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A RARE CASE OF AMNIOTIC BAND SYNDROME- A CASE REPORT

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ABSTRACT Amniotic Band Syndrome (ABS) happens when the amnion, a membrane around the embryo, breaks early in pregnancy. This causes the development of thin strands (amniotic bands) that can wrap around the embryo. If these bands tighten around body parts, they can cause constriction rings and, in severe cases, disrupt blood flow and lead to partial amputation. Even if the bands don't tighten, they can still cause malformations or deformations, like clefts. The exact cause of the amnion rupture is mostly unknown, though it has been linked to some maternal or fetal disorders like epidermolysis bullosa and connective tissue disorders. The term "pseudoamniotic band syndrome" is used when the rupture is caused by medical procedures, such as failed first-trimester curettage, fetoscopy, or septostomy for managing twin-twin transfusion syndrome.

KEYWORDS :

INTRODUCTION

Amniotic Band Syndrome (ABS) is a rare condition that happens during pregnancy. The inner lining of the amniotic sac, which surrounds the baby, is weak. This weakness creates fibrous bands or strings of tissue that can wrap around parts of the baby. These bands can cause tight rings around limbs or other body parts and, in severe cases, can even cut them off. ABS can sometimes lead to miscarriages. This condition affects both boys and girls equally and is slightly more common in people of Afro-Caribbean descent.

The first known case of ABS was described by a person named Portal in 1685. Later, in 1832, a researcher named Montgomery studied the causes of ABS. Research by Orioli and others found that ABS most often affects the arms and legs. Another study by Guzmán-Huerta and colleagues showed that 78% of ABS cases involved abnormalities of the face and head, while 70% involved problems with the limbs. The changes in the skull can be different, ranging from encephalocele, where brain tissue sticks out through a hole in the skull, to acrania, where parts or all of the skull are missing. Exencephaly is a type of acrania where the brain tissue is disorganized and comes out from the base of the skull.

Case Report

A 6-year-old boy was brought to the Pediatric Outpatient Department (OPD) by his parents. They were concerned about his persistent cough and runny nose. During the general examination, the medical team observed that the child had ring-like constriction deformities on the fingers of both hands. Additionally, some of his fingers were amputated, as shown in the accompanying images (1) and (2). Notably, there were no deformities observed in any other parts of his body.

The child's mother provided a detailed medical history. She mentioned that she and the child's father were related by blood, a consanguineous marriage. However, she reported no significant complications or unusual events during her pregnancy. She also confirmed that she did not take any medications known to cause birth defects (teratogenic medications) during her pregnancy.

The child was delivered via Cesarean section at full term. His birth weight was 2.6 kilograms, which is within the normal range for newborns. There were no significant issues or complications reported at the time of his birth. The deformities in his fingers were noticed immediately after birth. At that time, the parents were informed that these deformities were likely due to a weak uterus during pregnancy. No other deformities or abnormalities were observed in the child at birth.

Based on these observations and the child's medical history, the medical team diagnosed him with Amniotic Band Syndrome (ABS). This condition occurs when fibrous bands of tissue form within the amniotic sac and entangle parts of the developing fetus, leading to constrictions, deformities, or amputations of limbs and digits. In this child's case, the bands had caused constriction deformities and amputations of his fingers, but no other parts of his body were affected.

The detailed history provided by the mother, the absence of significant antenatal issues, the lack of teratogenic medication exposure, and the specific pattern of deformities observed all contributed to the diagnosis of ABS. The condition was identified as congenital, meaning it was present at birth, and it was determined that the deformities resulted from the formation of amniotic bands rather than any other underlying health issues or external factors. The diagnosis of ABS was confirmed, and the child was advised to receive appropriate follow-up care to monitor and manage his condition as he grows..

Figure 1

Figure 2

DISCUSSION

Amniotic Band Syndrome (ABS) consists of a group of birth defects that occur in 1% to 2% of all congenital anomalies in the general population. ABS is not hereditary, which means it does not run in families and there is no risk of it happening again in future pregnancies. Many theories have been proposed to explain how ABS develops. The most accepted theories are:

1. Extrinsic Theory: Proposed by Torpin in 1965, this theory suggests that the defects in the limbs or limb amputations are caused by the rupture of the amniotic sac. When the sac ruptures, fibrous bands form and compress the limbs of the developing baby.
2. Intrinsic Theory: Proposed by Streeter in 1930, this theory states that ABS results from a defect in the germplasm, which is the early tissue that forms the baby. This defect leads to problems with blood vessels and changes during the early stages of development, creating fibrous bands that cause limb damage.

Several genetic and environmental factors can cause ABS, including:
- Amniotic sac rupture
- Bacterial infection of the amniotic sac and membranes
- Abdominal trauma (injury to the mother's belly)
- Amniocentesis (a medical procedure where a needle is used to take a sample of amniotic fluid)
- Maternal exposure to certain drugs
- Oligohydramnios (low amniotic fluid levels)

One study by Higginbottom identified three ways that fibrous bands cause malformations:
1. Interruption of Morphogenesis: The normal development process is interrupted.
2. Deformation: Fetal parts are compressed by the bands or the baby itself, leading to malformations.
3. Injury: Already formed body parts, such as limbs, are injured by the bands, causing constrictions.

