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Case Report

Streeter Dysplasia in Association with Cleft Palate: A Case Report

Paediatrics Section

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ABSTRACT

Streeter dysplasia, a synonym of Amniotic Band Syndrome (ABS) is a wide spectrum of complex, controversial congenital anomalies. The disease varies in its severity which can range from a simple constriction ring to more severe and lethal forms like limb body wall complex or acalvaria .The usual manifestations include constriction rings around the digits, arms and legs, amputation of digits and limbs, club feet, club hands, etc. Here, we report a case of amniotic band syndrome with multiple digit anomalies and a cleft palate.

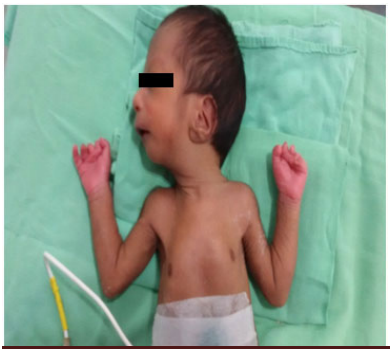
Keywords: Amputation, Constriction ring, Hypoplasia, Malformed limbs, Syndactyly

CASE REPORT

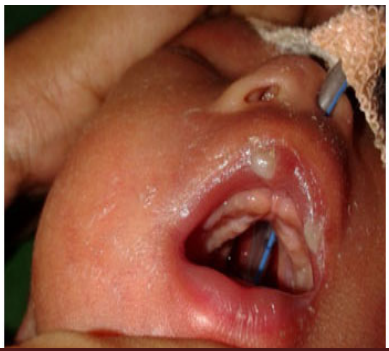
A 2 day old male newborn was brought to the Special Newborn Care Unit (SNCU) by his parents with chief complaints of abnormally formed upper and lower limbs [Table/Fig-1]. The baby who was the first issue of a non-consanguineous marriage was born by preterm vaginal delivery at 30 weeks of gestation with a history of premature rupture of membranes. There was no history of trauma or drugs taken by the mother during pregnancy. Pregnancy was uneventful. Her dating scan and anomaly scans were unremarkable. There was no history of maternal systemic diseases like diabetes, hypertension or obstructed labour. At the time of birth the child cried normally and there was no cyanosis. On examination, the baby was active weighing 1.3kg. General examination revealed stable vitals (HR-120/min, CRT=2 sec, RR-46/min, Temp-99F) and a cleft palate [Table/Fig-2]. Examination of the left upper extremity revealed that the thumb, index and middle fingers terminated at the interphalangeal joints with syndactyly of the other digits [Table/Fig-3]. Examination of the right upper extremity revealed amputation of the index and middle fingers at the proximal interphalangeal joints [Table/Fig-4]. The left lower limb revealed amputation of the second toe at the proximal interphalangeal joints with moderate development of the other digits and also the right lower limb [Table/Fig-5]. X-rays of all the limbs were advised for further evaluation which revealed absence of distal phalanges of the malformed bones with soft tissue fusion. Based on these clinical and X-ray findings a diagnosis of amniotic band syndrome (ABS) was made. In view of the very low birth weight and feeding difficulties on account of the cleft palate the child was admitted, feeding was stabilised and was discharged on day 21 of life with a weight of 1.5kg. The parents were counselled regarding the feeding techniques, micronutrient and vitamin supplementation and was advised regular follow up for the correction of the limb deformities.

DISCUSSION

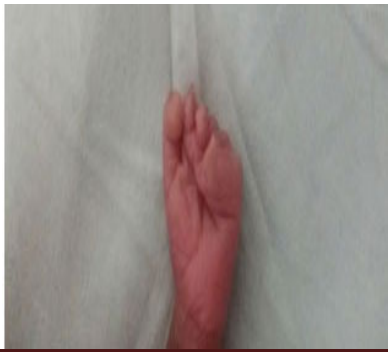
During intrauterine life, the fetal parts can be entangled by amniotic bands which in turn gives rise to a set of



