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Case Report

Amniotic band syndrome: a diagnostic dilemma

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ABSTRACT

Amniotic band syndrome can cause a broad spectrum of anomalies ranging from simple band constrictions to major craniofacial and visceral defects. It occurs when the amnion is damaged during pregnancy. Thin strands of tissue called amniotic bands form inside the amnion and can tangle around the developing fetus, restricting blood flow, thus affecting the growth of certain body parts. This can cause congenital deformities of limbs, and if vital organs are involved, it can result in significant neonatal morbidity. A 25-year-old primigravida was referred for fetal karyotyping as the targeted anomaly scan revealed bilateral limb defects along with a single umbilical artery. The right foot was reported as lymphedema, and the left foot showed clubfoot deformity. Here, we report a case of amniotic band syndrome, which was missed for “Milroy’s primary congenital lymphedema” in antenatal sonography but was revealed in fetal autopsy. Mostly, amniotic band syndrome is diagnosed after birth, but with the advancement of ultrasound, prenatal diagnosis is possible. Also, three-dimensional ultrasound and magnetic resonance imaging can be used for a more detailed and accurate diagnosis. Accurate diagnosis is very important for further management of the pregnancy and in counselling regarding recurrences in future pregnancies.

Keywords: Amniotic band syndrome, Antenatal sonography, Band constrictions, Fetal autopsy

INTRODUCTION

Amniotic band syndrome (ABS) is a congenital disorder caused by the entrapment of fetal parts in fibrous amniotic bands while in utero. ABS is an underappreciated cause of fetal and neonatal mortality and morbidity.¹ Synonyms for ABS include amniotic band disruption complex, constriction band syndrome, and amniotic deformity, adhesions, mutilations (ADAM) complex.² The spectrum of anomalies depends on which part gets entrapped and at what point of gestation. Hence, the clinical presentation can be extremely variable. Early detection of such cases using ultrasonography is challenging due to the small size of the fibrotic bands. A prenatal ultrasound scan can help indirectly show swelling of digits or limbs distal to the constriction.³ Three-dimensional ultrasound and magnetic resonance imaging can be used for a more detailed and accurate diagnosis when there is suspicion of amniotic

band syndrome. Here, we present a case of amniotic band syndrome in a fetus at 20 weeks of gestation, which was referred for amniocentesis and genetic analysis for limb defects noted in sonography. It is a non-recurrent cause of fetal malformations involving the limbs, craniofacial region, and trunk. Prenatal diagnosis is important for counselling and managing the present pregnancy. Here, we report a case of ABS, which was missed in sonography but was noted on autopsy.

CASE REPORT

A 25-year-old primigravida was referred to our institute for fetal karyotyping as the targeted anomaly scan revealed bilateral limb defects along with a single umbilical artery. The right foot was reported as lymphedema, and the left foot showed clubfoot deformity (Figure 1).

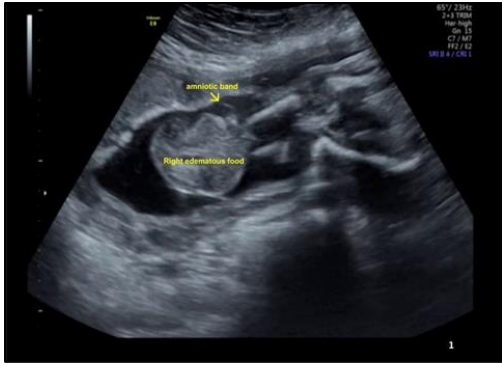


Figure 1: USG image showing right edematous foot with amniotic band attached to the lower leg.



Figure 2: Right edematous foot with amniotic band attached to the lower leg and left foot showing club foot deformity with syndactyly of 2nd and 3rd toe with a band attached to it.

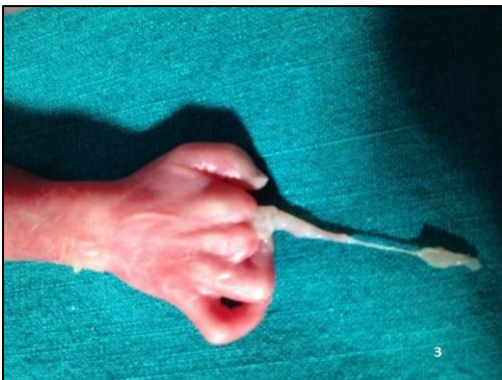


Figure 3: Left hand showing band attached to the amputated index and middle fingers.

