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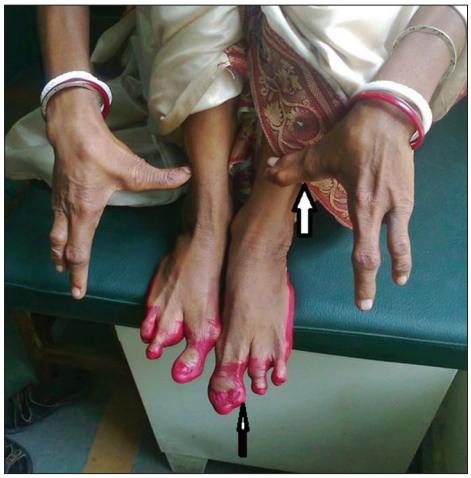

Figure 1: Lobster claw syndrome

Lobster claw syndrome: A rare entity

Sir,

A 50 year old female patient presented with congenital absence of middle finger in both hands with bilateral syndactyly of thumb and index finger [Figure 1; white arrow]. The third toe in left foot and second and third toe in right foot were absent with syndactyly of great and second toe in left foot [Figure 1; black arrow]. Systemic examination and Intelligence Quotient (IQ) were normal. She had no similar family history, had no children, did not have any significant functional deficit and hence did not opt for surgery. This rare congenital (autosomal dominant) defect also known as split hand foot malformation (SHFM) or ectrodactyly has an incidence of 1 in 90,000 live births. The most common associated genetic mutations are in homeobox genes DLX 5 and DLX6 (DLX: Distal-less-like-genes; chromosome 7q). Two expressions of SHFM occur, one with isolated involvement of the limbs, the non-syndromic form and the other, the syndromic form[1], with associated anomalies such as tibial aplasia, mental retardation, orofacial clefting and deafness. The manifestations of cleft hand may vary from only a very minor cutaneous cleft without absence of a finger to a severe form in which only the little finger remains. The case we report belongs to the non-syndromic type as there is no associated anomaly. It can be treated surgically or with prosthesis for functional improvement. Broad indications for surgery include progressive deformity (caused by deforming syndactyly or transverse bones), deficient first web space, deformity of the cleft, syndactyly between digits of unequal length (especially the thumb and index finger) or absent thumb. Many cleft hands, although stigmatizing the child, function well. A family with one or more generations of cleft hands may avoid surgery for their children because they perceive function as excellent and aesthetic improvement as marginal.[1,2] There is a report[3] in the literature on treatment of 4 children with bilateral lobster-claw deformity of the feet. The aim of treatment was to achieve balanced plantigrade feet that are narrow enough to be accommodated in normally shaped shoes; thus, improving both appearance and function. Early physical and occupational therapy can help these patients adapt and learn to write, pick things up and be fully functional. Most surgical procedures may be postponed until the child is between 1 and 2 years of age.[2]

Transvaginal ultrasonography with three-dimensional ultrasonography performed at 14-16 weeks of gestation demonstrates deformities of hands and feet and can help diagnose this condition prenatally.[4]

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REFERENCES

1. Cyriac MJ, Lashpa E. Lobster-claw hand: A manifestation of EEC syndrome. Indian J Dermatol Venereol Leprol 2006;72:54-6.
2. Wolfe SW Deformities of hand and fingers: Central hand

deficiencies. In: Wolfe SW, Hotchkiss RN, Pederson WC, and Kozin SH, editors. Green's Operative Hand Surgery 6th ed., Ch. 40: Churchill Livingstone; Philadelphia, USA 2010.
3. Dwyer FC, Maisels DD. Lobster-claw deformities of the feet. Proc R Soc Med 1976;69:178-81.
4. Koo BS, Baek SJ, Kim MR, Joo WD, Yoo HJ. Prenatally diagnosed ectrodactyly at 16 weeks′ gestation by 2-and 3-dimensional ultrasonography: A case report. Fetal Diagn Ther 2008;24:161-4.

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