Case Report

Amniotic Band Syndrome (ABS) - A Rare Congenital Disorder

Mahmuda Hassan[1], Afsana Mukti[2], Hasibul Haque[3], Masuma Khan[4], Marium Begum[5], Sudipta Roy[6], Taslim Uddin Ahmed[7], Aashraf Uddin Ahmed[8], Sadia Islam[9], A R M Lutful Kabir[10], Hamidur Rahman[11], Zannatul Ferdous Sonia[12]

Abstract:

Amniotic band syndrome (ABS) is a rare congenital disorder that can lead to a wide range of physical abnormalities in the newborn infant.

In ABS 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 minor constriction rings and lymphedema of the digits alimbs to complex, bizarre multiple congenital anomalies incompatible with life. ABS is not seen very often, but should be considered in every newborn with congenital anomalies, especially defects of extremities and/or body walls. ABS can be diagnosed prenatally by ultrasound; otherwise, the defects are seen after birth. A team of specialists should be included in the treatment and follow-up of children with ABS, according to individual needs of every patient. Earlier surgical intervention is must for proper growth & development of child.

Keywords: Amniotic band syndrome, Limb reduction defects,

Introduction

Amniotic band syndrome (ABS) is a rare congenital disorder that is associated with a wide range of physical abnormalities[1] in the newborn infant, some of which are significantly disabling and disfiguring in nature. The commonest abnormalities usually involve the limbs and can range from simple construction rings to complete amputation(congenital amputation) occurring at various levels [2]. Abdominal wall defects and abnormalities of the cranio-facial region such as cleft lip and cleft palate are also associated with ABS [2] while in the more complex cases, visceral defects such as renal agenesis [3] and rarely septo-optic dysplasia [4] are also known to occur. Various studies estimate the incidence of ABS to be between 1 in 1300 to 1 in 15000 though, [4] 1: 70 instillborns[5]and amongabortuses as high as 178:10000[6]. Among total of 3% major congenitalmalformations in general population, [7] ABS isresponsible for 1-2%[8].

1. Professor, Department of paediatrics, AWMC
2. Assistant Professor, Department of paediatrics, AWMC
3. Associate Professor, Department of paediatrics, AWMC
4. Assistant Professor, Department of paediatrics, AWMC
5. Professor, Department of paediatrics, ABMC
6. Assistant Professor, Department of paediatrics, AWMC
7. Professor, Department of paediatrics, AWMC
8. Professor, Department of paediatrics, AWMC
9. IMO, Professor, Department of paediatrics, ShSMC
10. Professor, Department of paediatrics, AWMC
11. Professor, Department of paediatrics, AWMC
12. Assistant Professor, Department of paediatrics, AWMC

Correspondence: Dr. Mahmuda Hassan, Professor, Department of paediatrics, AWMC

Case Report:

A 35 weaker male neonate, weighing 2070gms, 1st issue of non-consanguineous parents was delivered by caesarean section as a result of non-progressive labour and breech presentation. The baby cried immediately after delivery. Age of the mother was 16 years. The pregnancy had been uneventful and there was no remarkable family history. Baby was delivered by lower uterine caesarian section. Physical examination at birth showed several constriction rings around right leg, 2 deep circumferential groove was present 1.5 cm and 3 cm above bothankle joint.(Figure 2) Also the left distal phalanges(4th and 5th) were lost . Amputation of the left thumb, also partial loss of the lefttoes with syndactyly of the 1st and 2nd, 4th and 5th toes. Therewere no signs of

Fig.-1: Consitriction groove above bothankle joints

Fig.-2: Distal phalanges malformations

Fig.-3: Righthand-congenital amputation of ring andsmall finger with hyperemia

limb perfusion abnormalities andthere was no neurological deficit (motor). Also a small ball like remnants of distal 4th left digit attached with a peduncle. On clinical basis diagnosis of amniotic band syndrome was present. Ultrasounds, echocardiography, X-ray were normal.

Discussion:

Amniotic band syndrome is a rare disorder. It often results in congenital physical defects in the infant which are disabling and disfiguring.

