SPOT DIAGNOSIS (IMAGE GALLERY)



CONSTRICTING BANDS OVER LIMBS

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A 3 months old boy was admitted for treatment of pneumonia and was incidentally noted to have constricting bands around both legs and right hand. There was either partial or complete auto-amputation of toes of right foot and fingers of right hand. Child was born out of a non-consanguineous marriage to a primigravida mother by a term vaginal delivery at home. The pregnancy was unbooked, unsupervised and no antenatal ultrasound was done. There were no any other associated anomalies.

What is the diagnosis?

A diagnosis of Congenital Constriction Ring syndrome was made based upon clinical findings, with Patterson's type IV deformity. (1) It is a rare sporadic congenital disorder without any genetic or hereditary disposition. The condition is also known as amniotic band sequence, Streeter's dysplasia, congenital constriction bands, and pseudoainhum. (2) The cause for this condition was attributed long back by Streeter to a focal developmental error affecting the subcutaneous germ cell layer. (3) Later on three pathogenesis theories has been suggested for this syndrome: (a) Exogenous due to constriction from fibrous bands formed from partial rupture of amniotic sac leading to array of deletions and deformations, (b) endogenous due to primary vascular compromise and (c) a third one postulates the involvement of intra-uterine trauma in the form of amniocentesis, choriovillous sampling or any fetal surgery. (4-12) Treatment mostly occurs after birth, but with advancement of prenatal radio-diagnosis, fetal surgery in utero has been tried. (7) Management of constriction band syndrome is essentially plastic and reconstructive surgery and is focused on improving function and development while providing a more acceptable aesthetic appearance. (8)

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