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Prenatal diagnosis of amniotic band syndrome

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Abstract

Amniotic band can cause a broad spectrum of anomalies ranging from simple band constrictions to major craniofacial and visceral defects. It can cause significant neonatal morbidity. Accurate diagnosis will help in the management of the present pregnancy and in counseling with regard to future pregnancies. Here we report three cases of amniotic band syndrome detected in the prenatal period.

Key words: Amniotic band; amniotic band syndrome; craniofacial anomaly; limb anomaly

Introduction

Amniotic band syndrome (ABS) is an underappreciated cause of fetal and neonatal mortality and morbidity. The synonyms for ABS include, amniotic band disruption complex, constriction band syndrome, and amniotic deformity, adhesions, mutilations (ADAM) complex. It is a nonrecurrent cause for fetal malformations involving the limbs, craniofacial region, and trunk.[1] Prenatal diagnosis is important for counseling and management of the present pregnancy. Deformities in ABS can mimic genetic syndromes, hence, an accurate diagnosis is important to counsel the couple regarding the low risk of recurrence in future pregnancies. Here, we report three cases of ABS detected *in utero.*

Case Reports

Case 1

A27-year-old primigravida was referred to us at 23 weeks of gestation, with a history of leaking per vaginum for two weeks. There was no history of fever or bleeding per vaginum in the first trimester.

Ultrasound done at our center showed a single live fetus, with parameters corresponding to 21 weeks. Imaging was difficult because of severe oligoamnios. Single vertical pocket measured 7 mm. The fetus was detected to have a lower limb anomaly.The left lower leg was short [Figure 1A] with the tibia and fibula measuring16mm compared to 31 mm on the right side. The left foot was not seen. The findings were suggestive of amputation of the left leg. Movement of the left lower limb confirmed amputation and absence of the foot [Video 1]. On the right side, a constriction was seen in the calf [Figure 1B]. The working diagnosis was amniotic band syndrome.

Labor was induced in view of maternal sepsis and she delivered a dead female fetus weighing 460g. The left lower limb showed amputation at the mid calf. There was an area of constriction on the right lower limb and below the constriction, the limb was edematous. The upper limbs were normal.The postnatal picture confirmed our diagnosis of amniotic band syndrome [Figure 1C]. The fetal karyotype was normal.

Case 2

A 25-year-old primigravida came for a first trimester ultrasound at 12 weeks of gestation. The first trimester was uneventful. USG done showed a fetus with parameters corresponding to 12-13 weeks of gestation.A membrane was

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seen attached to the lower limb, restricting the movement of the limb [Figure 2A, Video 2]. An ultrasound done at 19 weeks showed limb anomaly. The right foot was acutely dorsiflexed with oligodactyly- only one toe could be seen and the left foot had inversion [Figure 2B]. There was syndactyly in the left hand and oligodactyly in the right.A band was seen connecting the hands [Figure 2C]. The features were suggestive of ABS. The couple when counseled regarding the findings opted for termination in view of the morbidity. The pregnancy was terminated and the findings were confirmed. The fetal karyotype was normal.

Case 3

A 28-year-old primigravida with a fourth degree consanguineous marriage, came to us for an anomaly scan at 19 weeks. The fetus was detected to have multiple anomalies - bilateral cleft lip and palate [Figure 3A], distorted calvarium [Figure 3B], severe frontal bossing with discontinuity of the frontal bone [Figure 3C], and a band was seen near the fetal head moving along with the fetus [Figure 3D].

The couple, when counseled, opted for termination of the pregnancy. The fetus was found to have bilateral cleft lip palate, the right eyelids were missing. In the scalp there was a localized area of aplasia cutis and a membrane was attached to that area [Figure 3E]. The fetal karyotype was normal.

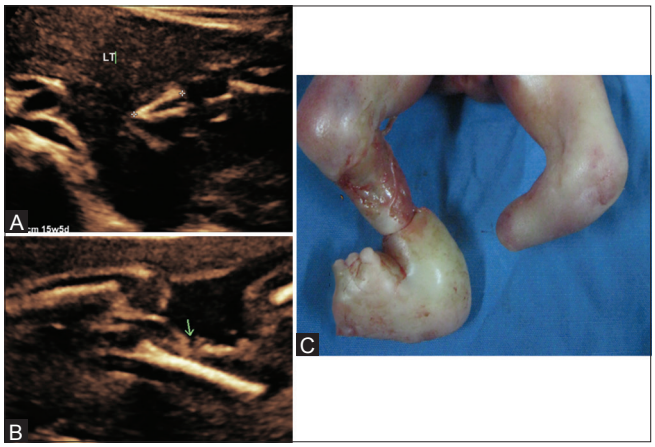

Figure 1 (A-C): (A) Short lower limb (B) Constriction in the lower limb (arrow) (C)Postnatal picture showing amputation of the left leg and constriction of the right lower limb

