

Case Report

Amniotic Band Syndrome: about Two Uncommon Cases

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• Antenatal diagnosis; Fetal malformations; Amniotic band syndrome

Abstract

Background: Amniotic Band Syndrome is a congenital disorder presenting with fetal anomalies associated with fetal placental fibrous bands that may cause disruptions, deformations or malformations without a consistent anatomical pattern. Anomalies are limited to external structures with or without body wall disruption or internal malformations that vary in severity and location. These malformations are characterized by being asymmetric, polymorphic and not respecting any embryological systematization. The pathophysiology of this syndrome is the subject of a debate that started more than seventy years ago and continues to this day.

Case presentation: We report here two cases of amniotic band syndrome. The first one is that of a 22-year-patient admitted for delivery at 37 weeks of amenorrhea. Newborn examination at birth revealed the presence of upper limb ischemia, whose therapeutic decision following the unfavorable evolution towards necrosis with gangrene was the amputation of the upper limb.

The second case is that of a 29-year-old woman pregnant at 27 weeks, whose ultrasound scan showed the presence of anencephaly associated with a myelomeningocele adherent to the placenta. The therapeutic decision was to convene the ethics committee to agree to the medical termination of pregnancy, which was deemed admissible given the lethality of the malformation.

Discussion: Amniotic Band Syndrome includes a wide variety of fetal anomalies associated with fibrous bands that entangle or constrict various parts of the fetus or umbilical cord in utero. Despite numerous studies, no pathogenesis theory has been proven to date. Diagnosis is possible as early as the first trimester, depending on the nature and severity of the malformations observed. The therapeutic decision varies according to the case.

Conclusions: ABS is uncommon but screening should be systematic in the neonatal period. In order to improve diagnosis, it is necessary to promote antenatal screening reference centers that assess suspected cases of rare malformations.

BACKGROUND

Amniotic Band Syndrome (ABS) is a congenital disorder presenting with fetal anomalies associated with fetal placental fibrous bands that may cause disruptions [1-3], deformations or malformations [4], without a consistent anatomical pattern [5,6]. Anomalies are limited to external structures with or without body wall disruption or internal malformations that vary in severity and location [7-9]. ABS, whose incidence is reported to be between 1/1,200 and 1/15,000 births, is a group of complex congenital malformations mainly affecting the limbs. The clinical aspects of the limbs are varied with presence of cutaneous grooves of amputation, pseudo-syndactyly and club feet [3-6]. More rarely, the cranio-facial region is affected (exencephaly, anencephaly, oblique and asymmetric facial clefts) as well as the thoracic-abdominal region (laparoschisis, omphalocele, bladder exstrophy). The presence of flanges is not necessary for the diagnosis [1]. Deaths in utero by constriction of the cord by an amniotic flange have also been reported [2]. The prognosis of ABS depends on the severity of the lesions which can range from a simple skin fold to life-threatening incompatible with life. The clinical signs have been well known for many years [1-6], but the

etiologies and especially the pathophysiology of this syndrome are the subject of a debate that started more than seventy years ago and continues to this day [3,4].

We hereby report a series of two cases of ABS through which we try to recall the diversity of clinical forms, the importance of prenatal diagnosis but also to discuss the supposed mechanisms of its physiopathology and the different therapeutic possibilities.

CASE PRESENTATION**First case**

The patient was 22 years old with no history of consanguinity, primigravida primiparous with an estimated pregnancy of 37 weeks of amenorrhea and 4 days according to the first trimester ultrasound, with a history of pre-eclampsia on methyldopa 500mg (3 times daily) discovered at 31 weeks of amenorrhea, referred to our department for delivery. The admission examination found blood pressure 13/7, heart rate 89bpm, no neurosensory signs, afebrile with a BMI of 24.4, fetal heart sounds present and regular. Vaginal examination revealed a soft median cervix 80% effaced and dilated to 3 cm with cephalic presentation and intact

membranes. Ultrasound showed an evolving singleton pregnancy with fetal weight estimated at the 32nd percentile, a fundial placenta and normal amniotic fluid. Therefore, the patient was managed in our facility for delivery at 37 weeks of amenorrhea on uncomplicated preeclampsia and was taken to the delivery room.

