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Amniotic Band Syndrome at the Van Norman Clinic in Burundi: A Case Series

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Abstract

Amniotic band syndrome is an acquired embryo-fetopathy. It is rare and is characterized by malformations mainly affecting the limbs but also the skull, face and thoraco-abdominal axis. Its etiopathogenesis remains poorly understood. Its diagnosis is essentially clinical and is classically based on the existence of signs such as furrows, amputations and pseudosyndactyly. To show the importance of antenatal diagnosis in resource-limited countries, we report the case of two newborns, one premature at 31 weeks and the other at term, in whom amniotic band syndrome was discovered incidentally at birth. It involved an amputation of the right leg for both cases. The premature baby was born in a context of neonatal sepsis and will succumb to the latter while the 2nd case was released from the hospital alive. Imaging examinations to search for probable congenital malformations could only be carried out for the 2nd case and no accessible congenital malformation had been identified. And as management of the disease, only psychological support to the parents was provided for the 2 cases. The antenatal discovery of a case of amniotic band syndrome in countries with low technical capacity such as Burundi should push clinicians to think in time about treatment options.

Keywords

Amniotic Band Syndrome, Embryo-Fetopathies, Antenatal Diagnosis, Limbs

1. Introduction

Amniotic band syndrome (ABS) is a relatively rare and sporadic condition of non-malformative disruptive origin, and questions about its management have

been raised for decades by the various practitioners involved in the care of these patients, namely obstetricians, geneticists, pediatricians and fetopathologists or pediatric surgeons [1]. It is characterized clinically by the association of pathognomonic signs: presence of flanges, cutaneous stricture furrows, amputations of the extremities and pseudosyndactyly. Also found are atypical facial clefts, club feet, exencephaly and lesions of the adnexa. Limb involvement is the most common. In cases of amniotic bridle associated with isolated limb constriction, antenatal resection of the bridle may be proposed to avoid the occurrence of in utero amputation [2].

In many countries with limited resources, monitoring pregnancies poses serious problems. The illiteracy of mothers, household poverty combined with limited access to quality health care and an insufficiently developed technical platform make antenatal diagnosis often inaccessible for a good number of embryofetopathies. In this context, there are a limited number of studies relating to embryofetopathies and no cases have yet been reported in Burundi. To show the importance of antenatal diagnosis in resource-limited countries, we report two cases, one of a premature newborn of 31 weeks and the other of a full-term newborn, in whom amniotic band syndrome with amputation of a limb in utero was discovered incidentally at birth.

2. Cases

2.1. Case 1

This is a newborn male, born prematurely at 31 weeks. He was the result of a third pregnancy poorly monitored with 2 antenatal consultations and an obstetric ultrasound done late at 30 weeks which did not show any fetal anomaly from a morphological point of view but which noted oligohydramnios. His mother, aged 28, unemployed, had no particular pathological history and even his father, aged 33, a trader, had no known pathological history. The mother's history does not reveal any notion of consanguinity or pathology during the pregnancy. Additionally, there was no history of taking toxic medications during pregnancy. He was born by cesarean section for severe oligohydramnios due to premature rupture of membranes lasting more than 12 hours. His birth weight was 1400 g and the Apgar score was normal. The physical examination revealed mild respiratory distress (Silverman score of 3). The right leg was amputated at the lower 1/3 with the wound on the stump still healing (**Figure 1**), (similar to images found in literature, **Figure 2** [3]). The patient's head circumference was 30 cm and his height was 40 cm. The family was shocked to see the newborn whose leg was amputated with a wound that had not yet completely healed. It was difficult for the care team to explain to the family that it was a malformation and not the result of witchcraft.

