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Anencephaly associated with amniotic bands: A case report and review of the association

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Abstract
Introduction: Anencephaly is a relatively common fatal neuro-developmental disorder. Routine prenatal screening using ultrasound diagnoses the condition frequently and aid in managing the cases appropriately. Anencephaly is characterized by a severe developmental anomaly of the brain and a defective skull. Amniotic band syndrome is a condition associated with multiple congenital anomalies, which are often a consequence of fibrotic amniotic bands extending from the placenta to the fetus. It is a rare association but can be recognized by first trimester or late obstetric ultrasound. Amniotic band syndrome may be associated with anencephaly and often attributed as a causative factor. Apart from the amniotic band syndrome, many other conditions are associated with amniotic bands and the syndrome may be labeled erroneously if only the bands are taken into consideration.
Case: A 27-year-old, G2P1 lady at 12 weeks and 2 days gestational age came for a routine ultrasound evaluation. The fetus was found to have a deformity of the head characteristic of anencephaly sequence. Multiple fibrotic bands were seen in the amniotic cavity extending from the placenta towards the fetus side, suspicious of amniotic band syndrome. The pregnancy was subsequently terminated on the patient's request.
Conclusion: The association of anencephaly and amniotic band syndrome is well known and the bands are often responsible for the spectrum of anencephalic anomalies. Asymmetric defects in the skull and brain are the norm in such cases along with multiple defects in the body parts. Both the anomalies are in general severe and may warrant early intervention. More benign amniotic bands like chorioamniotic separation may be encountered in pregnancy and should not be confused with more dangerous amniotic band syndrome. Recognition of amniotic bands is important, as many bands regardless of the etiology were found to be potentially harmful.

Keywords: Amniotic band syndrome, anencephaly, anencephalic spectrum, exencephaly, prenatal ultrasound, chorioamniotic separation

1. Introduction

Amniotic band syndrome (ABS) is a rare anomaly with an incidence of 1:200 to 1:500 live births with manifestations ranging from simple constriction rings to severe defects with potential for miscarriage. It affects both sexes equally and has a higher frequency among Afro-Caribbean descent [1]. Most of the reported deformities predominantly involve the extremities. Higher incidences of craniofacial abnormalities are also reported as high as 78% in some series in comparison to extremity anomalies in about 70% [2]. Cranial changes include encephalocele, acrania, exencephaly, and anencephaly [1]. Fetopathologic studies reveal three types of lesions in ABS: (1) constrictive tissue bands, which are caused by primary amnion rupture with subsequent entanglement of fetal parts (mostly limbs) by amniotic strands (2) amniotic adhesions, the result of a broad fusion between disrupted fetal parts (mostly cephalic) with intact amniotic membrane and (3) more complex anomaly patterns or limb-body wall complex including most of the craniofacial defects (encephalocele and/or facial cleft) occurring as a result of vascular disruption sequence with or without cephalo-amniotic adhesion [3]. Thus amniotic band syndrome produces a range of anomalies including craniofacial clefts, thoracic or abdominoschisis, amputations, ring constrictions, placental adhesions, and other internal malformations. Histologically amniotic bands are fibro-connective tissue and flattened epithelial cells together with neuroectodermal elements.