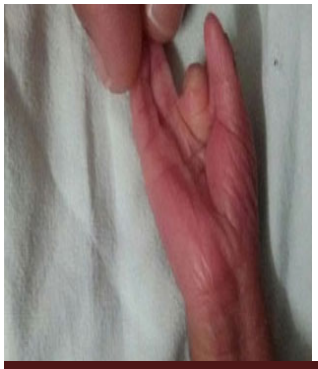
[Table/Fig-1]: Newborn with malformed limbs

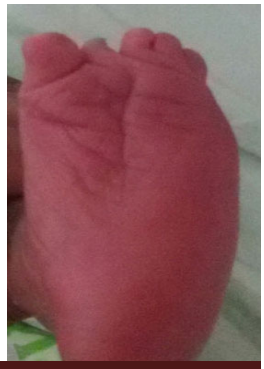


[Table/Fig-2]: Cleft palate



[Table/Fig-3]: Left upper limb





[Table/Fig-4]: Right upper limb **[Table/Fig-5]:** Left foot malformation

malformations comprising the amniotic band syndrome (ABS). The malformations may be less severe such as constriction rings or it could result in multiple complex anomalies which could even be fatal [1]. The entity has been addressed by multiple synonyms in literature such as congenital constriction ring, ADAM (amniotic, deformity, adherences, mutilations) complex, amniotic band rupture complex, amniotic band sequence and Streeter dysplasia. As early as 300 BC, noted pioneers like Aristotle and Hippocrates have discussed this condition. The explanation of constrictions and amputations attributable to the extrinsic pressure caused by rupture of amniotic membranes was suggested by Hippocrates. Notable scientists like Montgomery in 1832 and Simpson in 1836 first documented a case series describing the malformations in ABS. Although a novel germ-plasm theory was highlighted by George Streeter in 1930, it was overlooked by the Torpin's extrinsic theory in 1965. [2,3].

The exact mechanism of the occurrence of ABS is still a mystery; however there are two theories postulated to explain the pathogenesis. The "extrinsic model" theory by Torpin and Faulkner in 1966 is most accepted according to which the malformations arise due to amniotic rupture in early pregnancy with the formation of amniotic bands which in turn entrap the growing parts of the foetus. The less accepted Streeter's "Intrinsic model" states that the amputations are attributable to disturbances of the developing germinal disc of the embryo. [4-6]

In our case, the clinical presentation of digits and toes amputation with its associated cleft palate could be a result of local compression or adhesion. Therefore, the present case report may further support the concept of extrinsic theory in the aetiology of ABS.

The aetiology is unknown. There have been reports associating ABS with maternal trauma, oophorectomy during pregnancy, intra uterine contraceptive device and amniocentesis. Most cases of ABS have presented as sporadic events. However familial recurrence has been reported in many studies. Cases have been reported in families with connective tissue disorders like Ehler Danlos syndrome [7].

ABS has extensively polymorphic clinical findings, because the type of deformities depends on the time of amniotic rupture during pregnancy and the part of the foetal body which is entangled in amniotic bands. Early amniotic rupture, during first 45 days, leads to the most severe cranio-facial and visceral malformations [8]. Most often extremities, especially upper extremities (hands affected in 90%).However the bands can compress any organ part according to which the manifestations vary: Digits of extremities (amputations, syndactyly, hypoplasia, peripheral nerve palsies), head (facial clefts, anophthalmos, microphthalmos, anence-phaly, encephalocele, acalcaria), abdominal wall (gastroschisis) [9]. Hence in view of the heterogeneity we can seldom find two similar presentations of ABS which depicts its uniqueness.

Prenatal diagnosis is by antenatal ultrasound which can show the amniotic bands or the malformations, however mild forms are likely to be missed and hence post natal diagnosis depends mainly on a prompt physical examination.

Surgery remains the mainstay of treatment with a unique approach to every case. Multidisciplinary approach with coordinated effort of plastic surgeon, orthopaedic surgeon and others according to the affected part is unavoidable. Long term occupational and physiotherapy is often required. [9]

CONCLUSION

Hence, the entity of ABS should never be overlooked in any newborn presenting with malformations of the extremities, skull or body wall. If the diagnosis is missed in the antenatal scans, it can still be diagnosed after birth. Proper physical examination with minimal investigations like X-rays paves way for timely post natal diagnosis and ample treatment at an early stage.

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