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LETTER TO THE EDITOR

Amniotic Band Syndrome

GHRITLAHAREY R K

Sir,

I'm herein presenting a one-day, 2.4 kg old newborn, who presented with a single constriction mark over her right arm due to a congenital amniotic band. She was a full-term baby, born normally at home to a G1 P0, 22 years old mother, on May 15, 2007. Her antenatal history was nothing significant. Antenatal ultrasonography was done in the third month of pregnancy, which was normal. Clinical examination revealed a constriction mark over her right arm, and she also had associated congenital talipes equines varus (CTEV) on the left side ([Table/Fig 1]). Her axillary, brachial, radial, and ulnar arterial pulsations were normal on the affected side. Motor and sensory functions of the affected right upper limb were also normal. Other systemic examinations were within normal limits. Plain skiagram of her right arm showed soft tissue constriction without involvement of the humerus ([Table/Fig 2]). Doppler study of the right upper limb revealed normal blood flow in all the vessels, as well as at the constriction site ([Table/Fig 3]). A small releasing incision was given under local anaesthesia, over the lateral aspect of the arm across the constriction site, although there were no signs of vascular compression in the affected limb. An above knee plaster cast was applied for the CTEV. She is doing well at two months of age, and is awaiting corrective operation.

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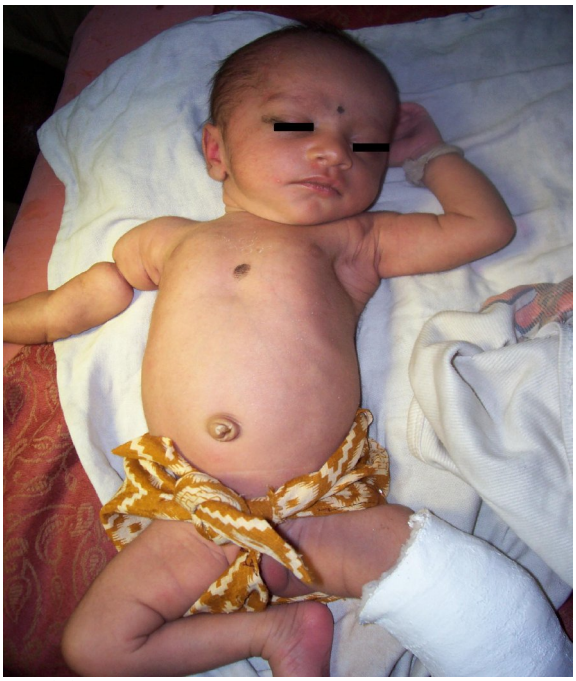
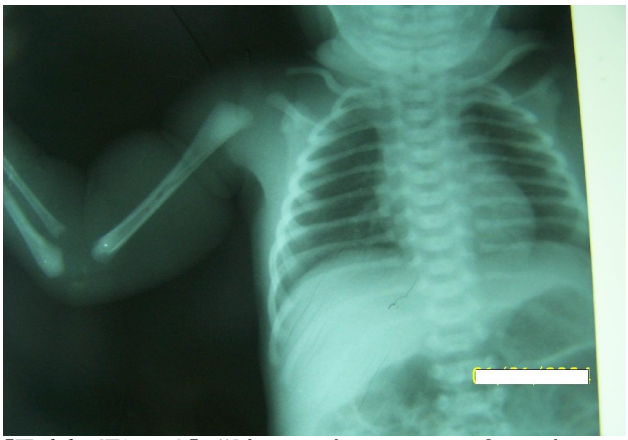
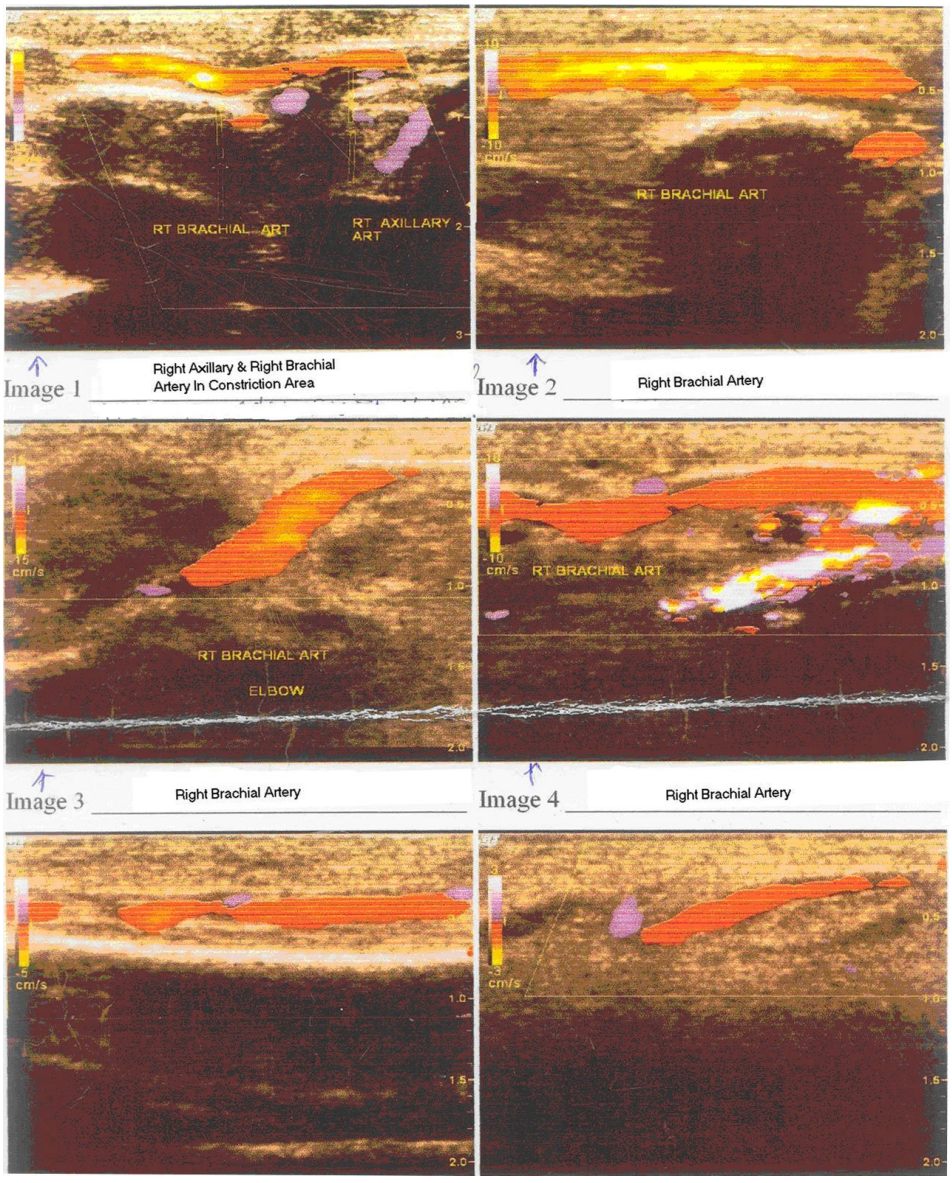