ABS occurs when the inner membrane of the amniotic sac tears and wraps around the developing baby and causes problems in the limbs, clefts in the face and band marks in different areas of the body. [9] The etiology is unknown. There have been reports associating amniotic band syndrome with maternal trauma, oophorectomy during pregnancy[10]intrauterine contraceptive device and amniocentesis.[11]

There are case reports in families with connective tissue disorders like lerDanlos syndrome. [12,13]

Amniotic band syndrome has very polymorphic clinicalfindings. Early amniotic rupture, during first 45 days, leadsto most severe cranio-facial and visceralmalformations. [12] Every part of the fetal body can bedamaged, but usually the extremities, especially upperextremities are affected. Most often there are minordefects, such as constriction rings or digit amputations[14]. Abnormalities of the extremities can be expressedin several ways: constriction rings of the soft tissueaccompanied by distal edema, shortening of the limb orintrauterine limb amputation, amputation of the digits(most often II, III and IV finger) and toes, syndactyly,hypoplasia of the digits, foot deformities,pseudoarthrosis, peripheral nerve palsy[9]. Our case had constriction rings on left leg and amputation of all toes and amputation of right 4th and 5th finger,syndactyly of 3rd and 4th right fingers with complete amputation of the right thumb. If bandscompress the fetal head or face, different cranio-facialdisturbances appear – asymmetric face clefts, orbitaldefects (anophtalmos, microphtalmos, enophtalmos),corneal abnormalities, central nervous system

Malformations (anencephaly, encephalocoele, asymmetric meningocoele), calvaria defect[9]. Amnioticbands can also cause abdominal wall defect andabdominal organs extrophy[9], chest wall defect withheart extrophy[15], umbilical cord strangulation with oftenlethal outcome[9]. Our case did not have these types of malformations. Amniotic rupture and consecutive olygoamnion by mechanical pressure on the fetus can cause deformities such as metatarsovarus, scoliosis[12] or hip dislocation[9]. Because of such a wide spectrumof possible anomalies and many combinations of theirsimultaneous appearance, there are no two identical cases of ABS. Beside all previously mentionedmalformations caused by amniotic bands itself, a subset of cases manifest additional findings that are notconsistent with that mechanism, such as congenital heartdefects, renal anomalies, hemangiomas, imperforate anus,polydactyly, septo-optic dysplasia, typical cleft lip andpalate[16]. ABS can be diagnosed prenatally by ultrasound, whichcan sometimes show amniotic bands, but more oftenmalformations consistent with ABS, as well asolygoamnion and reduction of foetal movements[17]. The most important ultrasound diagnostic criteria revisable amniotic bands, constriction rings on extremitiesand irregular

amputations of fingers and/or toes withterminal syndactily. Latest ultrasound techniques–three-dimensional and four-dimensional ultrasoundcontribute to more sensitive prenatal diagnostics of ABS and in complicated cases foetal magnetic resonance canbe helpful[18]. Placenta and amnion examination afterthe delivery should be obligatory part of the newborns health evaluation because it can show presence ofamniotic bands, among other things[18]. Physicalexamination is the main stay of postnatal diagnosis of ABS. However use of additional investigations like Ultrasound, echocardiography, X-ray is important inorder to establish potential malformations of different organs and body parts. ABS must be considered indifferential diagnosis of all complex or asymmetricmalformations, especially those on extremities, face andbody walls. ABS should be differentiated from the wholespectra of symmetric fusion defects of middle body line[19]. Therapy of ABS is mostly surgical, with an individualapproach to every single case. Interdisciplinary consultingand work is very often needed (plastic surgeon,orthopedic surgeon, orthodontist, ophthalmologist, neurosurgeon.[9]. Lately, there have been someattempts of prenatal ABS treatment - foetoscopiclasercutting of amniotic bands, before their compression onthe fetus makes malformations[20]. In cases when foetalanomalies incompatible with life are prenatalyseen,pregnancy termination is advised. Surgery is not needed for shallow constriction bands that are not circumferentialand without distal swelling. Distal edema or impairment of neurovascular function requires staged constriction band excision, Z-plasty or W-plasty. Multiple plastic surgical procedures are required for corrections of the complex craniofacial abnormalities.

Conclusion:

ABS is not seen very often, but should be considered inevery newborn with congenital anomalies, especiallydefects of extremities and/or body walls.

