

AN UNCOMMON CASE OF AMNIOTIC BAND SYNDROME

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ABSTRACT:

Amniotic band syndrome is a set of congenital malformations ranging from minor constriction rings to complex multiple congenital anomalies that are attributed to amniotic bands that entangle and disrupt fetal parts. Usual manifestations are constriction rings around the digits, arms and legs, amputation of digits and limbs, club feet, club hands, etc. Here we present a case of amniotic band syndrome with symmetric intrauterine growth retardation

(IUGR) with oligohydramnios. The baby had a small cleft adjacent to left big toe without any other obvious deformities. This case of amniotic band syndrome is reported to highlight the importance of how heightened clinical suspicion, appropriate obstetric intervention and neonatal care can improve the outcome of child wellbeing.

Key words : amniotic band syndrome, constriction rings, oligohydramnios.

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INTRODUCTION:

Amniotic band syndrome (ABS) has been studied since the time of Hippocrates and Aristotle.^[1] As early as 1968 Richard Torpin worked on the pathogenesis of amniotic band syndrome. The prevalence of amniotic band syndrome for live births is 7.7:10,000.^[2] Among pregnant mothers with amniotic band syndrome, spontaneous abortions is a common outcome and it can be as high as 178:10,000.^[3] Here we report a case of amniotic band syndrome with symmetric IUGR with oligohydramnios. The aim of this paper is to describe how a high clinical suspicion and appropriate obstetric management can help in improving fetal wellbeing in the case of amniotic band syndrome.

CASE REPORT

A 23 year old lady with 37 weeks of gestation was referred from a peripheral center with a diagnosis of amniotic band syndrome for further management. She had one previous history of abortion following which she conceived spontaneously and was on regular folic acid supplementation. She reported quickening at the fifth month and also received two doses of tetanus toxoid. Her dating scan and anomaly scan were normal. Ultrasonogram examination done at 36 weeks revealed the presence of amniotic bands with symmetric intrauterine growth retardation (IUGR) and oligohydramnios (Fig.1).

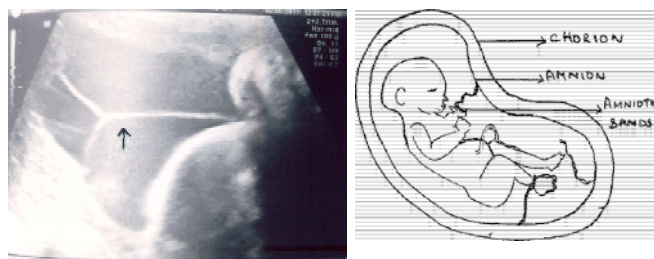


Fig.1 : Ultrasonogram image showing presence of amniotic bands; adjacent picture gives a diagrammatic picture of amniotic band

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At 36 weeks, two doses of steroids, injection Betamethasone 12mg were given 24 hours apart, (prophylactically in view of IUGR).

She had no significant past medical or surgical history. Her menstrual cycles were regular. Her first pregnancy was a spontaneous abortion at 8 weeks for which dilatation and curettage was done.

On examination she had no pallor, icterus, cyanosis or clubbing.

Her pulse rate was normal and her blood pressure was 110/70 mmHg. All the systemic examination was normal. Per abdomen: uterus corresponded to 32 weeks, which was relaxed with cephalic presentation. Clinically the liquor was decreased. Fetal heart rate was 140 beats per min. Her laboratory parameters were all within the normal range (Table 1). Her ultrasound scan reports revealed amniotic bands after which a repeat ultrasound was performed that confirmed the presence of amniotic bands, oligohydramnios and symmetric IUGR (Table 2). Her urine routine, serum electrolytes and liver function tests were normal.

Table 1. Laboratory parameters of the patient with Amniotic Band Syndrome.

INVESTIGATIONS	REPORTS
Haemoglobin	11.3g/dl
PCV	41.1
FBS	91mg/dl
Platelets	1.61 lakhs/cu mm
Blood Group	'A' Positive
HIV, HBSAG	Negative

All laboratory parameters were in the normal range.

