International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Rita D et al. Int J Reprod Contracept Obstet Gynecol. 2023 Jun;12(6):1944-1946
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20231591

Case Report

A rare case report of amniotic band syndrome associated with gastroschisis and limb body wall complex

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Received: 22 April 2023
Accepted: 11 May 2023

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ABSTRACT

The amniotic band syndrome can cause a wide range of anomalies and the most common associated anomalies include amputations, constriction bands, encephalocele, acrania, syndactyly, craniofacial defects, club feet and cleft lip. Vertebral and abdominal wall defects such as gastroschisis and scoliosis are extremely rare. A 19-year-old female, G1P0 (primi) presented to our outpatient department for her first antenatal visit at 14 weeks + 6 days' period of gestation according to her LMP. Ultrasonography (USG) features suggestive of limb body wall complex/ amniotic band syndrome, undergone MTP after counselling. In amniotic band syndrome there is no known inheritance pattern. The prognosis in case of amniotic band syndrome is dependent on the specific anomalies present. Severe and complex malformations as seen in this case are incompatible with extra uterine life and are an indication for termination of pregnancy. This condition is diagnosable prenatally. It can also lead to lethal deformation of vital internal organs in rare cases. Because limb body wall complex is incompatible with life, early diagnosis is crucial for parental counseling.

Keywords: Amniotic band syndrome, Gastroschisis, Limb body wall complex, Parental counselling

INTRODUCTION

Amniotic band syndrome (ABS), also known as amniotic band disruption complex or constriction band syndrome or amniotic deformity, adhesions, mutilations (ADAM) complex. The syndrome can cause a wide range of anomalies and the most common associated anomalies include amputations, constriction bands, encephalocele, acrania, syndactyly, craniofacial defects, club feet and cleft lip.[1]

Vertebral and abdominal wall defects such as gastroschisis and scoliosis are extremely rare. The estimated incidence is reported to be difficult to measure, with a wide range of one case in 1200 births to one case in 15,000 births.[2]

It is commonly sporadic with a low risk of recurrence, so the differential diagnosis of this syndrome from other structural and congenital anomalies is important.[3]

CASE REPORT

This rare case report was presented in 31st KSOGA state level conference and won first prize among hundred poster presentation.

A 19-year-old female, primi gravida presented to our OPD with history of 4 months of amenorrhea with no complaints for her first antenatal checkup. Her previous medical and surgical history were uneventful. No history of any trauma or infections or drug intake. Urinary pregnancy test was positive, O/E –pallor ++, pulse- 96 bpm, blood pressure (BP)-114/70 mmHg, CVS/RS- NAD, on per abdomen-uterus was 14-week size, and relaxed.

Investigations include haemoglobin (Hb) - 7.7 gm%, total count- 6350, platelet- 3.5 lakhs, (MCV, MCH, MCHC) - decreased, and peripheral smear shows microcytic hypo chromic anemia.

On USG, single live intrauterine fetus of 14 weeks + 2 days of gestational age with defective anterior abdominal wall and abdominal contents floating in amniotic cavity s/o gastroschisis. Kypho-scoliosis of thoraco-lumber spine, shortened left tibia and fibula, small curved right femur with absent tibia and fibula, left foot adjacent to femur, oligohydromnios, s/o limb body wall complex with amniotic band syndrome.

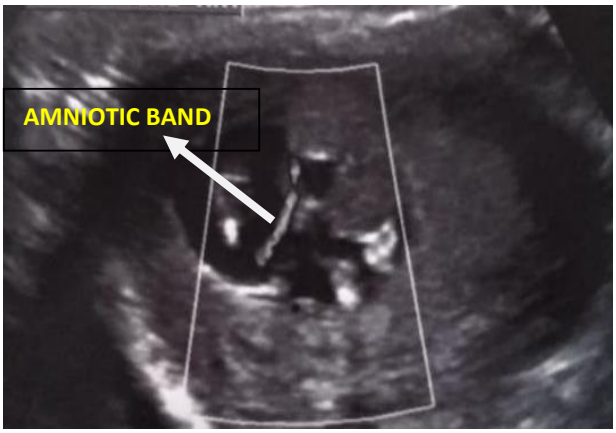

Figure 1: USG showing amniotic band attaching to body wall of fetus and gastrochisis suggestive of amniotic band syndrome.

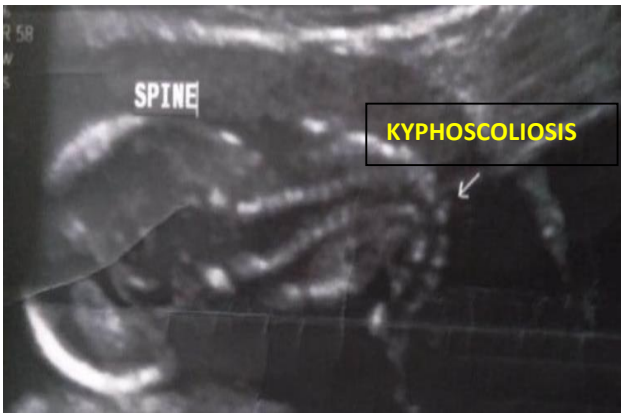

Figure 2: USG showing kyphoscoliosis in fetus.

