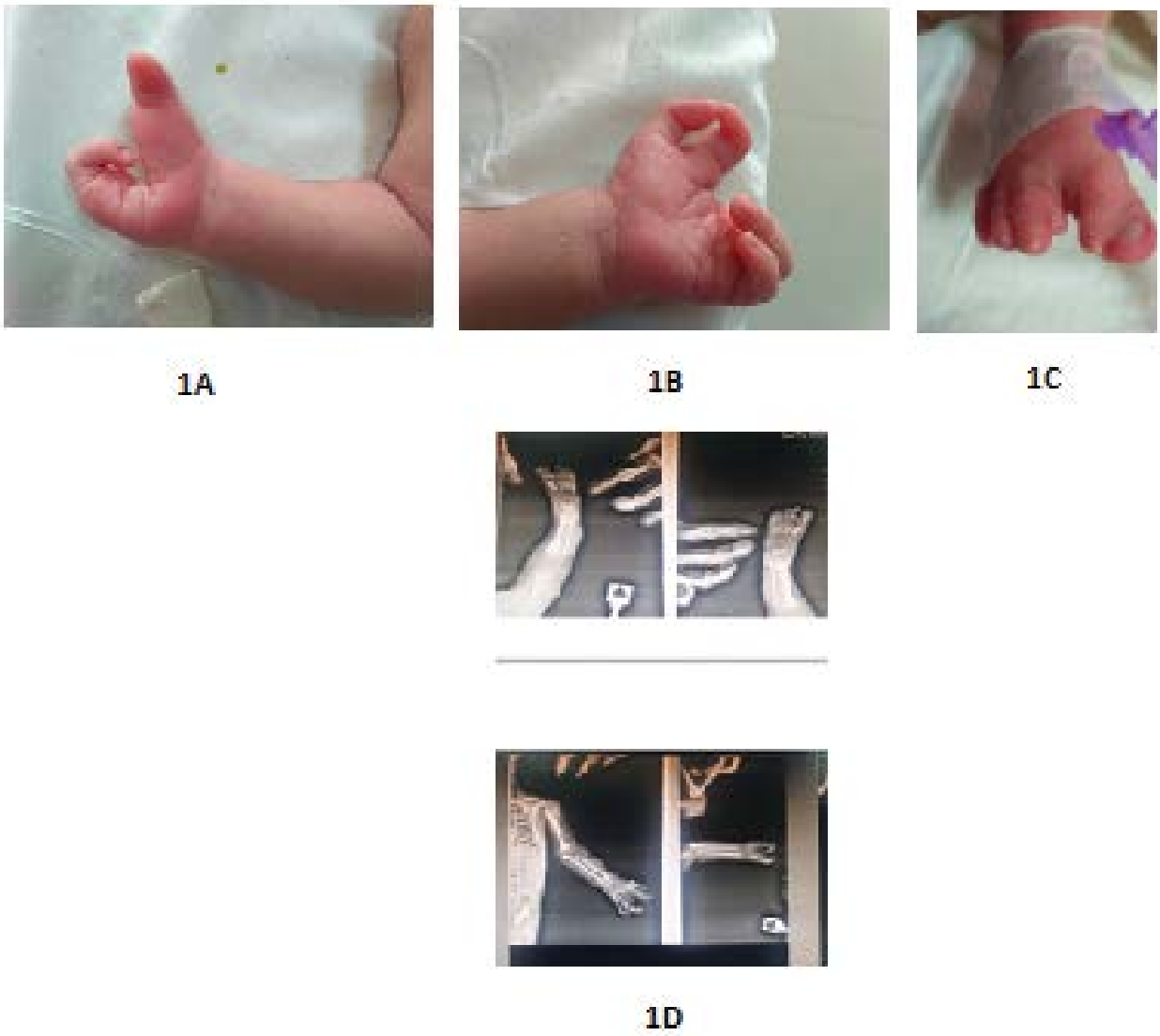

Figure 1: (A-D) Clinical bony malformations.

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Polydactyly and the presence of cleft in right foot with normal left foot. X-ray of Hands and Feet (Figure 1) was in accord to the above mentioned clinical bony malformations. The Baby had no other systemic malformations on thorough clinical and laboratory workup and no similar history in other family members.

This rare congenital SHFM affects the central rays of hands and feet, is a subtype of limb deficiency disorders which occurs due to longitudinal deficiency of central digits [1]. The clinical presentation may vary from only a very minor cutaneous cleft without absence of a finger to a severe form known as monodactyly in which only the little finger remains, and the severity may differ not only between patients but also between the limbs of a single individual as shown in our case [1,2]. In many cases of SHFM, there is syndactyly and hypoplasia of some of the remaining digits. Two expressions of SHFM occur, one with isolated involvement of the limbs, the no syndromic form and the other, the syndromic form with associated anomalies such as tibial aplasia, mental retardation, cleft lip and palate and deafness e.g., Cornelia de Lange syndrome, Acrorenal syndrome etc [1]. The case we report belongs to the non-syndromic type as there is no associated anomaly. The most common mode of inheritance is Autosomal dominant. Autosomal recessive, X-linked and chromosomal duplications and deletions are the rare mode of inheritance [2]. It can be treated surgically or with prosthesis for functional improvement.

References

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