



Amniotic Band Sequence: A Case Report

Aniruddh Agrawal^{1*}, Adit Singhal², Sohum Shah³, Arun Chutani⁴,
Anisha Agrawal⁵ and Siddhant Bhargava⁴

¹Topiwala National Medical College, 201 Mahalaxmi Building, 12th NS Road, JVPD Scheme, India.

²Commonwealth Medical College, Pennsylvania, USA.

³Georgetown University, Washington DC, USA.

⁴Topiwala National Medical College, Mumbai, India.

⁵Sathaye College, Mumbai, India.

Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/IJMPCR/2016/27122

Editor(s):

- (1) Daniel Laubitz, Steele Children's Research Center, Department of Pediatrics /Gastroenterology and Nutrition, Arizona Health Sciences Center, University of Arizona, Tucson, USA.
- (2) Erich Cosmi, Director of Maternal and Fetal Medicine Unit, Department of Woman and Child Health, University of Padua School of Medicine, Padua, Italy.
- (3) Syed A. A. Rizvi, Department of Pharmaceutical Sciences, College of Pharmacy, Nova Southeastern University, USA.

Reviewers:

- (1) Anonymous, All India Institute of Medical Science, New Delhi, India.
 - (2) Balakumar K, Balku's Scan, PVS Hospital, India.
 - (3) Hakan Timur, Zekai Tahir Burak Women's Health Education And Research Hospital, Ankara, Turkey.
 - (4) Michal Blaszczyński, Poznan University of Medical Sciences, Poznan, Poland.
 - (5) Subhajeet Dey, ESIPGIMSR and ESI Hospital, (GGSIP University), New Delhi, India.
- Complete Peer review History: <http://www.sciencedomain.org/review-history/15985>

Case Study

Received 20th May 2016
Accepted 23rd August 2016
Published 30th August 2016

ABSTRACT

Background: Amniotic band sequence (ABS) is a rare condition in which parts of a fetus get entangled by the amniotic bands attached to the fetal surfaces and structures. This can lead to grotesque structural fetal anomalies or constriction rings that appear in-utero. The severity of this condition ranges from visceral herniation or mild constriction to complete amputation of a digit or limb. This anomaly affects one infant out of 11,200 and this number has shown a stable trend during the last 17 years. Management of this condition is individualized and can include Z-plasty, W-plasty and other such cosmetic surgeries for mild conditions. Severe conditions may require neurovascular reconstruction and even amputations.

The Case: This paper reports a 6-month-old female who presented with constriction rings on both

*Corresponding author: E-mail: anirudhagrwal25@gmail.com;

lower limbs and on the upper left limb. She was the youngest of 3 siblings, none of which presented with amniotic band sequence at birth. The child was of an African descent and was born in Tanzania to parents of a standard economic class. The condition was observed at birth and not in-utero. The baby was brought to a tertiary care hospital in Mumbai, India where she underwent Z-plasty surgery. The baby has now recovered from the surgery and appears to be in good shape.

Conclusion: The etiology of the amniotic band sequence is still not clear. The mother of the patient provided a history free from illnesses or abnormal discomfort during the term of the pregnancy. Identification of this anomaly in the pre-natal periods is very important as one can initiate preventive measures. Prenatal ultrasonography is the only screening method to diagnose these deformities.

Keywords: Amniotic band syndrome; amniotic band sequence (ABS); constriction deformities; syndactyly.

1. INTRODUCTION

Amniotic Band Syndrome Amniotic band sequence is an abnormal congenital pathological condition that shows multiple anomalies in an infant, the most common being automated amputation of digits of the upper or lower limbs and the presence of constriction rings that usually appears in association with fibrous bands [1]. A number of different synonyms have been adopted for the condition among which are "ADAM Complex", "pseudoinhum" and "congenital constriction bands" [2] The exact prevalence of this condition is unknown but it is estimated to be between 1:11,200 [3] and 1:1,200 [4].

The cause of this disease has tried to be explained by two theories. Two theories have tried to explain the cause of this anomaly. One suggests a common origin for the bands, caused by a disturbance to the developing germinal disc of the embryo and therefore attributes the disorder to rise internally within the fetus [intrinsic model] [5]. The other theory suggests that the defects are caused by action of fibrous amniotic bands with the sequence rupture of amnion, followed by extrusion of all parts of the fetus into the chorion, therefore attributing the deformities to a factor which acts on the fetus externally [extrinsic model] [6].