After a smooth eight-hour labour and an uneventful partogram, the patient delivered a baby boy with an Apgar score of 10/10/10 and a birth weight of 2740g. The pediatrician was called in after finding ischemia of the right upper limb with a well-limited upper limit of circumferential appearance extending beyond the elbow and reaching the lower third of the humerus with the presence of a narrowing skin groove (Figure 1). Doppler ultrasonography showed patency of the arterial axes up to the elbow, with an absence of arterial flow distally without visualization of thrombosis. The biological work-up was initially normal. The treatment consisted of a bolus injection of 100 units/kg of unfractionated heparin, then a continuous injection at a rate of 20 units/kg/h, which resulted in an activated partial thromboplastin time of 2.4 times normal, in addition to a prophylactic antibiotic treatment with protected amoxicillin. The pediatric surgical team immediately rushed the neonate to the operating room to remove the constriction. However, the evolution was marked at 6 hours of life by the persistence of the absence of distal pulses with necrosis complicated by gangrene of the right upper limb (Figure 2). It was therefore decided to proceed with an amputation of the right upper limb. The neonate was hospitalized in a neonatal intensive care unit for 30 days with clinical and biological monitoring until the stump healed and then was discharged home.

Second case

This was a 29-year-old patient, without any notion of consanguinity, nor any notable personal or family pathological history, gravida 2 para 2 with a 3-year-old girl delivered vaginally in good health with good psychomotor development, referred to our structure by her referring doctor at 21 weeks of amenorrhea and 3 days for ultrasound suspicion of anencephaly. The morphological ultrasound performed in our facility showed an evolving monofetal pregnancy complicated by skull agenesis



Figure 1 First case at birth: ischemia of the upper limb with the presence of a cutaneous groove of stricture.



Figure 2 First case at six hours of life: evolutionary character of the ischemia with necrosis and gangrene.



Figure 3 Second case at birth: anencephaly associated with myelomeningocele.

in favor of anencephaly associated with a myelomeningocele adherent to the placenta with the appearance of the face difficult to appreciate and the existence of a probable hypertelorism. This ultrasound appearance was therefore in favor of a cranioschisis due to a neural tube closure defect at cranial level (Figure 3).

The decision was to convene the ethics committee for approval

of the medical termination of pregnancy, judged admissible given the lethality of the malformation. After agreement from the patient and her partner, we performed a misoprostol induction under strict surveillance to watch for any complications. The patient responded to the induction giving birth vaginally to a stillborn boy with a birth weight of 630g. Examination revealed a neonate with an externalized brain very adherent to the placenta consistent with ABS complicated by cranioschisis. The patient was discharged home at 3 days post-partum after initial care by the psychology team.

DISCUSSION

ABS is a congenital disorder presenting with fetal anomalies associated with fetal placental fibrous bands that may cause disruptions [1-3], deformations or malformations [4], without a consistent anatomical pattern [5; 6]. Anomalies are limited to external structures with or without body wall disruption or internal malformations that vary in severity and location [7-9]. The clinical aspects of the limbs are various with presence of cutaneous grooves of amputation, pseudo-syndactyly and/or club feet [3-6]. More rarely, the cranio-facial region is affected (exencephaly, anencephaly, oblique and asymmetric facial clefts) as well as the thoracic-abdominal region (laparoschisis, omphalocele, bladder extrophy) [1,2]. ABS is a relatively uncommon condition with an incidence from 1/1,200 to 1/15,000 live births [1,7].

These malformations probably result from multiple different pathological processes [3,10-13]. Two main theories have been discussed to explain its pathogenesis. According to Streeter et al. endogenous theory published in 1930 [14], it is a malformation of the germinal disc that leads to a disturbance of the development of the amnion. Streeter et al. [14], suggested that amniotic bands appeared early and were not the primary cause of defects but a consequence of an imperfect histogenesis that caused necrosis, scarring, constrictions and fusions; the deficiency in one or two cells resulted in a larger anomaly of the final structure [14,15]. McKenzie et al. [16], supported this theory and suggested that tissue alterations were an example of an abnormal distribution of cell death areas. Hartwig et al. [17], suggested that limb and lateral body wall anomalies were caused by defects in ectodermal placodes which resulted in mesodermal deficiency and, therefore, abdominal wall deficiency, that was substituted by the amnion and in the case of rupture, it allowed the abdominal content to fill the extraembryonic celom [15-17]. Facial and limb anomalies were attributed to a defect in the craniofacial placodes, whereas internal anomalies were explained by the alteration in lateral abdominal placodes that caused an intermediate mesoderm deficiency [14-17].

