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Case Report

Prenatal Diagnosis and Postnatal Treatment in a Case of Abdominal Obstruction and Polyhydramnios Caused by Amniotic Band Sequence

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ABSTRACT

Background: Amniotic band sequence (ABS) is a rare congenital disorder resulting from the entanglement of fetal parts by fibrous bands that may cause disruptions, deformations, or malformations that can range from mild to life threatening conditions. Prenatal diagnosis is usually based on ultrasounds visualization on amniotic bands attached to fetal parts, possibly causing fetal defects.

Case Presentation: A 19-year-old woman with an unremarkable clinical history and a low-risk pregnancy was referred to our Fetal Therapy Unit for a suspected fetal clubfoot at 22 weeks gestational age. A chorioamniotic separation, together with unilateral clubfoot was diagnosed. Due to a high risk of premature rupture of membranes a decision was made not to perform amniocentesis for genetic investigation. At prenatal follow up progressive polyhydramnios developed with a preterm spontaneous rupture of the membranes at 34 weeks. After caesarean section – due to breech presentation – an amniotic band was observed at the abdominal level causing a circular skin lesion, constriction, and a sub-occlusive intestinal status. Moreover, a left clubfoot with amputation of distal phalanx of the second toe and a right foot amputation of 2nd and 3rd toes were evident. The neonate underwent abdominal plastic surgery on the second day of life with complete recovery. Following the postnatal diagnosis of ABS, we re-examined a stored fetal 3D ultrasound volume acquired at 22 weeks and, focusing on the fetal surface rendering, we could notice the deep abdominal skin furrow that was evident at birth.

Discussion: This case represents an unusual antenatal presentation of an amniotic band sequence that escaped prenatal diagnosis with a progressive polyhydramnios as an indirect sign of fetal bowel obstruction caused by an abdominal constricting band. In the presence of chorioamniotic separation and additional ominous ultrasound findings, it is advisable to consider the possibility of an ABS.

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Introduction

Amniotic band sequence (ABS) is a rare congenital disorder resulting from the entanglement of fetal parts by fibrous bands that may cause disruptions, deformations, or malformations [1]. The severity of ABS can range from mild to life-threatening depending on where the bands are constricting and how tightly they are twisted, with the most common

manifestations involving limb deformities. The reported incidence of ABS varies from 1 in 1200 to 1 in 15,000 live births, and its aetiology remains uncertain and a matter of debate [1]. Prenatal suspicion of ABS is based on ultrasound visualization of amniotic bands that appear as thin echogenic strands attached to a fetal part and usually connected to the uterine wall. Constriction ring defects of the extremities and irregular amputations of fingers and/or toes may be visualized as well as clubfoot,

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partial or complete limb amputation, facial or cranial clefts and, less commonly, chest wall or abdominal wall defects. Fetal fixed position or restriction of certain movements can also be a diagnostic clue. The differential diagnosis includes body stalk anomaly, specific craniofacial defects or abdominal wall defects, amniotic sheets (where the amnion is wrapped around intrauterine synechiae with no evidence of fetal involvement), and chorion-amnion separation [1]. This latter condition is characterized by the evidence of separation of the amniotic sheet from the uterine wall without entrapment of fetal parts.

Intrauterine fetoscopic lysis of bands before severe progression of vascular compromise can restore normal perfusion and prevent amputation [2]. However, the efficacy of this investigational approach is still unproven, and no consensus exists regarding the selection criteria for candidates for in utero intervention. In fact, this surgical approach is burdened by the possible preterm premature rupture of the membranes and premature birth [2]. In addition, although fetoscopic release of the band with scissors or laser may restore blood flow and save the limb, plastic surgery may be needed after birth to limit the scar tissue damage.