[Table/Fig 1] "Clinical photograph of patient" showing shaft tissue constriction mark at right arm and CTEV left side.



[Table/Fig 2] "Plain skiagram of right arm" showing shaft tissue constriction without involvement of humerus.

Amniotic band syndrome is a set of congenital malformations ranging from minor constriction rings and lymphoedema of the digits, to complex, bizarre, multiple congenital anomalies that are attributed to the amniotic bands that stick, entangle, and disrupt foetal parts [1],[2]. Synonyms of amniotic band syndrome includes; ADAM complex (amniotic deformities, adhesion, mutilation), amniotic band sequence, amniotic disruption complex, annular grooves, congenital amputation, congenital constricting bands, Streeter bands, transverse terminal defects of limb, aberrant tissue bands, amniochorionic mesoblastic fibrous strings, and amniotic bands [2]. The prevalence of amniotic band syndrome among live births is estimated to be around 7.7:10,000, while for spontaneous abortions, it may be as high as 178:10,000. It affects males and females in the same proportion. Most of the cases are sporadic, with no recurrence in siblings or children of affected adults [1],[2]. The exact aetiology is not known, although some authors have suggested a teratogenic effect in some cases [2],[3].



[Table/Fig 3] "Doppler study of right upper limb" showing normal blood flow in all the vessels including at constriction site of right upper limb.[All the photographs displayed in this article were obtained after informed consent by the parents of the child.]

Amniotic band syndrome is often difficult to detect before birth, although the earliest amniotic band that was detected, was at 12 weeks of gestation, by vaginal ultrasound. Bands

may be difficult to detect by ultrasound, and are more often diagnosed by the effects they have on foetal anatomy. Doppler studies of the constricted limb could be of useful predictive value of in utero amputation, and therefore could be helpful to determine when in utero treatment should be considered [3]. Congenital amniotic band may result in; constriction rings around the digits, arms and legs, swelling of the extremities distal to the point of constriction, amputation of digits, arms and legs, asymmetric facial, cephalocele, anencephaly, multiple joint contractures, pterygium, clubfeet, clubhands and pseudosyndactyly, microphthalmia, uveal coloboma, corneal metaplasia and unilateral chorioretinal lacunae. Amniotic band syndrome needs to be differentiated from amniotic fold, body-stalk anomaly, and short umbilical cord syndrome.

Management of anomaly depends upon the extent of the anomalies. Termination of pregnancy is usually proposed at the time of the diagnosis of severe craniofacial and visceral abnormalities, whereas minor limb defects can be repaired with postnatal surgery [3]. Surgical correction of the deformity ranges from simple to complex surgical procedures, depending on the extent of the deformity. Recently, for postnatal surgical correction of constriction rings, a new technique (Mutaf procedure) has been reported, which advices filling the circular groove caused by the constriction ring with dermofat flaps. This eliminates the soft tissue deficiency, and provides a normal extremity contour. Moreover, since rectangular-plasty allows replacing the major limbs of the incisional scars within the relaxed skin tension lines, it provides a better scar in comparison with old Z-plasty techniques [4]. Foetoscopic release of amniotic bands in extremity amniotic bands offers the potential to prevent limb amputation [5],[6].

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