The exogenous theory of Torpin et al., published in 1965 [7], is opposed to the previous one; it is based on the premature rupture of the amnion, the debris of which forms constrictive flanges on the fetal skin surface, the authors adopted a mechanical explanation in which the flanges were the cause of the stricture grooves and the amputations of the ABS. The primary event would be an early rupture of the amnion leading to chorio-amniotic detachment [7]. Some elements of the fetus, especially the extremities, could then pass through these breaches into the chorionic cavity, which usually closes at 12 weeks of amenorrhea.

Bands made up of fibrous tissue of mesodermal origin, coming from the external surfaces of the amnion and the internal surface of the chorion, would be formed and would come to wrap themselves around the externalized parts of the fetus [7]. Therefore, it is by a phenomenon of ligation and striction that the typical lesions of the disease would be formed. Depending on the stage of pregnancy at which the amnion rupture occurs, the lesions would be more or less severe, the very early forms being lethal [18,19]. In this theory, the actual cause of the rupture remains unknown. Some have suggested a traumatic or infectious origin [20]. Amniocentesis has also been cited as potentially responsible [19-21]. In spite of a great deal of work, neither theory has been proven to date. It is highly probable that several non-exclusive mechanisms participate in the formation of the different lesions of this disease [3].

Diagnosis of ABS is possible as early as the first trimester, depending on the nature and severity of the malformations observed [8,9]. Craniofacial and thoracoabdominal anomalies can be detected at the first ultrasound scan at 10-13 weeks of amenorrhea whereas isolated limb anomalies are usually detected at the time of a new ultrasound examination [9]. In our first case, the diagnosis could only be made at birth as the constriction resulting from ABS only concerned the right upper limb. Whereas in our second case, it was the first ultrasound made unfortunately at 21 weeks of amenorrhea that detected the anencephaly and allowed a medical termination of pregnancy. A two-dimensional ultrasound allows to detect the ABS in the second trimester of gestation; however, unlike a three-dimensional ultrasound, it has certain limitations to obtain orthogonal multi-planar images of the cavity and the surface, which help to see the defect and the adjacent structures [22-25]. A prenatal ultrasound marker suggestive of ABS is the presence of detached amnion in the cavity and structural assessment of the fetus should then be performed to exclude other anomalies [9]. If an ultrasound image indicates the presence of amniotic bridges, a differential diagnosis with polylobed placenta, intrauterine adhesions and uterine septum should be made [23-26]. Antepartum detection of an amniotic flange is not essential for its diagnosis; however, pathological examination should confirm the diagnosis [24-26]. Thus, the diagnosis should be evoked when the following characteristic signs are present: constriction or asymmetric amputation of one extremity of a limb with lymphoedema downstream of the constriction; asymmetric craniofacial malformations (encephaloceles, cleft lip and palate); coelosomies; pseudo-syndactyly; presence of an amniotic flange in contact with the injured fetal pole [24-26].

Obstetrical prognosis of patients with a fetus with an ABS-related anomaly is not altered compared with the general population [2,12,13]. In particular, in cases of craniofacial or visceral malformation, there is no increased rate of presentation anomalies or dystocia [2,13,14]. At birth, the amount of amniotic fluid is considered normal in the majority of cases [2-4]. However, some authors [6,14], report an increased rate of late miscarriage and prematurity in cases of ABS. In our cases, the deliveries performed vaginally were uneventful and the amount of amniotic fluid was considered normal. While the first case of ABS in our series benefited from postnatal therapeutic management by amputation of the right upper limb, the second case was beyond

therapeutic resources and death was inevitable. Termination of pregnancy seems reasonable in this type of case.

CONCLUSIONS

ABS is uncommon but screening should be systematic in the neonatal period. In order to improve diagnosis, it is necessary to promote antenatal screening reference centers that assess suspected cases of rare malformations. The obstetrical attitude will be adapted on a case-by-case basis and will depend on the severity of the syndrome. A medical termination of pregnancy may be proposed when the fetal malformations are recognized as being incompatible with life. Otherwise, multidisciplinary management of the pregnancy, which also remains a challenge, is essential in order to explain to the couple the difficulty of establishing a functional prognosis in the ante-natal period, as well as the post-natal and even prenatal therapeutic possibilities.

DECLARATIONS

Guarantor of Submission

The corresponding author is the guarantor of submission.

Availability of data and materials

Supporting material is available if further analysis is needed.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethics approval and consent to participate

Ethics approval has been obtained to proceed with the current study. Written informed consent was obtained from the patient for participation in this publication.

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