The abnormal number of vessels in the umbilical cord is also a common pathological association. Studies correlating etiology found amniotic disruption, vascular disruption or genetic disruption could explain the amniotic band syndrome/limb body wall complexes, alone or in combinations [4]. Anencephaly is a spectrum of severe neural tube defects due to failure in the closure of the rostral end of the neural tube by 5-6 weeks. Similar pathology is also seen in acalvaria which is due to the absence of calvarial bones with normal development of chondrocranium but with the presence of cerebral hemispheres. Acrania, exencephaly, and anencephaly are the immediate differential diagnosis as facial structures and the base of the brain may be intact in all these conditions. In anencephaly, the brain is typically absent above the orbit with bulging eyes and a frog-like facial appearance. Exencephaly is advanced acrania with a large amount of disorganized brain tissue at the base of the cranium with convolutions or pseudo-sulcal pattern in ultrasound. Exencephaly is regarded as a precursor of anencephaly in embryological development. Acrania and exencephaly may eventually progress to anencephaly. About five percent of anencephaly may be associated with ABS. The presence of fibrous bands at constriction points and typical craniofacial deformities which are often bizarre in non- embryological locations may help in the diagnosis. ABS cases are sporadic events in otherwise normal families with no known genetic abnormalities [5, 6]. It is an easily diagnosable anomaly by prenatal ultrasound by 14 weeks. Missing in early ultrasound may be due to very early first trimester scans done before 11 weeks of gestational age [7].

2. Case presentation

A 27-year-old female, G2P1, came for a routine initial prenatal ultrasound scan at 13weeks and 1 day after her last menstrual period. Her medical history was insignificant with no major illness in the past. Her first pregnancy was unremarkable with a healthy baby. The initial sonogram revealed a small deformed head with a flat vertex. The bony calvarium was absent. The brain appeared deformed with no formed hemispheres. The brainstem appeared unremarkable. The thalamus and basal ganglia were not recognizable. A single irregular ventricular cavity was noted in the brain. No evidence of any meningo-encephalocele was noted. The face appeared unremarkable, except for the prominent globes. Findings were consistent with anencephaly (Figure 1). The fetus was reasonably mobile. No truncal or extremity anomalies were noted. Internal organs like the heart, kidneys, and spine were unremarkable. The placenta was on the posterior wall and appeared unremarkable. No abruption was noted. Multiple thin linear bands of membranes were noted extending from the placental surface towards the fetus. Peculiar association of anencephaly and multiple thin amniotic bands were noted. The bands were well walled off by a wall of an intact amniotic membrane from the fetus. Amniotic bands appeared to be an isolated feature with no definite anomalies directly attributable to it. Features indicate chorioamniotic separation with amniotic bands (Figure 2, 3). Because of anencephaly, the patient was counseled and agreed to an early termination of pregnancy by medications. The fetus was expelled intact which showed the cranial defects characteristic of anencephaly (Figures 4). The placenta and membranes were easily separable from the fetus.

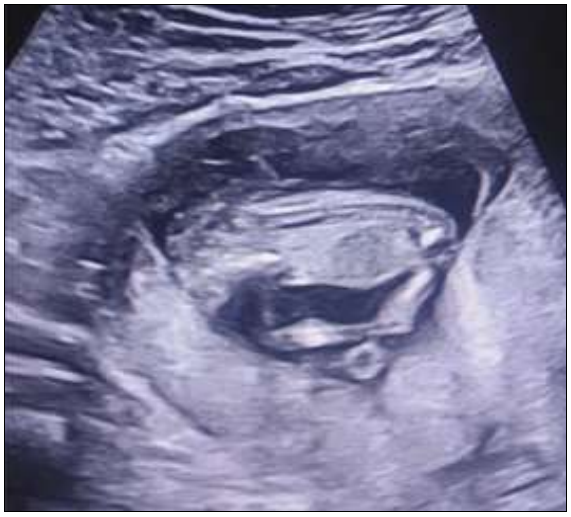

Fig 1: Sagittal ultrasound image of the fetus at 13 weeks 1 day gestational age showing absent cranium and brain characteristic of anencephaly.

