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Case Report

Amniotic Band Syndrome with Calvaria Involvement Mimicking Anencephaly During a Mid-Trimester Ultrasound Scan - A Case Report

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Abstract

Amniotic band syndrome (ABS) consists of multiple irregular fibrotic bands stretching out from placenta to the fetus that can result in many fetal abnormalities. The calvaria involvement may mimic anencephaly. A 30-year-old primigravida with anomaly ultrasound scan at a gestational age of 22 weeks showing a poorly formed fetal head with bulging eyes giving the typical 'frog eye' appearance of anencephaly and linear echogenic structures arising from the placenta into the amniotic cavity consistent with an amniotic band. A diagnosis of anencephaly was made. She was counseled on the prognosis of the fetal anomaly, and she opted for termination of the pregnancy. She expelled a fresh female stillborn with well-formed cranium which had a deep laceration on the scalp and extrusion of the brain substance. A diagnosis of amniotic band syndrome with calvaria involvement was made. Congenital anomalies caused by ABS are unique and when it involves the calvarium, it can mimic anencephaly.

Keywords: Amniotic band syndrome, anencephaly, fetal anomaly.

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Introduction

Anencephaly is a lethal anomaly resulting from a disturbed closure of the neural tube. It is the second most common neural tube defect after spina bifida occurring in 3 per 10,000 births. The diagnosis is usually made during the first trimester ultrasound between 11th and 14th week of gestation by the non-visualization of the ossification of the cranial box and exposure of the brain.[1]

Anencephaly is one of congenital anomalies that can be diagnosed with 98-100% accuracy during the routine anomaly ultrasound scan at 20 weeks and in the hand of experts, at 14 weeks. This accuracy is because of some sonographic features that have been associated with it. These include the 'Beret' sign in the first

trimester and the 'frog eye' sign in the second and third trimester.[1,2] We present a case of a 22-week fetus who had typical features of anencephaly on ultrasound and an incidental finding of a linear echogenic band suggestive of an amniotic band on ultrasound. However, a diagnosis of amniotic band syndrome was made after expulsion of the fetus.

Case Report

A booked 30year old primigravida booked for routine antenatal care at our hospital at a gestational age (GA) of 14 weeks and 2 days with a scan done at 9 weeks in outside facility confirming a life intrauterine fetus. The pregnancy was unplanned and spontaneously conceived. She had no underlying medical condition, and she was not on any routine drug apart from haematinics prescribed at the hospital. Her booking parameters were normal.

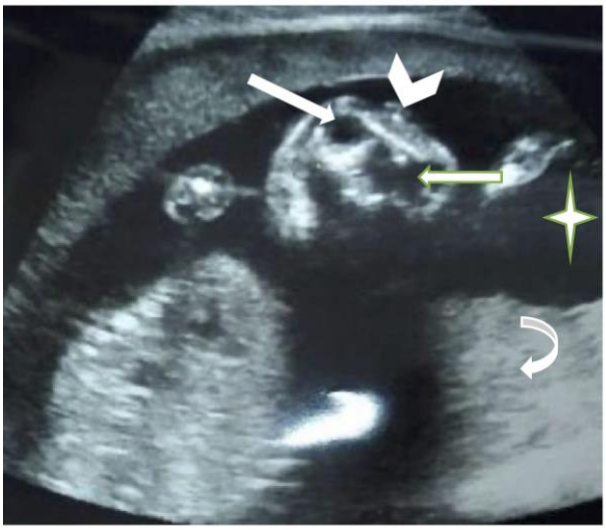

Figure 1- A gray scale ultrasound image of the fetus at 22wks gestational age showing the bulging eyes (white arrows) giving the frog eye appearance and small brain substance (arrowhead). The placenta is seen posteriorly, and the amniotic fluid was adequate(star).

An anomaly ultrasound scan at a gestational age of 22 weeks and 1 day, showed a single live fetus with non-visualization of the cranial bone with bulging eyes giving the typical 'frog eye' appearance (Figure 1). An incidental finding of a linear echogenic structure arising from the placenta into the amniotic cavity was also seen consistent with an amniotic band (figure 2). No other fetal anomaly was seen. The placenta was posteriorly placed, and the amniotic fluid was adequate.

A diagnosis of anencephaly coexisting with an amniotic band was made. The patient and her husband

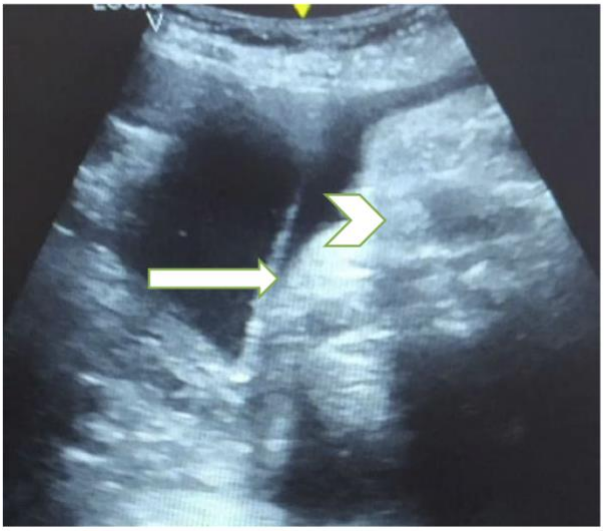

Figure 2: A coned down grey scale ultrasound image showing a linear echogenic structure(arrow) extending into the amniotic cavity from the placenta (arrowhead) consistent with an amniotic band.