Management

Patient was counseled about fetal condition and its lethal prognosis and patient expressed willingness for termination of pregnancy. Consent was obtained.

After correcting anemia by transfusing 2 pint PRBC, tablet mifepristone 200 mg given per orally, after 24 hours, Foleys induction done with 16 F catheter and cervix is smeared with tablet misoprostol 50 mcg. After 16 hours, Foleys catheter was expelled.

After 4 hours, single female dead fetus with multiple anomalies as explained in the scan with fetal weight approximately 400gm and placenta weight 500 gm was expelled.

Karyotyping of fetal specimen denied by patient side due to lack of affordability.

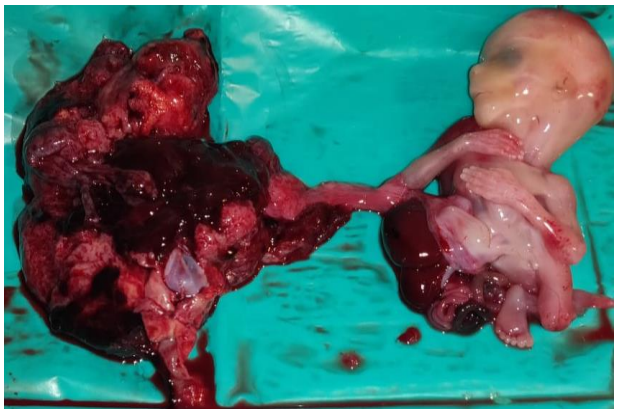

Figure 3: Expelled fetus with placenta.

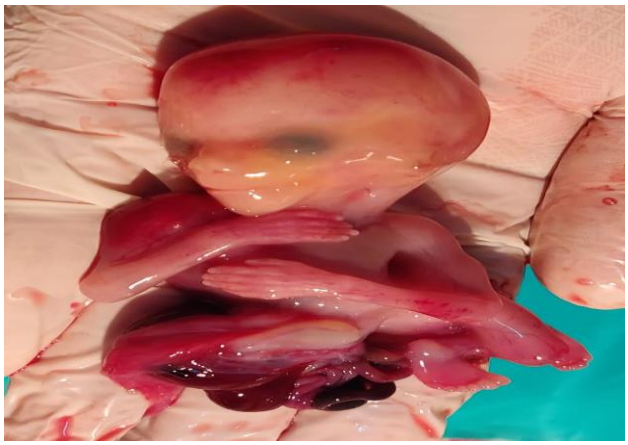

Figure 4: Expelled fetus with gastroschisis, limb body wall and spine defects.

DISCUSSION

Amniotic band syndrome shows no known inheritance pattern. Almost all cases are sporadic, but few examples of familial Amniotic band syndrome have been reported primarily with monozygotic twin gestation.[1]

Etiology is multifactorial with multiple proposed mechanisms; the most widely accepted theories include amniotic band attachment, vascular disruption, and rare genetic mutation.

There are four main categories of anatomical defects – constriction rings, limb defects, spine/neural defects, and craniofacial defects.[4] Elevated α-fetoprotein levels and sonographic evidence of scoliosis, limb defects, Abdominal wall defects, and abnormal placentation help in the early identification of limb body wall complex.[2]

In utero surgery can free the baby's limbs from amniotic bands that could prevent them from growing or that could threaten to amputate the limbs. Performing a minimally

invasive fetoscopy allows the fetal surgeon to insert a fetoscope that is able to cut the bands around the baby's limbs, fingers or toes and allow the blood flow to resume properly.[5]

But, according to Pant et al, the prognosis in case of amniotic band syndrome is dependent on the specific anomalies present. Severe and complex malformations as seen in this case are incompatible with extra uterine life and are an indication for termination of pregnancy.[3]

CONCLUSION

Amniotic band syndrome is diagnosable prenatally. It can also lead to lethal deformation of vital internal organs in rare cases. Because limb body wall complex is incompatible with life, early diagnosis is crucial for parental counseling.

Management begins with early fetal diagnosis and early collaboration between obstetrics, neonatal- perinatal specialist, and maternal-fetal medicine specialist. This may help achieve the best outcome for individual patients. Lastly, parents should receive proper counseling from the clinical inter professional team that most amniotic band syndrome cases are sporadic with no known re-occurrence risk unless in cases of familial amniotic band syndrome.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Rita D, Pushpalatha VS. A rare case report of amniotic band syndrome associated with gastroschisis and limb body wall complex. Int J Reprod Contracept Obstet Gynecol 2023;12:1944-6.