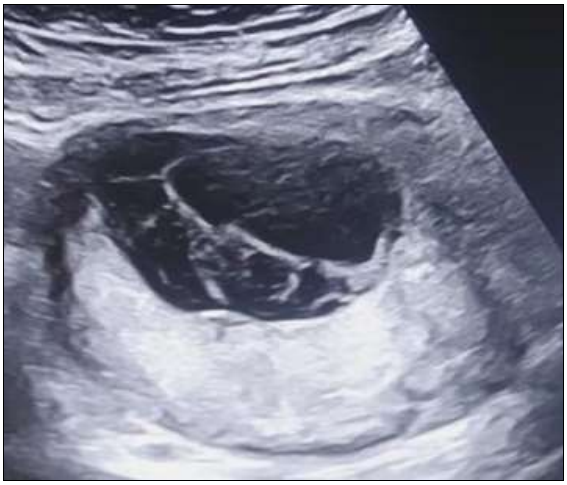

Fig 2a: Oblique section of the uterus showing placenta with radiating bands enclosed by the amniotic membrane indicating chorioamniotic separation.

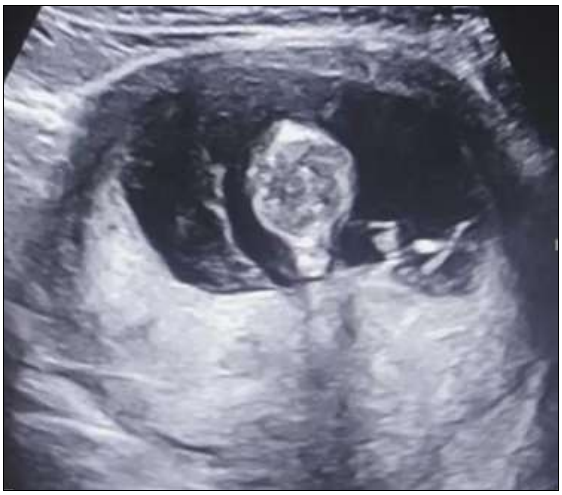

Fig 2b: A transverse section shows the body of the fetus separated from the amniotic membrane and chorionic bands.

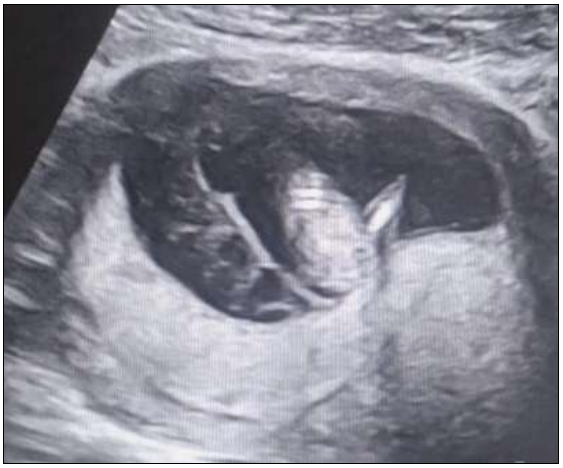

Fig 3a: Oblique ultrasound section showing chorionic bands, membrane, and the fetus.

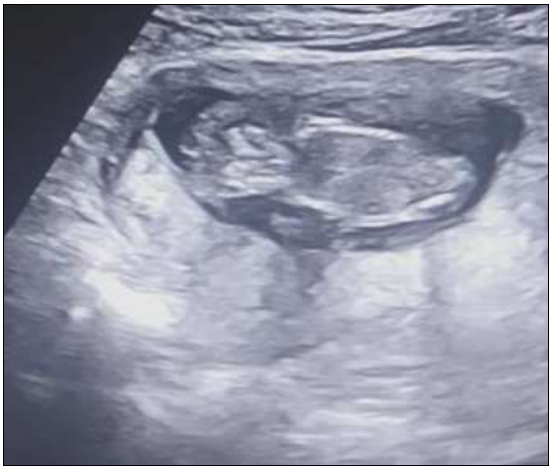

Fig 3b: Coronal section of fetus showing relatively normal facial and body parts.

