



Case report

Amniotic band syndrome: A case report

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ABSTRACT

Amniotic band syndrome is a rare disorder, which consists of a combination of malformations in which the main feature is the existence of an amniotic band that can envelop the limbs, the body wall, and/or the viscera. We report a case of an antenatal diagnosis of amniotic band syndrome in a 21-year-old female patient at 20 SA + 1 day, who had a medical abortion.

1. Introduction

The amniotic band syndrome is a group of disorders involving the limbs, the craniofacial region and the thoraco-abdominal area, with a constricting skin band formed by fibrous tissue of chorioamniotic origin, enveloping the limbs, the body wall and/or the viscera, as the main feature. The origin of these malformations is multifactorial. Early and accurate prenatal diagnosis is the key element of management. The prognosis depends on the severity and location of the malformation. Medical abortion may be proposed in case of severe malformations. The work has been reported with respect to the SCARE 2020 criteria [1].

2. Observations

Mrs. M.E, 21 years old, non-consanguineous marriage, no particular pathological history, second gesture (one child alive by vaginal route), presented to our unit-care center for a 20 weeks of amenorrhea and one day pregnancy. The morphological ultrasound showed a progressive monofetal pregnancy, a type III caudal regression syndrome with sacral agenesis, associated to a spina bifida, myelomeningocele, scoliosis, omphalocele liver, spleen, intestine and gastric pouch with visible amniotic band. The rest of the morphology was quite normal (Fig. 1). Medical abortion was indicated because of the poor prognosis of the major malformations described above. After agreement of the couple, misoprostol induction was started, according to the International Federation of Gynecology and Obstetrics FIGO 2017's protocol. The patient successfully responded to the induction, giving birth to a male stillbirth by vaginal delivery, with a birth weight of 400 g. The clinical examination of the newborn had objectified the presence of a parietal

defect revealing the abdominal viscera, with a large skin band (Fig. 3). Moreover, the fetopathological examination was refused by the parents.

3. Discussion

Amniotic band disease is a group of congenital malformations involving the limbs, the skull, the face, and the thoraco-abdominal axis. These malformations can be described in two forms: amniotic band sequelae (ABS) and limb body wall complex (LBWC) syndrome Fig. 2 [2]. The origin of malformations is multifactorial, involving multiple pathological processes (genetic, infectious, or environmental). It is usually difficult to identify the exact cause. Authors have developed many theories (exogenous, endogenous, vascular or genetic) based on clinical findings, animal experiments etc., but none has been proven so far [3]. According to Streeter in 1930, it is an endogenous malformation of the germinal disc causing an amnion development anomaly. As for Torpin, his theory is opposed to the one previously described, and concerns the premature rupture of the amnion, the debris of which forms bands on fetus skin surface [2,4-6]. It is a relatively rare pathology. Its incidence is between 1/1200 and 1/15,000 births worldwide [2]. Its incidence in Africa is not known despite the frequency of congenital malformations in common practice. In Morocco, Hillali reported 24 cases of MBA in his study in 2013. As for Nagalo K from Burkina Faso, he reported only five cases [6,7]. Antenatal diagnosis is possible in the first trimester by early ultrasound, performed between the 10th and 12th gestational age, looking for craniofacial and thoraco-abdominal anomalies; limb anomalies are diagnosed around the 20-22nd gestational age. The diagnostic is suspected in front of constriction or amputation of the extremities, asymmetrical, associated or not with a downstream

