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Amniotic Band Syndrome with Amputation of Digits and CTEV: A Case Report

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ABSTRACT

Amniotic band syndrome (ABS) is defined as a continuous spectrum of manifestations due to intrauterine rupture of amnion range from simple soft tissue constrictive bands to amputation of digits or more severely the whole limb due to dysplastic vasculature. Therefore, we report a case with the objective of accenting the subsistence of ABS in Tamilnadu as throwing light on the prenatal diagnostic challenges and the socioeconomic problems faced by families living with physical disabilities.

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Introduction

Amniotic band syndrome (ABS) is a set of congenital malformations attributed to amniotic bands that entangle fetal parts during intrauterine life, which results in a broad spectrum of anatomic disturbances ranging from ring like constrictive bands, either in upper or in the lower limbs, occasionally in the trunk and lymphedema of the digits to complex, bizarre multiple congenital anomalies incompatible with life¹. Abnormalities in growth lead to amputation of extremities may be observed in severe cases. Thus the prognosis of the disease may vary from isolated digital amputation to severe lethal anomalies of the central nervous system, face and viscera².

Amniotic band syndrome may be detected by ultrasonography with the observation of asymmetrical limb deformities or defects and visualization of the amniotic membranes wrapping around foetal portions. The most common defect is constriction bands around foetal extremities. Management of the disease may change from expectant management to the termination of pregnancy, according to the severity of disease. Spontaneous resolution of amniotic bands was also described²⁻⁴.

Results of amnion rupture are external so no internal anomalies are associated⁵. Associated defects include hydra-anencephaly, porencephaly, cranio-facial abnormalities and spinal dysraphism⁶. Most cases are sporadic with no recurrence in siblings of affected patients⁷. The Prevalence is estimated to be about 7.7/10,000 live births⁸ and as high as 178/10,000 among abortuses⁹. ABS occurs in 77% of fetuses with multiple anomalies. Males and females are equally affected⁷.

Case Report

A 27 year old primigravida presented with 5 months amenorrhea with spontaneous

conception in the first year of married life. No history of fever with rashes, trauma, irradiation, teratogenic drug intake. Routine ultrasound examination showed a single live intrauterine pregnancy of 19 weeks gestation in breech presentation with posterior placenta previa (Figure 1). No obvious fetal malformations. There was a thin membranous band seen extending from the anterior surface of placenta in fundal region extending to the opposite uterine wall. So, differential diagnosis of ABS or uterine septum was made. The patient was lost to follow up.

She presented with PPROM and labor pain at 36 weeks gestation and had a normal vaginal delivery of a girl baby of weight 2.34 kg with normal APGAR 8/9, Baby was found to have an amputation of second to fifth digits at the metacarpophalangeal joint (MCP) with lymphedema and normal thumb in right hand. Left hand showed club hand with normal thumb, amputation of second digit at the interphalangeal joint (Figure 2) and constriction band in the MCP joint with amputation of other digits at MCP joint (Figure 3). Right leg showed congenital talipes equino varus (Figure 4). The left leg was normal. No other obvious anomalies seen.

Discussion

Incidence of ABS is estimated to a wide range of 1:1200 - 1:15000 live births⁸,¹⁰ in 1: 70 in stillborns¹¹ and among abortuses as high as 178:10000. Among a total of 3% major malformations in the general population, ABS is responsible for 1-2%^{12,13}. The visualization of the amniotic band is considered diagnostic of ABS. This precludes the need for further etiological investigations, such as karyotyping and genetic studies. There are various theories attempting to explain the pathogenesis of ABS. Some study suggested that once the

amnion is ruptured, the fetus lies outside the amniotic cavity and bands extending from the chorionic side of the cavity entrap various parts of the fetus and disturb normal development⁹.

According to Pattersons diagnostic criteria for ABS, there should be the presence of at least one of the following: 1) simple ring constrictions, 2) ring constrictions with distal deformity plus or minus lymph edema, 3) ring constrictions accompanied by syndactyly or acrosyndactyly, 4) amputation¹⁴.

A case of a one year old male who had constrictive circumferential grooves just above his left ankle with edema distal to the constriction was also reported¹⁵. Surgical excision of the fibrous bands on the leg and foot resulted in subsequent normal development of the extremity distal to this band.

The severity depends upon the period of gestation during which the bands develop, especially when accompanied with oligohydramnios¹⁶. The most frequent organs involved in ABS are the fingers and toes, with or without association with cleft lip and palate. Feet abnormalities such as club feet and finger abnormalities such as syndactyly, cranial, cardiac, abdominal wall defect and abdominal organs exstrophy, chest wall defect with heart exstrophy are reported¹⁰. Etiological associations have been reported with abdominal trauma, intrauterine devices, cerclage and chorionic villous sampling, however, none is confirmed. ABS can be diagnosed prenatally by ultrasound, which can sometimes show amniotic bands, but more often malformations consistent with ABS, as well as oligoamnios and reduction of foetal movements¹⁷. ABS can be diagnosed as early as 12 gestational weeks¹⁸; in the second trimester of gestation, most of ABS defects could be seen during routine ultrasound examinations¹⁷. The most

important ultrasound diagnostic criteria are visible amniotic bands, constriction rings on extremities and irregular amputations of fingers and/or toes with terminal syndactyly. Mild defects, however, are less likely to be diagnosed prenatally, in which case defects are seen after birth¹⁸. Latest ultrasound techniques including three dimensional and four dimensional ultrasound contribute to more sensitive prenatal diagnostics of ABS, and in complicated cases fetal magnetic resonance can be helpful. Placenta and amnion examination after the delivery should be an obligatory part of the newborns health evaluation because it can show presence of amniotic bands, among other things. Physical examination is the main way of postnatal diagnostic of ABS, with usage of addition, searches in order to establish potential malformations of different organs and body parts: ultrasound, echocardiography, X-ray. The treatment of ABS is mostly surgical, with an individual approach to every single case. Interdisciplinary consulting and work is very often needed (plastic surgeon, orthopedic surgeon, orthodontist, ophthalmologist, neurosurgeon). Lately, there have been some attempts of prenatal ABS treatment - fetoscopic laser cutting of amniotic bands, before their compression on the fetus makes malformations¹⁹.

There is a need for improvement of the diagnostic capabilities of prenatal USS. Families with disabilities resulting from ABS need to be supported to reduce the economic and social consequences. This type of case reports revealed large gaps in service provision for people with disabilities with unmet needs for welfare, assistive devices, education, vocational training and counseling services. Thus the authors suggested performing differential diagnostic including amniotic folds, short umbilical cord syndrome and extra-amniotic pregnancy. Amniotic folds which are

recognized by prenatal ultrasonography as reflecting membranes floating freely in the amniotic fluid are considered as a major clue to determine the issue.

Ethical committee approval

Not Applicable.

Informed consent

Informed consent was received from the parents.

Author's contribution

Both the authors contributed equally during the preparation of this manuscript.

Conflict of interest

No conflict of interest was declared by the authors.

Financial disclosure

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bands. *Ultrasound Obstet Gynecol* 1997; 10(5):316-20.



Figure 1. Amniotic band UCG



Figure 2. Amniotic band UCG



Figure 3. LT digit constriction band



Figure 4. CTEV