It was suspected as AV malformation and/or Milroy's primary congenital lymphedema. There was only one umbilical artery noted. After counselling, the couple opted for termination due to the morbidity. The pregnancy was terminated, and the fetus was subjected to genetic testing and autopsy with the parents' consent. The fetus weighed 450 gm. The right foot was grossly edematous, and a band was seen attached to the lower end of the leg [Figure 2]. The left foot showed clubfoot deformity with syndactyly,

and a band was seen attached to the third toe. A band was also seen attached to the fingers with distal agenesis of the 2nd and 3rd fingers (Figure 3). The features were suggestive of ABS. Retrospectively, the band-like structure was appreciated on a careful search of the ultrasonographic video (Figure 1). No other abnormalities were observed on autopsy. The band-like structure was sent for histopathological examination, which resembled amniotic membrane histopathologically. The fetal karyotype was normal. Amniotic band syndrome can cause malformations that range from mild deformities to severe anomalies that are incompatible with life. ABS is difficult to diagnose, and its incidence varies widely from 1 in 1200 to 1 in 15000 live births and is responsible for 178 in 10,000 miscarriages.³

Though different theories have been proposed for ABS, the exact pathogenesis is not well known. The exogenous theory suggests the disruption of the amnion, and the endogenous theory suggests vascular compromise as the etiological factor. Genetic predisposition has been suggested by some authors but without any conclusive evidence.⁴ Other risk factors shown by various studies are inhabitants of high altitudes where hypoxia might play a role. Vasculopathy seen in diabetes adds to the risk of vascular disruption seen in amniotic band syndrome. Obesity and diabetes are possibly causes of early amnion rupture.⁵ The deformities seen in ultrasound are head/face entrapment like acrania, anencephaly, and facial clefts; truncal entrapment like abdominal defects, rib clefting, and congenital scoliosis; and extremity entrapment: variable levels of limb amputation, variable levels of limb constriction, lymphedema, pseudosyndactyly, and phocomelia.⁶ The constriction of appendages by amniotic bands may result in constriction rings around the digits, arms, and legs. Swelling of the extremities distal to the point of constriction (congenital lymphedema); amputation of digits, arms, and legs (congenital amputation).⁷

Milroy's primary congenital lymphedema (PCL) (hereditary lymphedema type I, Milroy disease) is present at birth and mostly affects the dorsal aspects of feet. PCL is an autosomal dominant disease with incomplete penetrance due to a mutation in the gene locus encoding for VEGFR3 with resultant dysgenesis of microlymphatic vessels.⁸ The swelling varies in degree and distribution. Limb swelling of all extremities may occur, while bilateral but asymmetric lower-limb lymphedema is most typical. Other features include prominent veins, cellulitis, hydrocele, papillomatosis, and typical up slanting 'ski-jump' toenails. In antenatal sonography, it appears as an edematous foot, but clubfoot deformity or amputation of digits have not been reported. 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. All the limbs should be evaluated thoroughly to find out if there is any kind of deformity like syndactyly or oligodactyly or other minor digit anomalies, which could be an important clue to support the diagnosis of ABS. 3D ultrasound and MRI can be used for a more detailed and accurate diagnosis of bands and the resulting damage/danger to the fetus, karyotype to be done to rule out the differential diagnosis.⁹ Color Doppler should be done to differentiate this condition from lymphatic and vascular complications.¹⁰

CONCLUSION

The amnion, which is a protective structure of the fetus, becomes destructive in this syndrome. The present report highlights that this syndrome, though rare, should be kept in mind during antenatal scanning when a lower limb deformity is noted in the form of an edematous foot. Deformities in ABS can mimic genetic syndromes; therefore, an accurate diagnosis is important to counsel the couple regarding the low risk of recurrence in future pregnancies.

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