

Rare association of Amniotic Band Sequence (ABS) with meningocele

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Abstract

Amniotic Band Syndrome also known as amniotic band sequence (ABS) is a group of congenital birth defects caused by entrapment of fetal parts in fibrous amniotic bands while in utero. Other synonyms are ADAM complex, Congenital constriction bands and Pseudoainhum. Defects usually involve longer central digits of the hand (middle, long, and index fingers). Up to 50% of cases have other congenital anomalies including cleft lip, cleft palate, and clubfoot deformity. Here we present a rare case of ABS with lumbar meningocele.

Keywords: Amniotic band sequence, pseudoainhum, meningocele.

1. Introduction

Amniotic band syndrome occurs due to a partial rupture of the amniotic sac[1]. This rupture involves only the amnion; the chorion remains intact. Fibrous bands of the ruptured amnion float in the amniotic fluid. Accidentally it may start constricting longer central digits of hand/foot. This constriction reduces blood circulation, hence causes congenital abnormalities. In some cases a complete "natural" amputation may occur. They are usually full-term; in most cases, the pregnancies were uncomplicated. Amniotic banding affects approximately 1 in 1,200 live births. The hands are affected in almost 90% of cases [2] mostly central fingers are affected. In rare cases, the thumb or small finger is involved. The same rule holds true for the feet, where constriction bands most commonly involve the hallux. In Light and Ogden's series, the average gestation was 37.5 weeks, with an average birth weight of 3.0 kg[3]. It has no risk factor for recurrence. A sonographic study by Barzilay et al suggested that prior uterine surgery may be a risk factor for ABS [4]. Less common findings associated with ABS are craniofacial abnormalities; complications from ABS include severe lymphatic or venous congestion at the time of birth due to tight bands. Sometimes neurovascular involvement may lead to necrosis and gangrene.[5]

2. Case Presentation

A three day old term male baby born by NVD of nonconsanguinous marriage presented to our SNCU with amniotic band sequence and meningocele mother was a

booked case, first order, on regular ANC visit with no history drug intake, no radiation exposure, TORCH screening negative. On examination baby of birth weight -2.4 kg, cried immediately after birth, alert, HR-140/min, RR-42/min, temp-37C, length-51cm, HC-34cm, cry, reflex, tone and activity was good. On head to toe examination there is constriction band at right wrist, amputation of central fingers of left hand [Figure 1A and 1B], and amputation of 2nd toe and band in other toes in left foot [Figure 2]. There is a lumbar meningocele [Figure 3]. On systemic examination no significant findings present. All routine investigations are normal. Otherwise child is active, alert & feeding well.

Figure 1: Examination of head to toe showing constriction band at right wrist, amputation of central fingers of left hand





Figure 2: Amputation of 2nd toe and band in other toes in left foot



Figure 3: Lumbar meningocele



2.1 Case Discussion

Amniotic band syndrome usually consists of three things

- 1) Constriction rings around the digits, arms and legs
- 2) Swelling of the extremities distal to the point of constriction (congenital lymphedema)
- 3) Amputation of digits, arms and legs (congenital amputation)

Other associated findings include the following: Encephalocele, Cleft lip or palate, Renal abnormalities, Cardiac defects, Hemihypertrophy, Anterolateral bowing of the tibia, Tibial pseudarthrosis Leg-length discrepancy.

Regarding etiology many theories are there. These are:

- Hippocrates suggested that extrinsic pressures from a ruptured amniotic membrane lead to the formation of bands or digital amputations. Montgomery [6] in 1832 and Simpson [7] in 1836 subsequently described series of amniotic band-associated deformities and discussed the differences between agenesis- and amniotic band-induced amputations. Streeter postulated a germ plasm defect as one plausible etiology. [8] Torpin, who examined many placentae and infants with the disorder [9]. In 1965, Torpin reintroduced the idea originally held by Hippocrates. He proposed that maternal trauma leads to rupture of the amniotic membrane. This is the most accepted one. After rupture, a transient oligohydramnios occurs due to extravasation of amniotic fluid. Clubfoot deformities seen with ABS because of very little space to move in. If the amniotic bands are swallowed while still partially attached to the placenta, the tether may lead to bizarre facial clefts and palate deficiencies.[10]

