

Amniotic band disruption complex: Case report and review

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ABSTRACT

Amniotic band sequence refers to a highly variable spectrum of congenital anomalies that occur in association with amniotic bands. The pathogenesis of amniotic band syndrome is not firmly established. Clinical features are variable and include amniotic band adhesion, placental adhesions, craniofacial clefting, amputation, ring constriction, thoraco-abdominoschisis and internal malformations. We report a newborn presented at birth with multiple constriction rings, partial amputation, syndactyly, cleft palate, malformed nose and bilateral micro-opthalmia. These findings were suggestive of amniotic band disruption complex.

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KEYWORDS

Constriction rings;
Dysmorphism;
Malformations;
Amniotic band
disruption complex.

INTRODUCTION

Amniotic band sequence is well-known. In newborns, it has been estimated to occur 1 in 1300–2000 births^[1]. It affects both sexes equally. There is variable spectrum of congenital anomalies that with amniotic bands^[2]. There is heterogeneity in clinical manifestations, etiology, pathogenesis, and recurrence risk. It was also known as amniotic band sequence, amniotic band syndrome, amniotic band disruption complex, limb body wall complex, body wall complex with limb defects, amniotic deformity, adhesion, and mutilation (ADAM) sequence.

CASE REPORT

A male newborn baby born to a non-consanguineous couple with emergency lower segment caesarian

section. Child was second in birth order. The first baby was a 3 year old male child alive and healthy. Mother was an unbooked case but she had attended a health care facility locally once. She took iron and folic acid post conceptionally. She has no history of teratogenic drug intake, radiation exposure and fever with rash. Quickening was started at 5 months of pregnancy. Level 1 USG was missed. USG just before delivery found multiple congenital anomalies.

Baby cried immediately but developed severe respiratory distress soon after birth and needed mechanical ventilation. On examination the baby had multiple constriction rings at leg and fingers [Figure 1]. The fingers of upper and lower limbs were partially amputated and fused [Figure 2,3]. The baby had sub mucosal cleft palate. There were malformed nares, tip and septum of nose [Figure 4]. The baby had bilateral micro-opthalmia



Figure 1



Figure 2



Figure 3



Figure 4

[Figure 4]. The majority number of cases those survived has constriction rings of variable severity, otherwise normal mental development and no obvious dysmorphism. But there were few case reports; those have mentioned the other features also. So the diagnosis with these additional features was amniotic band

disruption complex.

DISCUSSION

Amniotic band sequence is not much uncommon and refers to a variable spectrum of congenital anomalies. It is called a sequence because the pattern of con-

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genital anomalies results from a single defect that can be produced by a variety of different etiologies. In contrast, a syndrome refers to a pattern of congenital anomalies that are known, or at least assumed, to result from only a single etiology. The pathogenesis of both amniotic bands and ABS are not well established. There were two different theories. According to extrinsic theory, amniotic band syndrome occurs when the inner layer (amnion) of the amniotic sac ruptures, exposing the fetus to strands of fibrous tissue. These bands of tissue can disrupt the normal development of an embryo or fetus^[3]. The symptoms depend on the part of the body and how tightly it has wrapped by fibrous bands. If the amniotic bands are still partially attached to the amniotic sac, they may wrap around a fetal body part and tether (anchor) that body part to the amniotic sac. This can restrict movement and proper development of an embryo or fetus. The extrinsic theory fails to explain why there is an intact amniotic sac in some infants with amniotic band syndrome; why there are a high number of malformations affecting internal organs in some cases; and why some infants have defects of parts of the body not affected by constriction bands. The intrinsic theory attributes the development of amniotic band syndrome to vascular disruption^[4]. The exact, underlying cause of vascular disruption is not known. In areas of poor blood flow injury occurs to the blood vessel walls of the fetus. This leads to bleeding and tissue loss in the affected areas, which in turn results in the varied symptoms associated with the disorder.

The symptoms are variable associated with amniotic band syndrome. The most severe complications occur when amniotic band syndrome develops early in the first trimester. Several different patterns have been identified with amniotic band syndrome. The three most common patterns are the limb-body wall complex, craniofacial abnormalities and neural tube defects complex.

Most infants with amniotic band syndrome have some deformity of the arms and legs or fingers and toes. Upper limbs are affected more commonly than lower limbs. Certain fingers or toes may have partial or total amputation and syndactyly. The constriction bands can cause depressions or grooves but do not enough to damage the limb or digit. Therefore, despite the presence of this band or ring of tissue, the distal portion of the limb or digit retains its normal size and function. In some cases, strands of tissue are attached to the ends

of the fingers.

In limb body wall complex affected infants have defects of the abdominal wall that occur in association with defects of the arms and legs and other abnormalities^[5]. Infants usually have protrusion of a portion of the brain through the skull defect (encephalocele), facial clefts, protrusion of the viscera (internal organs abdominal or chest cavities) through a fissure in the abdominal wall (abdominoschisis) or the chest wall (thoracoschisis), and a variety of defects affecting the arms and legs. Additional abnormalities can occur in infants with limb body wall complex. Amniotic band syndrome involves craniofacial abnormalities such as cleft palate, cleft lip, facial clefts, micro-ophthalmia, choanal atresia and malformations of the skull. In severe cases, neural tube defects such as anencephaly may occur. Craniofacial abnormalities and neural tube defects occur along with serious complications of the arms and legs called terminal transverse limb deficiencies. In such cases, affected infants may be missing a portion or all of a limb, ranging from one finger or toe to an entire arm or leg.

Amniotic band syndrome is often difficult to detect before birth as the individual strands are small and hard to see on ultrasound. Often the bands are detected indirectly because of the constrictions and swelling upon limbs, digits, etc. Misdiagnosis is also common, so if there are any signs of amniotic bands, further detailed ultrasound tests should be done to assess the severity. 3D ultrasound and MRI can be used for more detailed and accurate diagnosis of bands and the resulting damage/danger to the fetus.

Treatment In rare cases, if diagnosed in utero, fetal surgery may be considered to save a limb which is in danger of amputation or other deformity. This typically would not be attempted if neither vital organs nor the umbilical cord were affected. This operation has been successfully performed on fetuses as young as 22 weeks.

Treatment usually occurs after birth and where plastic and reconstructive surgery is considered to treat the resulting deformity^[6]. Physical and occupational therapy may be needed long term.

Prosthetics may help some ABS sufferers to live more functional lives. The prognosis depends on the location and severity of the constricting bands.

Prevention Amniotic band syndrome is considered

an accidental event and it does not appear to be genetic or hereditary, so the likelihood of it occurring in another pregnancy is remote.

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