Another theory by Hunter et al. suggests that ABS results from a defect or deficiency in the ectoderm, the outer layer of the embryonic disc. Depending on where the defect occurs, different areas of the body can be affected. Craniofacial (head and face) defects are classified into four categories:
1. Exencephaly/Encephalocele with Amniotic Connections: Brain tissue protrudes through a skull opening without facial cleft.
2. Similar Cranial Lesions with Facial Clefts: May or may not involve amniotic bands.
3. Abdominoschisis and Thoracoabdominoschisis: Defects in the abdominal and thoracic (chest) walls.
4. Limb Abnormality: Defects in the arms and legs.

Diagnosing ABS can be challenging and is successful in only 29% to 50% of cases. Craniofacial deformities caused by ABS are often irregular, and abnormalities of internal organs are rare.Exencephaly, a severe form of cranial deformity, can be diagnosed by measuring the level of alpha-fetoprotein in the amniotic fluid. An ultrasound can also be used as early as the 11th week of pregnancy to detect the absence of bone coverage in the skull.The prognosis for ABS varies depending on the severity and location of the anomalies. In the case described, the child has only upper limb abnormalities and is otherwise healthy. However, ABS can cause severe complications, so when detected at birth, genetic studies should be performed, and parents should be informed about the potential risks and the possibility that the condition could be life-threatening.

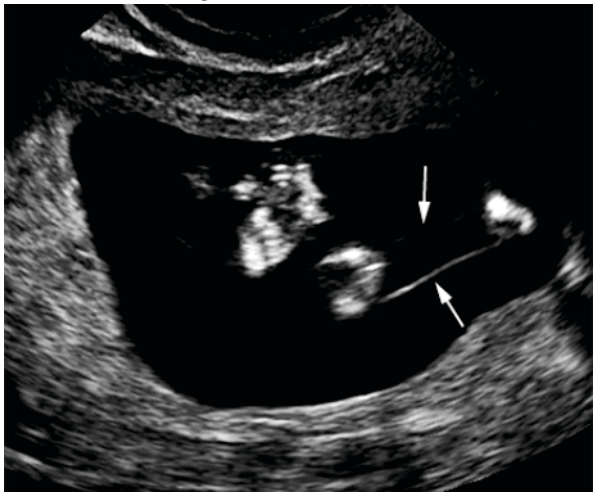

Fig 3

Image of the extremities shows amniotic bands (arrows) stretching between the extremities.

CONCLUSION

Amniotic Band Syndrome (ABS) is a rare condition, but it is important to check for it in every newborn who has congenital anomalies, especially if there are defects in the extremities such as fingers, hands, arms, toes, feet, or legs. Detecting ABS early is crucial because it can help guide the appropriate care and management for the affected child.Postnatal diagnosis of ABS typically begins with a thorough physical examination of the newborn. Doctors will look for the characteristic signs of ABS, such as ring-like constrictions, amputations, or other deformities in the limbs. If any abnormalities are found, additional examinations may be needed to check for potential malformations of internal organs, although these are rare in ABS cases.One important step in the diagnostic process is the analysis of the child's chromosomal karyotype. This test examines the baby's chromosomes to ensure there are no genetic abnormalities that might be causing the symptoms.

Conducting a karyotype analysis helps exclude other possible diagnoses and prevents the dissemination of incorrect information about the risk of recurrence to the parents. Since ABS is not hereditary, confirming the diagnosis through karyotype analysis reassures the parents that the condition is unlikely to occur in future pregnancies.Given the complexity of ABS, treatment and follow-up care require a multidisciplinary approach. A team of specialists, including pediatricians, geneticists, orthopedic surgeons, and physical therapists, should be involved in the care of the child. Each child with ABS may have different needs based on the severity and location of their deformities, so the treatment plan must be tailored to the individual patient.In some cases, surgical intervention may be necessary to release constrictions, improve functionality, or correct deformities. For example, surgery might be performed to separate fingers that are fused together or to improve the range of motion in affected limbs. Physical therapy can also play a significant role in helping the child develop strength and coordination in their limbs, as well as adapting to any physical limitations.Long-term follow-up is essential to monitor the child's progress and address any emerging issues. Regular check-ups with the medical team ensure that any complications or new symptoms are promptly managed. Additionally, psychological support for the child and family can be beneficial, as living with congenital anomalies can sometimes lead to emotional and social challenges.Parents should also receive comprehensive education and support to help them understand the condition and the care their child needs. This includes information on how to monitor for any changes or complications, guidance on physical therapy exercises, and advice on how to create a supportive environment for their child's development.Overall, early detection and a well-coordinated, individualized care plan are key to optimizing the outcomes for children with ABS. By addressing both the physical and emotional aspects of the condition, healthcare providers can help these children lead healthy and fulfilling lives despite their congenital anomalies.

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