Extremity deformities in ABS are classified into Patterson's four types, as follows:

- Type I - Simple ring constriction
- Type II - Ring constriction accompanied by fusion of the distal bony parts, with or without lymphedema
- Type III - Ring constrictions accompanied by fusion of soft-tissue parts
- Type IV - Intrauterine amputations amniotic band syndrome diagnosed after doing a physical exam when your baby is born.

Then x ray is needed to see how a band affects deeper tissues. If a band is deep, MRI (magnetic resonance imaging) scan or other imaging scans to see how the band affects their blood vessels and nerves.

Sometimes, but not usually, doctors can diagnose amniotic band syndrome based on what they see during an Ultrasound before a baby is born.

Treatment varies widely. Often, the deformity is not severe and no treatment is needed. In more serious cases, major surgery may be needed to reconstruct all or part of an arm or leg.

In severe cases experts from Orthopedics, Occupational Therapy, Orthotics and Prosthetics, Plastic Surgery, Neurosurgery, Craniofacial and other areas of healthcare are needed for better functioning of child. Plans should be made for careful delivery and management of the problem after birth. Our child presented with amniotic band sequence with very rare association of lumbar meningocele. There was no neurological defect. In our patient manifestation is not so severe, so no treatment is needed patient is on regular follow up.

3. Conclusion

Amniotic band syndrome is an accidental event with no genetic relationship. There is no chance of recurrence in subsequent pregnancy. If we are getting any case of ABS first of all we have to search any associated finding, then imaging study including X ray and MRI to detect the depth of involvement. If constrictions are not so deep, no surgical treatment needed. Baby may need urgent surgery right after birth if bands press deeply on blood vessels or nerves, or if they cause serious swelling (lymphedema) or other problems. If not, we can wait until the baby is at least 6 months old.

Conflict of interest: Declared None.

References

- [1] Ross MG. Pathogenesis of amniotic band syndrome. *Am J Obstet Gynecol.* 2007 Aug;197 (2):219-20.
- [2] Light TR. Growth and development of the hand. Carter PR, ed. *Reconstruction of the Child's Hand.* Philadelphia: Lea & Febiger; 1991: 122.
- [3] Light TR, Ogden JA. Congenital constriction band syndrome. Pathophysiology and treatment. *Yale J Biol Med.* 1993 May-Jun. 66 (3): 143-55.
- [4] Barzilay E, Harel Y, Haas J, Berkenstadt M, Katorza E, Achiron R, et al. Prenatal diagnosis of amniotic band syndrome - risk factors and ultrasonic signs. *J Matern Fetal Neonatal Med.* 2015 Feb. 28 (3): 281-3.
- [5] Zych GA, Ballard A. Congenital band causing pseudarthrosis and impending gangrene of the leg. A case report with successful treatment. *J Bone Joint Surg Am.* 1983 Mar. 65 (3): 410-2.
- [6] Montgomery W. Spontaneous Amputation in Utero. *Dublin J Med Sci.* 1832; 2:49.
- [7] Simpson J. Essays on diseases of the placenta. *Dublin J Med Sci.* 1836; 10:220.
- [8] Streeter G. Focal deficiencies in fetal tissues and their relation to intrauterine amputations. *Contributions Embryol Carnegie Inst.* 1930; 22: 1-46.
- [9] Torpin R. Amniochorionic mesoblastic fibrous strings and amniotic bands: associated constricting fetal malformations or fetal death. *Am J Obstet Gynecol.* 1965 Jan 1. 91:65-75.
- [10] Bouguila J, Ben Khoud N, Ghrissi A, Bellalah Z, Belghith A, Landolsi E, et al. [Amniotic band syndrome and facial malformations]. *Rev Stomatol Chir Maxillofac.* 2007 Dec. 108 (6):526-9.