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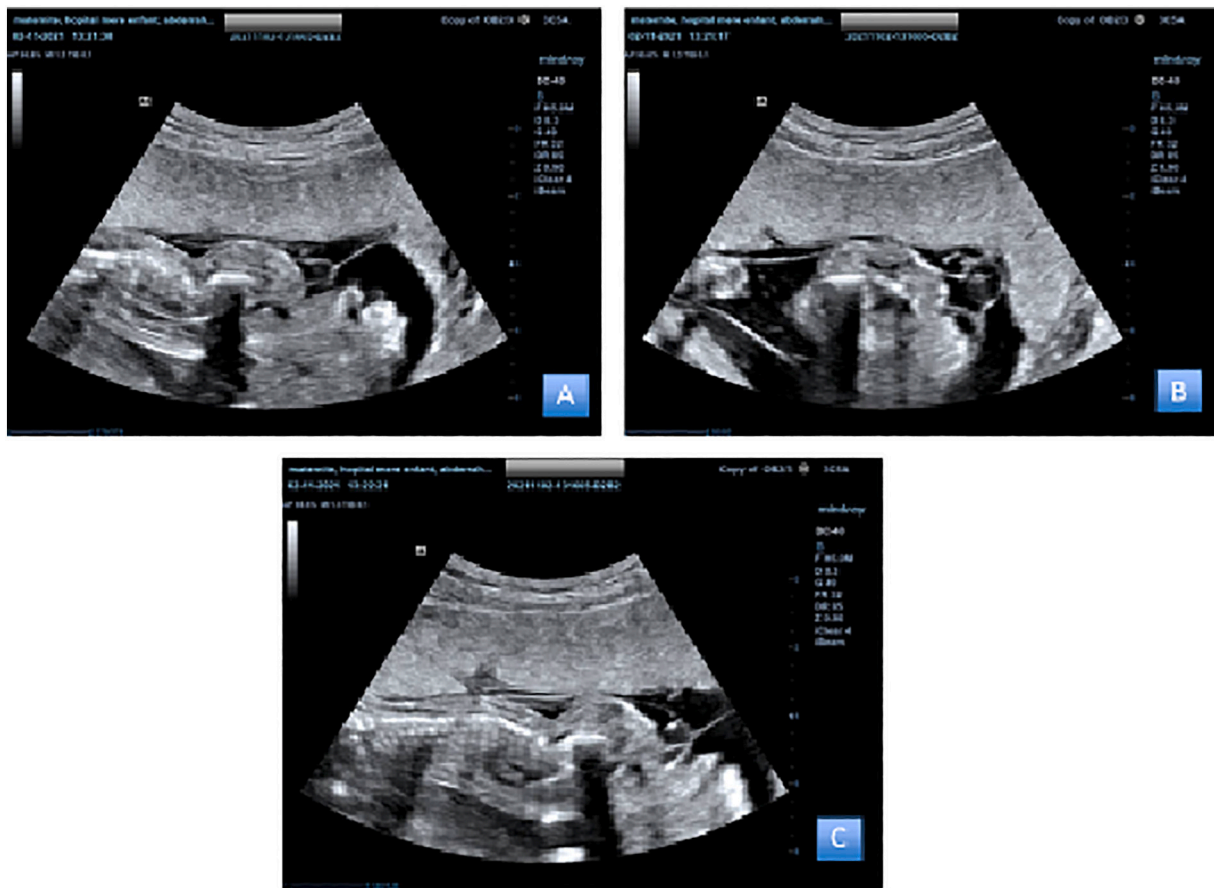


Fig. 1. Morphological ultrasound showing multiple spetations in the amniotic cavity (B) establishing the diagnostic of amniotic band syndrome associated with polymalformative syndrome. In figure 1 A and c we can see scoliosis and sacral agenesis.

oedema, atypical craniofacial anomalies without embryological systematization, coelosomies, and fenestrated syndactylies. The presence of an amniotic membrane in contact with these injuries is not essential for the diagnosis. In our case, the diagnosis was made following an ultrasound scan at 14 weeks' gestation. The contribution of three-dimensional imaging should improve the diagnosis and the use of pulsed Doppler should make it possible to monitor the evolution of vascular flow downstream of a stricture groove [8]. Fetal CT is a second choice examination to support antenatal diagnosis, allowing accurate analysis of the fetal skeleton [8]. The karyotype is always normal in amniotic band syndrome and should be performed as part of the differential diagnosis. Fetopathological examination confirms the diagnosis. The prognosis of amniotic band disease depends on the severity and the site of the malformations, medical abortion should be proposed in the presence of severe craniofacial and visceral malformations, whereas isolated limb malformations are amenable to surgical treatment. Furthermore, the obstetrical prognosis of patients does not seem to be altered compared to the general population [9–11]. In our case, the vaginal delivery was adopted and was uneventful.

4. Conclusion

Amniotic band disease is a rare embryo-fetopathy. Antenatal diagnosis is possible with pregnancy monitoring and early first trimester ultrasound. The obstetrical attitude should be individually adapted. Medical abortion may be justified in case of seriousness and lethality of malformations, with the couple's consent, considering the socio-cultural and religious context.

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Ethical approval

I declare on my honor that the ethical approval has been exempted by my establishment.