Figure 3: Pictures of the baby after termination showing a well-formed skull. A deep laceration is noticed on the skin over the frontal bone with brain tissues extruding from it.

were counseled on the severity and prognosis of the condition viz-a-viz incompatibility with extra-uterine life and pregnancy associated complications. They subsequently opted for a medical termination of pregnancy and gave an informed consent.

Termination of the pregnancy was done the following day with the use of prostaglandin analogue (misoprostol). Patient expelled a fresh stillborn female baby with a well-formed cranium. A laceration was noticed on the skin over the frontal bone with band of amniotic membrane attached and extrusion of the brain tissues (figure 3). A diagnosis of amniotic band syndrome (ABS) was then made.

She had manual removal of placenta done in the theatre on account of a retained placenta. She was placed on antibiotics and discharged home thereafter.
Consent for the case to be published (including images and case history) was obtained from the patient.

Discussion

Amniotic band syndrome (ABS) also known as Familial amniotic bands; Streeter dysplasia and Congenital constricting bands is a syndrome that consists of multiple irregular fibrotic bands stretching out from placenta to the fetus. Many deformities or malformations result from it and the severity depends on the part of the body entangled or entrapped by the constricting band. Many of the reported cases of deformities arising from this syndrome usually involve the extremities however cases of craniofacial abnormalities have been reported.[3,4] Craniofacial deformities that could arise from ABS include encephalocele, acrania, exencephaly, anencephaly, cleft lip and palate, nasal deformities and asymmetrical microphthalmia. The calvaria involvement of the amniotic band syndrome (ABS) may result in atypical anencephaly as seen in the presented case which was as a result of deep skull laceration that caused extrusion of all the brain tissue making it to mimic anencephaly. The difference between the two is the asymmetric preservation of the calvarium in ABS which makes it different from typical anencephaly where there is complete absence of the calvarium.[5,6]

The syndrome occurs equally in both gender and there is no racial predisposition though some rare familial cases have been documented in literature.[4] The case presented was found in a female fetus.

Amniotic bands are caused by damage to a part of the placenta called the amnion and this damage produces fiber-like bands that can trap parts of the developing fetus. A rare cause of amniotic band is persistent chorioamniotic separation (CAS) after 17 weeks gestation and this has been linked to preterm delivery, premature rupture of membrane, umbilical cord strangulation, fetal growth restriction and fetal death. Chorionamnion separation can occur spontaneously which is called primary CAS. It is however very rare, with only few cases documented in the literature. Secondary CAS could occur after amnioncentesis, amnio-reduction, fetal surgery and fetal blood sampling. It can also occur from maternal abdominal trauma and the use of some drugs in early pregnancy, for example acetaminophen.[3,5-8] None of the risk factors was identified in the index patient.

The mainstay of diagnosis of ABS has been prenatal ultrasound in which the amniotic bands appear as thin, mobile lines, which may be seen attached to or around the part of the fetus involved. In the presented case the amniotic band was seen on ultrasound, but it was separate from the part involved. This may be because the fetus is small and had adequate amniotic fluid enhancing her movement. It should however be noted that it is not in all cases that the bands are detected by ultrasound and sometimes diagnosis are made when the abnormalities are seen on ultrasound and are unique such in the case presented.[7-10]

For the management of amniotic band syndrome generally, the parents are adequately counselled on options of treatment and prognosis based on the severity of the affected parts of the fetus involved. For the mild cases, no definitive treatment is recommended except monitoring of the bands as growth occurs to watch for progressive constriction and swelling. Other constrictions may require surgical management. Surgical options available depend on the abnormality and these include fetoscopic laser therapy and the use of prosthesis after delivery for the amputated limbs. However, for severe cases like the case reported that was diagnosed in utero involving the calvarium with extrusion of the brain tissues, multidisciplinary management is needed, and options of management include termination of pregnancy after adequate counselling. This is because most of the fetuses will suffer early neonatal deaths.[6,10-12] Our patient was counselled, and she opted for termination of pregnancy.

Conclusion

It is important to differentiate cranial abnormalities due to amniotic band syndrome from neural tube defects. Congenital anomalies caused by amniotic band syndrome are unique but when it involves the calvarium, it can mimic anencephaly therefore high index of suspicion is needed during anomaly scanning so as not to miss it.

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