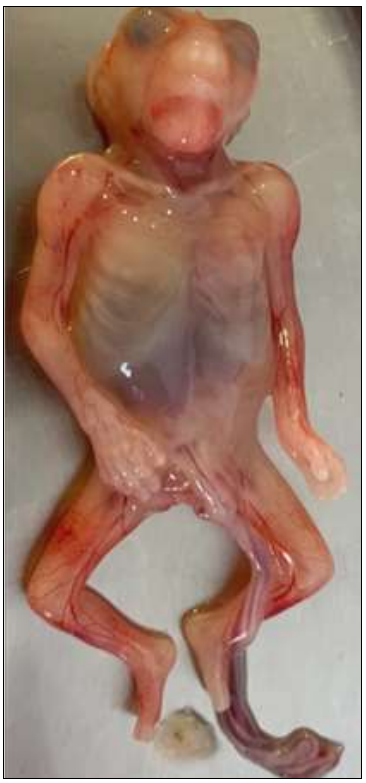

Fig 4a: Frontal profile picture of the fetus showing face and prominent frog-like eyes.

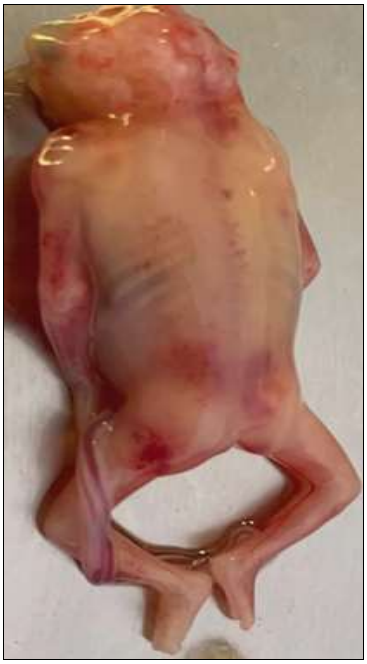

Fig 4b: Lateral profile of the fetus showing absent skull, covering membrane and a rudimentary brain matrix near the base of skull indicating anencephaly.

3. Discussion

Anencephaly is easily diagnosed prenatally with sonography after 14 menstrual weeks and the sonographic diagnosis is primarily based on the absence of brain and calvarium superior to the orbits on coronal views of the fetal head. This typical appearance may be absent in about 45% of cases with the presence of echogenic tissue superior to the orbits corresponding to angiomatous stroma (area cerebrovasculosa). Sizable angiomatous stroma in the form of solid or mixed solid- cystic tissue mimicking normal brain may be seen in up to 20% of fetuses [8]. Anencephaly is not an uncommon anomaly occurring about 1 in 1200 to 10 in 1000 pregnancies worldwide [8, 9]. The etiology is believed to be multifactorial with any individual factor alone is insufficient to produce this severe defect in the neurulation. Anencephaly is more common with mothers with previous anencephaly, among twins, female fetuses, and certain genetic diseases like trisomy 13, trisomy 18, and triploidy. Other nongenetic factors include gestational diabetes, metabolic syndrome, maternal hyperthermia, drugs like valproate, and environmental pollution. Inadequate maternal nutrition mainly affecting the folate-related metabolism is believed to be a cause. It includes deficiency of folate, Vitamin B12, zinc, and abuse of alcohol, caffeine, smoking etc [9]. Anencephaly may be part of a spectrum of anomalies starting with acrania (absence of cranium, which exposes the brain to mechano-chemical effects of amniotic fluid), leading to exencephaly (defective brain) to anencephaly (near-complete absence of brain parenchyma) [9, 10, 11]. Anencephaly spectrum needs to be distinguished from the cranial defects associated with the amniotic band syndrome which is asymmetrical unlike the symmetric cranial defect of typical anencephaly. The absence of typical anomalies of amniotic band syndrome including limb, body wall, and spinal abnormalities may further aid the diagnosis [8]. The amniotic band syndrome (synonyms-amniotic band disruption complex, limb-body wall complex) has a frequency of 1 in 1200 live births with manifestations