The laboratory assessment revealed a positive CRP at 12 mg/L and a normal blood count. The newborn was admitted to the neonatology department where he was treated for neonatal infection due to extreme prematurity and amniotic

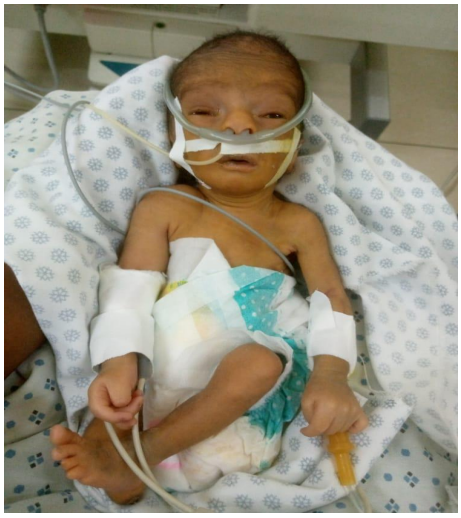

Figure 1. Jaundiced newborn with right leg amputated at the lower 1/3, stump visible under the left foot.

Figure 2. Right leg amputation.

band disease affecting the right leg. Antibiotic therapy based on cefotaxime (100 mg/kg/day in two doses), ampicillin (100 mg/kg/day in two doses) and gentamycin (5 mg/kg/day in one dose) was started. A dose of vitamin K1 (1 mg IM) was administered. He had also been put on aminophylline to prevent apnea in such a premature baby. He was also put on oxygen therapy (2 l/min) and a monitoring system was put in place.

Imaging examinations looking for anomalies and congenital malformations such as a chest X-ray, a cardiac ultrasound, an abdominal ultrasound and a transfontanellar ultrasound, although desired, could not be carried out.

After 3 days of hospitalization, the patient's condition worsened with worsening distress and the appearance of other signs such as fever, jaundice and petechiae. The laboratory assessment showed hyperbilirubinemia at 267 micromol per liter, an increased CRP at 96 mg/L. The newborn's condition continued to worsen despite adjustments in care. Antibiotic therapy was changed, the dose of cefotaxime was increased to 200 mg/kg/day, ampicillin was stopped and replaced by ciprofloxacin at 25 mg/kg/day. He had also been put on conventional photo-

therapy. The clinical signs have not improved. Respiratory distress worsened and other hemodynamic parameters continued to collapse. The newborn died on day 5 of life in a picture suggestive of severe sepsis.

2.2. Case 2

This was a full-term male baby, born vaginally from a poorly monitored pregnancy with 3 antenatal consultations without any obstetric ultrasound, born to a 32-year-old mother (gravidity of 3, Parity of 3, 3 living children) who was a farmer, and a father of 38 years old, farmer. No known pathological history in the parents was found in the anamnesis and there was no notion of consanguinity or pathology during pregnancy, nor of taking toxins during pregnancy. He was born vaginally at full term at the Van Norman Clinic with a weight of 3200 g. He had cried at birth. The physical examination did not find any abnormalities except the amputated right lower limb. The amputation took place on the upper 2/3 of the leg and there was no wound at the amputation site (**Figure 3**). A chest X-ray, cardiac ultrasound, abdominal ultrasound and transfontanellar ultrasound performed to look for anomalies and congenital malformations did not note any abnormalities. The parents received psychological support before the mother and her child left the maternity ward.

3. Discussion and Review of Literature

Amniotic band syndrome (ABS) is a set of malformations ranging from constriction and lymphedema of the fingers to multiple congenital anomalies mainly affecting the limbs, but also the craniofacial region and the thoracoabdominal axis [2]. These malformations are asymmetrical, polymorphic, and do not respect any embryological systematization, they probably result from multiple different pathological processes (vascular, hemorrhagic origin, early rupture of the amnion, primary damage to the germinal disc) [2].

Amniotic band disease (ABD) is relatively rare, its incidence is between 1/1200 and 1/15,000 live births [2] [4] but this is in reality probably higher due to early lethal forms [1]. In developed countries some authors estimate its incidence

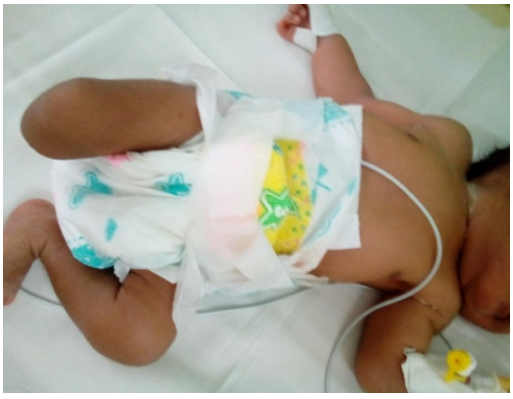

Figure 3. Full-term newborn with upper 2/3 amputated right leg with completely healed wound.

between 0.11/10,000 [5] and 1600 births [2]. There is no racial predisposition or link to sex [2] [4]. The incidence of ABD is difficult to specify due to the great diversity of its clinical presentations. Limb malformations are found in 65% of cases, and facial malformations are present in 48% of cases [6].