Consent

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

Author contribution

Lamrissi Amine: Corresponding author writing the paper and operating surgeon

Mourabbih Mariam: writing the paper and operating surgeon

Oujjih Oumaima: writing the paper

Jalal Mohamed: study concept

Fichtali Karima: study concept

Bouhya Said: correction of the paper and operating surgeon

Registration of research studies

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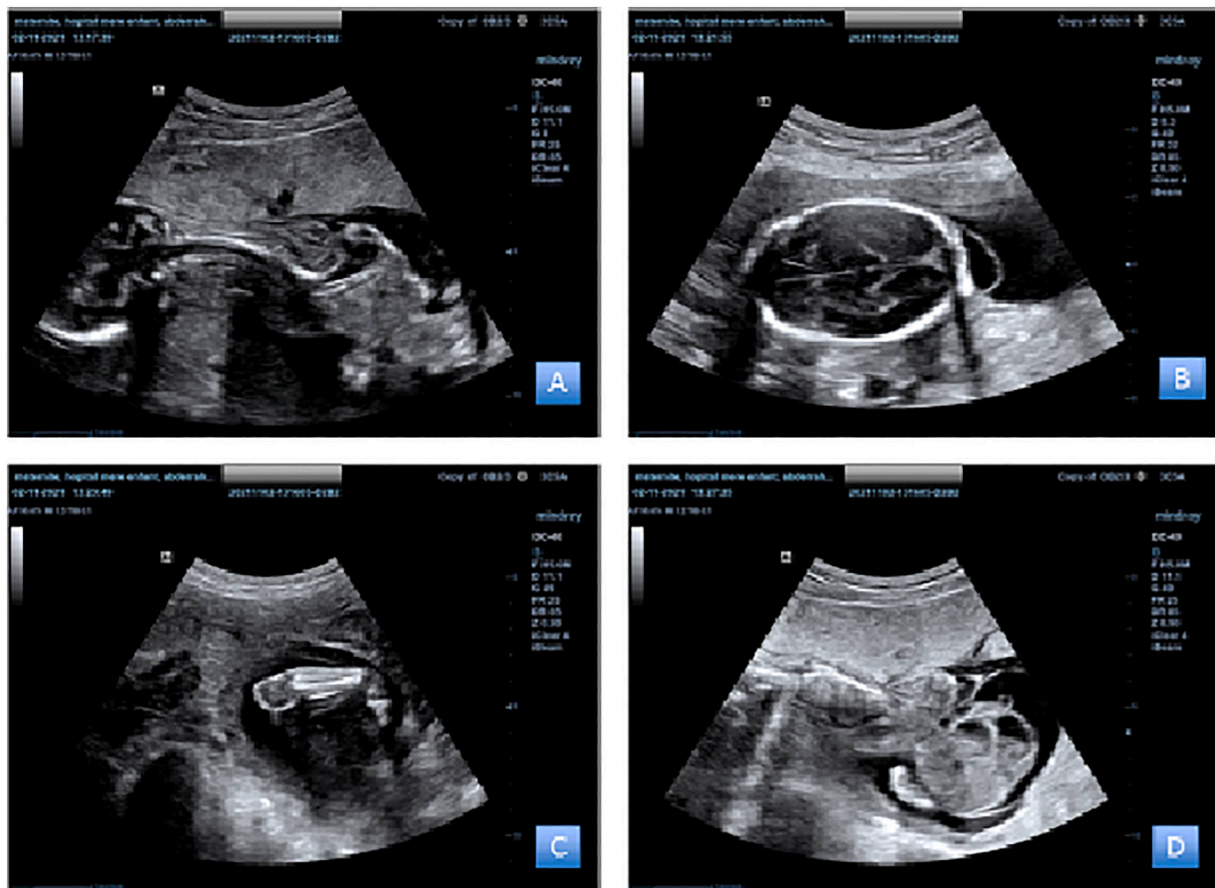


Fig. 2. Obstetrical supra- pubic gray scal ultrasounud showed complications of amniotic band syndrome. A: open myelomeningocele with ventriculomegaly, B encephalocele C: degeneration of extremities, D: huge omphalocele that contains all the intestines, liver and gastric pouch. sugging abdomen, with thor- acique hypoplasia.



Fig. 3. Post natal Macroscopic malformations of the Fetus showing adherent amniotic bands to the body and limbs of the fetus with absence of memebrial segments and omphalocele: limb body wall complex syndrome (LBWC).

Guarantor

Amine Lamrissi.

Declaration of competing interest

The authors declare having no conflicts of interest for this article.

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