Case Report

Herein we present the case of a 19-year-old woman, gravida 1, who was referred to the Fetal Therapy Unit "U. Nicolini" for a suspected fetal clubfoot at 22 weeks gestational age (GA). No screening test for aneuploidy was performed since the first ultrasound in pregnancy was carried out at 16 weeks. Screening for common infections was negative, exposure to teratogenic substances was ruled out and personal and family histories were unremarkable. The ultrasound scan was performed at 22.0 weeks GA following ISUOG guidelines for neurosonographic, echocardiographic and anatomic evaluation of the fetus [3]. A diagnosis was made of unilateral clubfoot, with degree of severity defined by antenatal criteria for an adequate counselling [4]. Due to the presence of chorioamniotic separation, in an otherwise normal fetus, a decision was made not to perform amniocentesis for fetal karyotype and CGH-microarray, considering highest risk of premature rupture of membranes.

At 30 weeks GA, the woman was hospitalized for threatening preterm delivery with mild polyhydramnios, defined by a maximum vertical pocket of 10 cm. The 75 gr oral glucose tolerance test was negative, and so were maternal laboratory tests, investigated according to international guidelines [5, 6]. Daily fetal cardiotocographic monitoring was performed. The mother was discharged after 3 days in stable clinical conditions with planned weekly monitoring. Although the polyhydramnios worsened, due to chorioamniotic separation and its consequent risk of premature delivery, the woman decided to avoid amnio-drainage.

At 34 weeks a preterm spontaneous rupture of the membranes (pPROM) occurred. Antibiotic therapy was initiated as well as prematurity prophylaxis with steroids. Intensive feto-maternal monitoring was regular with negative inflammatory markers. Elective caesarean section was performed at 36.1 weeks GA for prolonged pPROM and breech presentation with the birth of a 2262 gr female new-born, Apgar scores of 8 at 1 minute and 9 at 5 minutes. At delivery, the surgeon faced difficulties in fetal extraction and had to remove a placental membrane attached to the fetal abdomen where a circular skin tear was evident at the belt level (Figure 1). A careful neonatal examination revealed a left

clubfoot with amputation of distal phalanx of the second toe and a right foot amputation of 2nd and 3rd toes (Figure 1).



Figure 1: Postnatal findings: **A)** Circular skin tear at the fetal abdomen belt level and **B)** left clubfoot with amputation of distal phalanx of the second toe and a right foot with amputation of 2nd and 3rd toes. **C & D)** 3D ultrasound volume of the fetal surface at 22 weeks with the deep abdominal skin furrow that was evident at birth.

A thorough postnatal evaluation was carried out by neonatologists and pediatric surgeons. The abdomen was extremely distended, and the baby presented vomiting and oral feeding difficulties linked to the extrinsic abdominal constriction. Abdominal x-ray evaluation showed an enlarged stomach associated with sub-occlusive intestinal status. On the 2nd day of life, the baby underwent surgery. The circumferential amniotic abdominal scar was completely removed to free the abdominal fascia deeply. An extended abdominal plastic surgery was performed to mobilize the skin and to reduce the adhesions between the subcutis and the abdominal fascia. A circumferential subcutaneous fat advancement flap was followed by a stretched direct closure. The histological examination of the removed tissue showed the presence of fibrotic material characterized by chronic inflammatory infiltrate.



Figure 2: Scar appearance, one month after surgery.

Post-surgical follow-up was uneventful, with no more signs of subcutaneous strictures and no abdominal distention (Figure 2). Following the postnatal diagnosis of ABS, we were able to re-examine a

stored 3D ultrasound volume of the fetal skeleton acquired at 22 weeks and, focusing on the fetal surface rendering; we could notice the same deep abdominal skin furrow that was evident at birth (Figure 1).

Discussion

This case represents an unusual antenatal presentation of an amniotic band sequence in a low-risk pregnancy complicated by fetal congenital clubfoot, early second-trimester chorioamniotic separation, and late-onset polyhydramnios. The cause of ABS is unclear but can be ascribed to both intrinsic and extrinsic factors. The intrinsic hypothesis suggests the development of constriction bands secondary to disruption of the developing embryonic disk due to impaired blood flow [7]. This hypothesis is supported by cases in which an intact amniotic sac was demonstrated, and it may also explain the involvement of internal organs observed in some subjects with ABS [7]. On the other hand, the more widely accepted extrinsic hypothesis suggests that ABS arises when the inner layer of the amniotic sac is damaged, resulting in the formation of thin strands of fibrous tissue that leads to the entrapment of fetal structures by mesodermic bands [7].