In Africa, its incidence is not known; reported cases come from hospital data [5] [7]; many cases are certainly not reported. In Burundi, no cases have yet been reported to our knowledge. This low reporting in countries with limited resources is not a sign of the rarity of cases of ABD but it is above all evidence of the lack of diagnostic means and also of the lack of knowledge of the pathology from a diagnostic and even therapeutic point of view. Antenatal screening for ABD should allow the clinician to plan care for the baby to be implemented from birth, taking into account the type of lesions present.

Malformations mainly affect the extremities but also the skull, face and thoraco-abdominal axis [2] [8]. For our two cases, the right lower limb was involved. It involved a complete amputation of part of the right leg.

The diagnosis of ABD must be antenatal, the craniofacial and thoraco-abdominal anomalies being able to be visualized from the first ultrasound at 10 - 12 weeks. Unfortunately, when it comes to distal limb involvement, ultrasound detection is uncommon [9] [10]. According to the two cases published in Morocco in 2019, the diagnosis was made on the 2nd trimester (respectively at 25 weeks + 04 days and 22 weeks) via obstetric ultrasound [11]. For Kisito Nagalo *et al.*, in Burkina Faso, for one in five cases the malformation even went unnoticed during obstetric ultrasound and was only discovered in early childhood [12]; For our two cases, the diagnosis was made at birth, even if in one case (the premature baby) the ultrasound had been carried out at 3rd quarter.

At birth, the diagnosis of ABS is purely clinical. It was the same for our two cases.

Among the 5 cases of ABS published in Burkina Faso in 2015, a case of premature birth at 32 weeks by cesarean section with indication of severe oligohydramnios with premature rupture of amniotic membranes is reported.

Which is close to the case of the premature baby presented in our two cases even if the malformations are not the same.

According to the classification of Levy *et al.* [6], our cases corresponded to ABD (Amniotic Band Disease) which has a better prognosis, characterized essentially by limb anomalies. There are malformations associated with ABS such as cleft lip and palate as described in one of the five cases in Burkina Faso [10]. For our cases, there were no other malformations observed on physical examination.

The prognosis of SBA depends on the severity of the malformations. In mild forms, surgical [5] [10] or prosthetic palliative treatment can allow the patient to have an optimal quality of life [11]. On the other hand, in the case of Limb Body Wall Complex syndrome (LBWC) which is the other form of ABS alongside ABD and which is beyond any therapeutic resource, death is inevitable either in utero or shortly after birth [8]. The three cases out of five published in the Bur-

kina Faso study are an example of this [12].

Medical termination of pregnancy is generally proposed in the presence of severe craniofacial and visceral malformations; isolated limb malformations are accessible to surgical treatment at birth. In cases of amniotic band associated with isolated constriction of the limb, antenatal resection of the bridle by fetoscopy may be proposed to avoid the occurrence of in utero amputation [10]. In most countries with limited resources where the diagnosis of isolated limb lesion can be made at birth and not during antenatal diagnosis as in the case of our patient released alive, surgical treatment remains possible. In these cases, collaboration between the maternity department and the orthopedic surgery department is necessary with a view to setting up a palliative care project before leaving the maternity ward.

4. Conclusion

Amniotic band syndrome is an acquired embryo-fetopathy which is rare. Its diagnosis is often made clinically at birth while an antenatal diagnosis is possible as well as in utero surgery if the disease affects the limbs, thus avoiding amputation in utero. But in developing countries including Burundi where the technical platform is lacking, it is important that the disease is known to practitioners. This would make it possible to adequately manage cases with their families through surgical or even prosthetic palliative treatment for children or psychological treatment for families as soon as the case is known.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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