Discussion

Amniotic band syndrome can cause malformations that range from mild deformities to severe anomalies that are incompatible with life. The birth prevalence rate of ABS has been reported to be 0.89 per 10, 000 births.[2]

The most common defects associated with ABS are limb defects such as focal constrictions, amputations, psuedosyndactyly, and brachydactyly.An amputation with the distal bone protruding beyond the soft tissue at the site of the amputation is diagnostic of ABS. Focal constrictions caused by the amniotic band may occur with distal lymphoedema.Craniofacial anomalies caused by ABS include acalvaria, acrania, asymmetric encephalocoele, asymmetric microphthalmia, severe nasal deformity, and nonembryological facial clefting. Nonembryological clefts, asymmetric encephaloceles, and gastropleuralschisis can be suggestive of ABS, even when the bands are not apparent.[1,3-6]

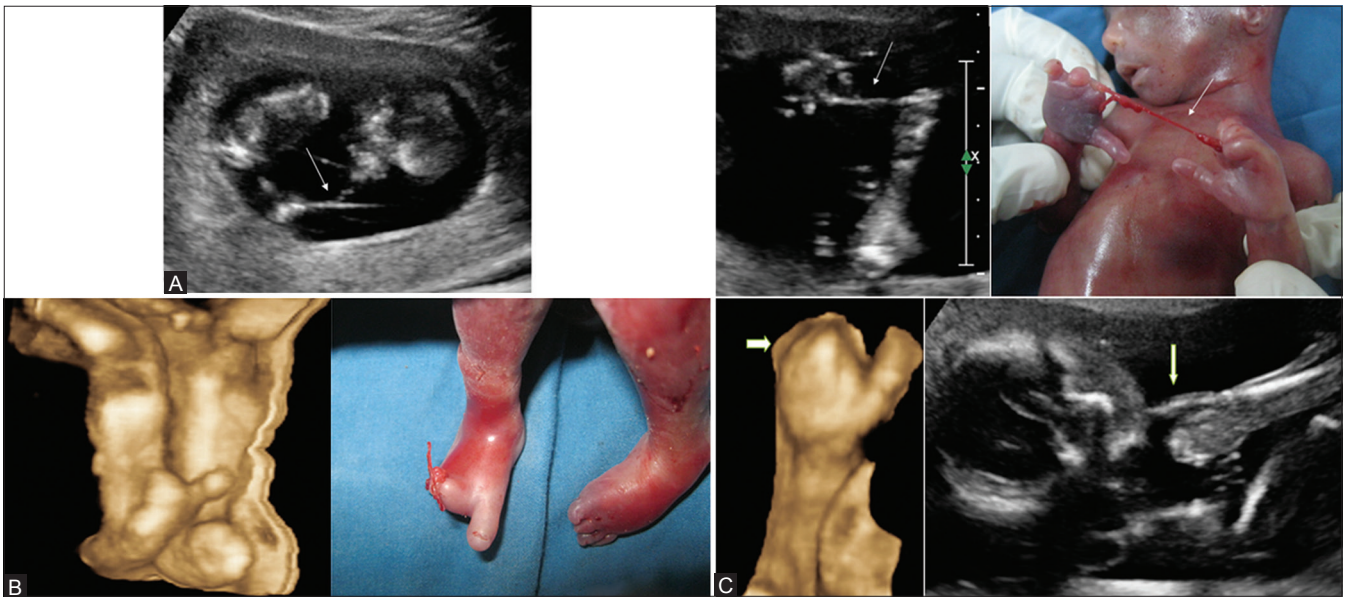

Figure 2 (A-C): (A) Amniotic band seen attached to the lower limb (arrow) (B) Ultrasound image and post-abortal picture of the lower limbs showing oligodactyly and inversion of foot (C) Ultrasound image and post-abortal picture of the fetus, with the amniotic band bridging the hands(arrow), with brachydactyly and syndactyly (broad arrow)

