Amniotic Band Syndrome with CTEV and Meningocele: A Rare Case Report

Sandeep Shrestha[1]*, Kamal Prasad Thani[1], Uttara Gautam[1], Nishan Bhattarai[1], Annie Shrestha[1], Munna Keshari[1], Akina Sainju[1], Santosh Lamichhane[1], Pooja Thapaliya[2]

[1]Department of Paediatrics & Neonatology, Karnali Academy of Health Sciences, Jumla
[2]Department of Medicine, Bayalpata Hospital, Achham, Nepal

Corresponding Author: Dr. Sandeep Shrestha, Associate Professor, Email: sandeepshrsth1@gmail.com

ABSTRACT

Amniotic band syndrome (ABS) is a group of rare congenital abnormalities caused by wrapping of parts of the foetus by fibrous amniotic bands during intrauterine life. It can be seen in infants without any known genetic mutations. Band formation most frequently affects the distal segments, including the hand. Here, we report a case of a neonate who presented with multiple congenital abnormalities and clinical features that suggest the Amniotic Band Syndrome. It was delivered by a 17-year-old female patient at 28 weeks period of gestation, who had a medical abortion.

Key words: Amniotic band syndrome; Malformation; Neonate

INTRODUCTION

Amniotic band sequence (ABS) is a constellation of complex fetal morphological abnormalities associated with intrauterine amniotic bands.[1] It is a rare disorder and with varied degrees of abnormalities.[2] The clinical aspects of the limbs are varied with presence of cutaneous grooves of amputation, pseudo-syndactyly and club feet.[3] The incidence varies from 1 in 1,200 to 1 in 15,000 live births.[4] Both sexes are equally impacted.[5] Anomalies can range in severity from simple craniofacial clefts that are ultimately life-incompatible to single-digit amputations.[6]

Portal first described this malformation in 1685.[7] ABS has several names that have been reported in the literature, including congenital ring constriction, constriction ring syndrome, amnion rupture sequence, amniotic band sequence, and amniotic band disruption complex.[2,8,9] Several explanations have been put out regarding this disease that includes: I. membrane disruption linked to the development of the amniotic band; II. genetic factors; and III. vascular abnormality resulting in an ischaemic event.[10]

CASE REPORT

An 18 year old primigravida who was 28 weeks pregnant came to Karnali Academy of Health Sciences (KAHS) with history of decreased fetal movement and vaginal leak since one day. There was no previous prenatal checkup. There was no significant past medical history for the patient and drug history during the pregnancy and history of consanguinity. Also, congenital anomalies were not found in the family in last 3 generations. A uterus of around 28 weeks' size was noted during an obstetric examination with absent fetal heart sound. Ultrasonography at the time of examination showed intra uterine fetal demise (IUFD) with multiple anomalies. Written consent was taken before the induction with misoprostol regarding the prognosis of outcome. The patient responded well to the induction, giving birth vaginally to a stillbirth boy with a birth weight of 1200 grams (Figure 1).

Clinical examination showed partial sectioning of the index finger, middle finger and ring finger of right hand along with syndactyly of the stumps and normal thumb and little finger (Figure 1). The left hand was unremarkable. Congenital Talipes Equino Varus (CTEV) was seen in right foot (Figure 2) whereas the left leg was normal. A sac protruding from the spinal column likely meningocele was noted (Figure 3). There was flat nasal bridge and low set ears. The fetopathological examination was not done as the parents did not give consent to do so.

DISCUSSION

Amniotic band syndrome is a complex disorder that can result in a number of fetal defects. Clinically, it can be identified by the presence of bands, cutaneous strictures,

Article information		
Received:26 July 2023	Accepted: 21 December 2023	Published online: 31 December 2023

Figure 1: Partial sectioning of the index finger middle finger and ring finger of right hand along with syndactyly of the stumps

Figure 2: CTEV in right foot

Figure 3: Meningocele

amputations of the extremities, and pseudosyndactylies, among other pathognomonic symptoms.[11]

ABS etiopathogenesis is still unknown, but there are two main theories.[12] The first is the intrinsic explanation, which George Streeter put forth in 1930. According to this view, an abnormality in embryogenesis would result in a faulty histogenesis of the fetal tissue, which would result in the fibrous bands. However, it is unable to explain the majority of the anomalies observed.[13] The extrinsic idea, put forth by Torpin in 1965, claims that amnion rupture would lead to the production of fibrous tissue strings that would be responsible for constriction bands, amputation, and other deformities.[14] Congenital abnormalities of the illness can range from limb or extremity amputations to potentially fatal craniofacial, thoracic, and abdominal problems. Amputated limbs or digits, contractures in the limbs, Congenital Talipes Equino Varus (CTEV) also known as club feet, syndactyly, and absent limbs are among the reported defects in the extremities.[15] Craniofacial abnormalities have been reported as encephalocele, anencephaly, cleft lip and cleft palate whereas Thoraco-abdominal complications include omphalocele and gastroschisis.[16]

Prenatal ultrasound can be used to identify ABS. This test may occasionally reveal amniotic bands, but more frequently it will detect abnormalities that are characteristic of ABS, such as oligoamnios and a decrease in fetal activity.[17] The most crucial and important ultrasound diagnostic criteria are constriction rings on the extremities, visible amniotic bands, and irregularly amputated fingers and/or toes with terminal syndactyly.[18]

The appropriate course of action relies on the nature of the anomalies that are present, which can range from life-threatening to functionally and aesthetically detrimental. Because there are so few reliable studies, prenatal therapy, such as fetoscopy to remove constriction bands, is still debatable. In addition, prenatal procedures demand a multidisciplinary organization.[8]

CONCLUSION

Amniotic band syndrome is a rare congenital anomaly affecting the fetus during pregnancy. When fetal deformities are recognized as being incompatible with life, a medical termination of pregnancy may be recommended. And multidisciplinary monitoring is necessary for patients who survive in order to enhance their quality of life.