Intraoperatively the placenta showed marginal insertion of the umbilical cord with multiple areas of calcification. Amniotic bands were seen extending from each avascular area.

She delivered a girl baby weighing 1.43kg with an Apgar score of 8/10, 9/10. Baby was immediately assessed by paediatrician. Baby had a small cleft adjacent to left big toe (Fig. 3). No other obvious congenital deformities was



Fig.3 : Presence of Cleft near left big toe.

TABLE 2: OBSTETRIC ULTRASOUND FINDINGS OF THE PATIENT

	29.04.2011	02.05.2011
Gestational Age (Dates)	36weeks	36 weeks + 3 days
Gestational Age (Scan) [lags by 4 weeks compared to the gestational age according to last menstrual period]	31-32 weeks	31-32 weeks
Presentation	Cephalic	Cephalic
Placenta	Posterior	Posterior
Amniotic fluid index	8.9 cm	8.5 cm
Estimated fetal weight	1523 +/-152.3 g	1674 +/-251.17g
	Symmetric IUGR with few amniotic bands with oligohydramnios	HC falls at 2 nd percentile and AC falls less than first percentile for 36 – 37 weeks suggestive of symmetric IUGR. Multiple amniotic bands seen applied closely to fetal body but not attached to it. Limited assessment of fetal anatomy. No obvious defect in parts assessed. Normal Doppler study.

noted. Post operatively the baby was monitored in neonatal intensive care unit in view of low birth weight.

Baby was followed up for 2 weeks after delivery. The baby was on direct breast feed. For further management of small cleft seen adjacent to left big toe, the patient was referred to the Department of Orthopaedics and General Surgery. No surgical intervention was suggested.

DISCUSSION

Amniotic band syndrome is a set of congenital malformations ranging from minor constriction rings to complex multiple congenital anomalies that are attributed to amniotic bands that entangle and disrupt fetal parts.

ABS occurs when the inner membrane of the amniotic sac tears and wraps around the developing baby and causes problems with limbs, clefts in the face and band marks in different areas of the body.^[4]

The etiology is unknown. There have been reports associating amniotic band syndrome with maternal trauma, oophorectomy during pregnancy^[5], intra uterine contraceptive device, and amniocentesis.^[6] There are case reports in families with connective tissue disorders like Ehler Danlos syndrome.^[7]

The clinical features^[8] range from minor constriction rings around the digits, arms and legs. Swelling of the extremities distal to the point of constriction (congenital lymphedema). Severe complications like amputation of digits, arms and legs (congenital amputation) can occur.^[8] Congenital deformities like clubfoot, clubhands, cleft lip, cleft palate and hemangioma can occur.

Amniotic band theory states that ABS occurs due to a partial rupture of amniotic sac. This rupture involves only the amnion; the chorion remains intact.^[9] The bands can constrict fetal parts reducing blood circulation and hence causes congenital abnormalities.

Vascular disruption theory suggests an “intrinsic” defect of blood circulation because the constricting mechanism of the amniotic band theory does not explain the high incidence of cleft palate and other cleft defects occurring in ABS.^[10]

Amniotic band syndrome can be diagnosed prenatally by ultrasound which can show amniotic bands, congenital malformations, oligohydramnios.^[11] The most important diagnostic criteria are visible amniotic bands, constrictions rings and irregular amputations of toes or fingers. Three dimensional and four dimensional ultrasound and MRI contribute to more sensitive prenatal diagnostics of amniotic band syndrome.

TREATMENT

The accepted modality of treatment for ABS in utero is by foetoscopic laser surgery before the bands can compress fetal parts.^[12]

Plastic and reconstructive surgery to treat any resulting deformity has been performed after birth. Physical and occupational therapy for long term rehabilitation must be considered.

RECURRENCE

Amniotic Band Syndrome is often sporadic with no recurrence in subsequent pregnancies. However there are some reports of ABS occurring among families with collagen disorders like Ehler-Danlos syndrome.^[13]

CONCLUSION

This case report illustrates how heightened clinical suspicion can help in pre-operative diagnosis of amniotic band syndrome.

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