In the present case, a spontaneous chorioamniotic separation occurred early in pregnancy. Chorioamniotic separation is a physiologic finding in the first trimester while it is always abnormal beyond 16 weeks of gestation [8]. The iatrogenic separation after invasive procedures (such as amniocentesis or fetal surgery) has been well documented. According to a recent literature review, spontaneous chorioamniotic separation in singleton pregnancies is a rare condition associated with preterm delivery, premature rupture of membranes, low birth weight, and higher perinatal mortality [9]. Accurate estimations of these risks are not available since data are largely based upon case reports and small series, but a 5.1% incidence of amniotic bands sequence in association with spontaneous chorioamniotic separation was reported [9].

A case of circumferential amniotic band around the lower fetal abdomen that caused significant constriction of the viscera – with particular deformation of the urinary bladder – and associated with complete spontaneous chorioamniotic separation was recently reported [10]. Even in that case report, the significant abdominal scar was noted only after delivery and the neonate showed an appropriate feeding and voiding with a normal bowel function, likely because the constriction was at a lower level compared to our case. When neonatal abnormalities due to constriction bands are diagnosed, surgical repair should be performed as soon as possible to avoid immediate functional complications and long-term aesthetic sequelae. Even in the abdominal site the recurrence of atrophic aspects is represented by the appearance of a depression characterized by the “hourglass sign” constriction bands. This complication should be prevented since the first surgical procedure and the removal of the tissue scars must be followed by extended abdominal plastic surgery. Some Authors recommend an approach that mostly increases tissue intake rather than tissue removal, to reduce the contractile effect in the site of the scar with the aim to limit aesthetic complications. In neonates the large circumferential subcutaneous fat advancement flaps, that we performed in our case, may support this approach.

A singular clue in our case was the association with progressive polyhydramnios. Polyhydramnios is an abnormal increase in amniotic

fluid volume, typically diagnosed in the second or third trimester of pregnancy, that can be defined through sonographic assessment by a deepest vertical pocket ≥ 8 cm, or an amniotic fluid index ≥ 24 cm [5]. When polyhydramnios developed in the third trimester, many possible causes were investigated and further excluded. Due to amniotic sac misplacement, our diagnostic capability was limited, as we could just observe reduced fetal movements and feet malposition without being able to visualize toes' amputations and abdominal cutaneous furrow. This sonographic finding can be associated with a wide variety of underlying pathologies. Conventionally, three causes of polyhydramnios are possible: fetal malformations (in particular digestive atresia and cardiopathies), maternal disorders (e.g., diabetes and alloimmunization), and placental abnormalities (e.g., chorioangioma), but no cause is found in as many as 40 to 60% of cases identifying an idiopathic polyhydramnios [6]. Idiopathic polyhydramnios may be associated with macrosomia; instead, fetal growth restriction associated with polyhydramnios presents a high risk for an underlying fetal abnormality, including trisomy 13 or 18 [6]. The risk of maternofetal anomalies increases with the severity of polyhydramnios. Diagnostic ultrasound can reach a diagnosis in the majority of severe cases and amniocentesis offers a complementary assessment.

In our case, the fetal growth was within the normal ranges, and fetal anatomy was evaluated as normal, except for a unilateral clubfoot of moderate degree. To the best of our knowledge, this is the first report of an amniotic band sequence presenting with polyhydramnios as indirect sign of fetal bowel obstruction due to an abdominal circular band. We recommend always to keep in mind the possibility of ABS in the presence of chorioamniotic separation and additional ominous ultrasound findings.

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Availability of Data and Materials

All data presented in this paper are available upon reasonable request.

Consent

The patient gave her informed consent to publish her case (including publication of images).

Competing Interests

None.

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