Data Availability: Related photograph of this case will be available on request

Informed Consent: Consent from patient's father as well

as department of pediatrics, KAHS Teaching Hospital was taken.

Source of Funding/Support: No external funding/ support available.

Conflict of Interest: None declared.

Authors Contribution: SS, KPT, UG (Case Identification and Description, Initial drafting and finalization of manuscript, Submission and Communication) UG, KPT, NB, AS (Critical Review and finalization of manuscript), MK, AS, SL, PT (Literature Review and finalization of manuscript)

REFERENCES

1. Kalousek DK, Bamforth S. Amnion rupture sequence in previable fetuses. Am J Med Genet 1988;31(1):63–73. [Google Scholar] https://doi.org/10.1002/ajmg.1320310110
2. Askins G, Ger E. Congenital constriction band syndrome. J Pediatr Orthop 1988;8:4616. [Google Scholar] [PubMed] https://doi.org/10.1097/01241398-198807000-00016
3. Hennekam RC, Biesecker LG, Allanson JE, Hall JG, Opitz JM, Temple IK. Elements of morphology: general terms for congenital anomalies. Am J Med Genet A. 2013; 161A: 2726-2733. [Google Scholar] [PubMed] https://doi.org/10.1002/ajmg.a.36249
4. Poeuf B, Samson P, Magalon G. Amniotic band syndrome. Chir Main 2008;27 Suppl 1:S13647. [Google Scholar] [PubMed] https://doi.org/10.1016/j.main.2008.07.016
5. Allington NJ, Kumar SJ, Guille JT. Clubfeet associated with congenital constriction bands of the ipsilateral loer extremity. J Pediatr Orthop 1995; 15(5): 599 – 603. [Google Scholar] [PubMed] https://doi.org/10.1097/01241398-199509000-00010
6. Ossipoff V, Hall BD. Etiologic factors in the amniotic band syndrome: A study of 24 patients. Birth Defects Orig Artic Ser 1977;13(13D):117–32. [Google Scholar] [PubMed]
7. Portal P. La Pratique des Accouchements Soutenue d'un Grand Nombre D'observations. Paris: Gabriel Martin; 1685. [Full Text]
8. González ZI, Padilla F. Complex of amniotic deformities, adhesions, mutilations: Endless debate. Bol Med Hosp Infant Mex 2015;72:159-68. [Google Scholar] [PubMed] https://doi.org/10.1016/j.bmhimx.2015.06.006
9. da Silva G, Cammarata SF, González-Coira M, Lacruz MA, Rendon B. Amniotic band syndrome: 3 case reports. Rev Chil Pediatr 2008;79:172-80. [Google Scholar]
10. Iqbal CW, Derderian SC, Cheng Y, Lee H, Hirose S (2015) Amniotic band syndrome: a single-institutional experience. Fetal Diagn Ther 37(1): 1-5. [Google Scholar] [PubMed] https://doi.org/10.1159/000358301
11. Hilali A. La maladie des brides amniotiques aspects orthopédiques et thérapeutiques : à propos d'une série de 24 cas. Université Mohammed. 2013. [Google Scholar]
12. Idris UA. Amniotic band syndrome. Arch Med Health Sci. 2018;6(2):2601. [Google Scholar]
13. Streeter GL. Focal deficiencies in fetal tissues and their relation to intrauterine amputation. Contrib Embryol Carnegie Inst 1930;22:1-44. [Google Scholar]
14. Torpin R. Amniochorionic mesoblastic fibrous strings and amnionic bands: Associated constricting fetal malformations or fetal death. Am J Obstet Gynecol 1965;91:65-75. [Google Scholar] [PubMed] https://doi.org/10.1016/0002-9378(65)90588-0
15. Levy R, Lacombe D, Rougier Y, Camus E. Limb body wall complex and amniotic band sequence in sibs. Am J Med Genet A. 2007; 143A: 2682- 2687. [Google Scholar] [PubMed] https://doi.org/10.1002/ajmg.a.32018
16. Lowry RB, Bedard T, Sibbald B (2017) The prevalence of amnion rupture sequence, limb body wall defects and body wall defects in Alberta 1980-2012 with a review of risk factors and familial cases. Am J Med Genet A 173(2): 299-308. [Google Scholar] [PubMed] https://doi.org/10.1002/ajmg.a.38016
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. [Google Scholar]
18. Merrimen JL, McNeely PD, Bendor-Samuel RL, Schmidt MH, Fraser RB. Congenital placental-cerebral adhesion: an unusual case of amniotic band sequence. Case report. J Neurosurg 2006; 104 (5 Suppl):352-5. [Google Scholar] [PubMed] https://doi.org/10.3171/ped.2006.104.5.352