References:

1. Nardozza LMM, Araujo E, Caetano ACR, Moron AF. Prenatal Diagnosis of Amniotic Band Syndrome in the Third Trimester of Pregnancy using 3D Ultrasound. J Clin Imaging Sci [Internet]. India: Medknow Publications & Media Pvt Ltd; 2012;2:22.
2. Cignini P, Giorlandino C, Padula F, Dugo N, Cafà EV, Spata A. Epidemiology and risk factors of amniotic band syndrome, or ADAM sequence. J Prenat Med. CIC EdizioniInternazionali. 2012;6(4):59–63.
3. Van Allen MI, Curry C, Walden CE, Gallagher L, Patten RM. Limb-body wall complex: II. Limb and spine defects. Am J Med Genet. 1987;28(3):549–65.
4. Jabor MA, Cronin ED. Bilateral cleft lip and palate and limb deformities: a presentation of amniotic band sequence? J Craniofac Surg. 2000;11(4):388-93.
5. Ross MG. Pathogenesis of amniotic band syndrome. American Journal of Obstetrics and Gynecology 2007; 197(2):219-20.
6. Burk CJ, Aber C, Connelly EA. Ehlers-Danlos syndrome type IV: keloidal plaques of the lower extremities, amniotic band limb deformity, and a new mutation. J Am Acad Dermatol. 2007;56(2): S53-S54.
7. Brent RL. Environmental Causes of Human Congenital Malformations: The Pediatrician's Role in Dealing With These Complex Clinical Problems Caused by a Multiplicity of Environmental and Genetic Factors. Pediatrics. 2004;113(4):957-68
8. Martínez-Frías ML. Epidemiological characteristics of amniotic band sequence (ABS) and body wall complex (BWC): are they two different entities? Am J Med Genet. 1997;73(2):176-9.
9. Poeuf B, Samson P, Magalon G. Amniotic band syndrome. Chir Main 2008;27: S 136-47.
10. Tanaka O, Koh T, Otani H. Amniotic band syndrome in a fifth month fetus and in newborn from maternal oophorectomy during early pregnancy. Teratology 1996;33:187-193.
11. Khon G. The amniotic band syndrome: a possible complication of amniocentesis. Prenatal Diagnosis. 1987; 8:303-305.
12. Jabor MA, Cronin ED. Bilateral cleft lip and palate and limb deformities: a presentation of amniotic band sequence? J CraniofacSurg 2000;11(4):388-93.
13. Burk CJ, Aber C, Connelly EA. Ehlers-Danlos syndrome type IV: keloidal plaques of the lower extremities, amniotic band limb deformity, and a new mutation. J Am AcadDermatol. 2007;56(2):S53-S54.
14. Evans C, Marton T, Rutter S, Anumba DO, Whitby EH, Cohen MC. Cranial vault defects: the description of three cases that illustrate a spectrum of anomalies. PediatrDevPathol. 2009;12(2):96-102.

15. KutluDilek TU, Yazici G, Gulhan S, Polat A, Dilek B, Dilek S. Amniotic Band Syndrome Associated With Cranial Defects and EctopiaCordis: A Reportof Two Cases. J Turkish German Gynecol Assoc 2005; 6(4): 308-10.
16. Robin NH, Franklin J, Prucka S, Ryan AB, Grant JH. Clefting, amniotic bands and polydactyly: A distinct phenotype that supports an intrinsic mechanism for amniotic band sequence. American Journal of Medical Genetics. 2005;137A(3):298-301.
17. Allen LM. Constriction Rings and Congenital Amputations of the Fingers and Toes in a Mild Case of Amniotic Band Syndrome. Journal of Diagnostic Medical Sonography 2007;23:280-5.
18. Merrimen JL, McNeely PD, Bendor-Samuel RL, Schmidt MH, Fraser RB. Congenital placental–cerebral adhesion: an unusual case of amniotic band sequence. J Neurosurg. 2006; 104(5): 352–5.
19. Bower C, Norwood F, Knowles S. Amniotic band syndrome: a population-based study in two Australian states. Paediatric and Perinatal Epidemiology. 1993;7(4):395-403.
20. Quintero RA, Morales WJ, Phillips J, Kalter CS, Angel JL. In utero lysis of amniotic bands. Ultrasound Obstet Gynecol. 1997;10(5):316-20.