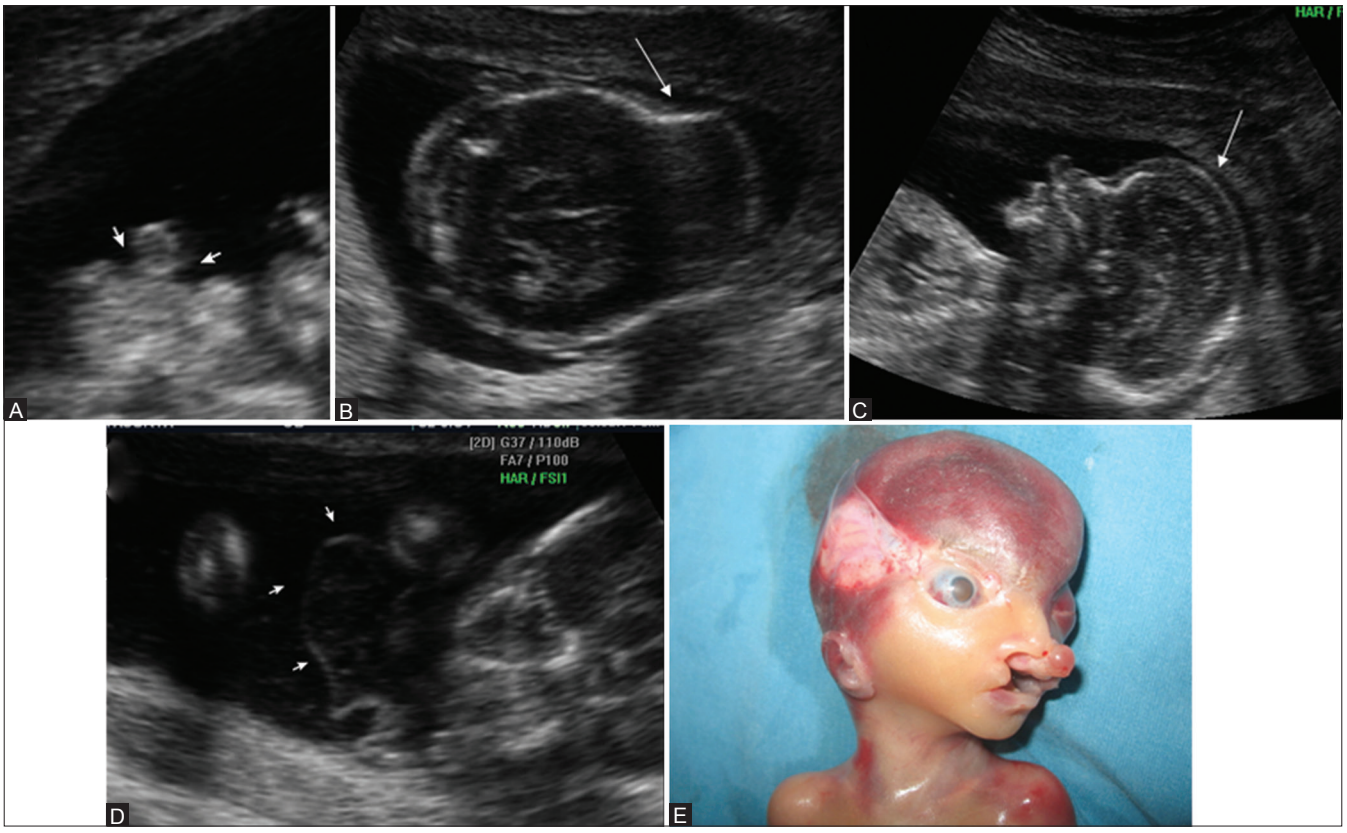

Figure 3 (A-E): (A) Bilateral cleft lip palate (arrows) (B) Image showing distortion of calvarium (arrow) (C) Image showing frontal bossing with discontinuity of bone (arrow) (D) Membrane seen by the side of the head (E) Post abortal picture

The most accepted etiology for ABS is the one proposed by Tropin.[7] He proposed that disruption of the amnion initiates a cascade of secondary events that cause ABS- the amniotic fluid and the fetus exit the amniotic cavity and come to lie within the chorion, transient oligoamnios occurs and from the chorionic side of the amnion multiple mesodermic strands emanate that entangle the fetal parts. These strands can cause anomalies by malformation, deformation or disruption. Amniotic rupture occurring early in gestation can cause multiple malformations, whereas, rupture occurring later causes limb anomalies.[1,3] Vascular insult occurring early in the embryogenesis is also proposed to be a cause for ABS.[8]

Orioli *et al*. have reported an increased incidence of ABS in the population living at a high altitude, in primipara, in women with a history of febrile illness in the antenatal period, and in women with a history of vaginal bleeding in the first trimester.[2] In our case series, all three women were primipara.

Antenatal detection of an aberrant sheet or bands of tissue attached to the fetus with characteristic deformities and restriction of movements is diagnostic of amniotic band syndrome. Antenatal detection of the sheet or band in the amniotic cavity without associated deformity does not warrant a diagnosis of ABS. Diagnosis is important for the management of the present pregnancy and for reassurance regarding future pregnancies, as the chance of recurrence is low.[5]

Conclusion

In conclusion, we have described three cases of amniotic band syndrome, which were detected in the antenatal period. In two of the cases, we could see the amniotic band antenatally.In the case where it was not seen, the anomaly was diagnostic of ABS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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