ranging from mild deformities to severe anomalies incompatible with postnatal life. The pathogenesis of amniotic band syndrome (ABS) is thought to be a disruption of the amnion. Thus the embryo or fetus may enter the chorionic cavity. This may expose the fetal parts to the fibrous septa that traverse the chorionic space and result in random entrapment by these bands. The fetal extremities, head, or trunk may be involved in combination or isolation [12, 13, 14]. Calvarial involvement in ABS may result in encephalocele and if severe may resemble anencephaly. Still, anencephaly associated with ABS may show asymmetric preservation of calvarium, unlike the complete absence of calvaria in typical anencephaly [12]. Two main theories for the etiology of amniotic band syndrome are the intrinsic theory of inherent developmental defects proposed by Streeter and the more favored extrinsic theory proposed by Torpin which suggests adhesive bands around the fetus due to slippage of the ruptured amnion from the chorion, leading to oligohydramnios and growth abnormalities [15, 16]. Amniotic band syndrome characterized by multiple irregular fibrotic bands extending from the placenta to the fetus is a serious diagnosis. It must be differentiated from other benign and frequent causes of intrauterine linear echogenicity in a gravid uterus which includes membranes of multiple gestations, uterine synechiae with amniotic sheets, uterine duplication anomalies, and circumvallate placenta. Other rarer causes are chorioamniotic separation and inter-membrane hemorrhages. Amniotic band syndrome is rare and diagnosis is ideally deferred without typical fetal defects [8, 16]. Nevertheless, early detection of such membranes may be significant, so that appropriate follow up may be done to watch for any developing anomalies in the future. Early scans demonstrating just membranes without typical anomalies should not be advised for immediate termination, but worthy to take close follow up to make a precise diagnosis of ABS later and monitor for any bad fetal outcomes [17]. The chorionamnion separation before 14 weeks of gestation is regarded as physiologically normal. Amnion-chorion fusion usually occurs between 14 and 16 weeks. Persistent chorioamniotic separation (CAS) after 16 weeks is uncommon and abnormal [18]. CAS can occur spontaneously and are called primary. More often, they are secondary after an intrauterine intervention like amniocentesis, fetal blood sampling, and fetal surgery. The literature on spontaneous CAS is sparse with only a small number of cases [19]. After 17 weeks CAS is associated with adverse perinatal outcomes, such as amniotic band syndrome, umbilical cord strangulation, and fetal death [18, 19, 20]. CAS has also been associated with preterm delivery, premature rupture of membranes, and fetal growth restriction [18]. CAS is associated with a preterm birth rate of 40% and intrauterine fetal demise or miscarriage of 28% [20]. Benign looking amniotic membranes may be possible as in this index case, where the association with exencephaly-anencephaly appears merely incidental. As more and more cases of amniotic membranes are identified in early pregnancy before 14 weeks thanks to high sensitive sonography, appropriate follow-up and management strategies can be adopted. Missing bands including those iatrogenic amniotic bands are a concern that can prove fatal in later fetal life and maybe benefited from follow-up ultrasound [21]. Recently intrauterine fetal interventions including fetoscopic laser surgery are explored as a treatment option for amniotic band syndrome [22] and thus making early diagnosis more important. Amniotic bands that develop secondary to iatrogenic complications such as amniocentesis, amnioreduction, and septostomy in twins, known as pseudoamniotic band syndrome, may be associated with fetal anomalies later in life similar to amniotic band syndrome [15].

4. Conclusion

Amniotic bands are becoming common findings in prenatal ultrasound examinations thanks to sensitive imaging studies. It is worthy to characterize them and followed up to exclude a more serious anomaly of amniotic band syndrome and pseudoamniotic band syndrome. Anencephaly may be associated with amniotic band syndrome which typically produces asymmetric defects. Identification of all amniotic bands with further characterization is warranted as they have the potential for defects later in pregnancy regardless of etiology and may be followed up by ultrasound.

Competing interests

The authors did not report any potential conflict of interest.

5. Acknowledgments

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