ABSABS is usually diagnosed by antenatal ultrasonographic screening or right after birth based upon the characteristic physical findings. These characteristic findings in mild-degree involvement refer to ring-like constrictions or amputation defects. The severity of the bands has a wide range. Bands that wrap around fingers and toes are considered mild as they may result in syndactyly or amputation of only the digits. However, severe bands can form around the limb and even constrict it causing a

decreased blood supply, which may result in amputation of the whole limb.

In this paper, we report a 6-month old female infant of African descent presented with constriction bands in both legs and the left hand.

2. CASE REPORT

A 6-month-old girl with constriction rings present on both lower limbs and the upper left limb was brought to our hospital. The child did not present with cleft lip. The child was born in Tanzania and was born to parents of an African descent. The child was the youngest amongst three siblings, born to parents of a standard economic class. No other siblings presented with amniotic band sequence. The maternal age at conception was 31. There was no history of abnormal discomfort or induced fever during the gestation period. The patient took no medication other than the general ones prescribed by the antenatal clinic. The patient was brought to India for medical intervention. The deformity was not observed during pre-natal diagnostics due to lack of antenatal ultrasound screening facilities, but at the time of delivery. The team of specialists that examined the patient consisted of an orthopedic surgeon, plastic surgeon, and a cardiologist.

The patient presented with syndactyly in the left hand between 3rd and 4th fingers, severe congenital constricting bands just above the ankle joints in both legs, significant edema in distal part of both feet and hypoplastic toes in the left foot.

Clinical examinations confirmed a structurally and functionally normal heart. Inspection of the other systems showed no abnormalities. No other associated congenital abnormalities were detected. The team made a clinical diagnosis of

Amniotic band sequence. Since then, the child has undergone standard Z-plasty surgeries for both legs in a single sitting. Patient is doing well 6 months post-surgery and the swelling on the right leg, where the constriction band was severe, is gradually regressing.



Fig. 1. Shows the right leg of the infant, pre-operatively. Apparent edema is observed along with presence of severe constriction rings above the proximal part of the foot



Fig. 2. Shows the left leg of the infant pre-operatively. Distal phalanges have been cut off from the toe and the second finger. Severe constriction bands are observed. Edema is less marked

3. DISCUSSION

Amniotic Band Syndrome Amniotic band sequence comprises of a set of observable malformations that range from minor constriction rings and lymphedema of the digits to complex multiple anatomical disturbances that can be attributed to sticky amniotic bands that entangle and disrupt fetal systems [7,8]. Familial history rarely reveals any information that could lead to establishment of an inheritance pattern because this symptom does not occur in association with any known chromosomal or genetic disorder. However, it has been hypothesized that ABS is

associated with families suffering from genetic connective tissue abnormalities such as Ehlers-Danlos syndrome and osteogenesis imperfect [9]. The patient presented in this paper however, reported no familial history of genetic collagen disorders or connective tissue diseases.



Fig. 3. Shows the left leg of the infant 3 months post-operatively. The constriction band has been alleviated and is presented with a small circular protrusion on distal part of leg



Fig. 4. Shows the right leg of the infant 3 months post-operatively. One can observe reduced edema in the feet as well as alleviation of amniotic bands

The anatomical cause of occurrence of amniotic band sequence is the tearing of the inner membrane of the amniotic sac of the growing fetus. This membrane wraps around developing baby and results in the formation of the anatomical disturbances like visible constriction bands, automated amputation, cleft palate, etc [10]. Early rupture can cause major anomalies (at about 5 gw- craniofacial anomalies and placental attachment to head or abdomen; at about 7 gw- thoracoabdominal defects, scoliosis, limb

reduction; after 9 gw- limb abnormalities like hypoplasia, amputation, deformities) and late rupture results in ring constrictions, amputations and digital fusion. The patient however, did not present with cleft palate or automated amputation but instead presented with severe constriction bands on three limbs.

In sub-Saharan countries, where this patient hails from, in-utero prevalence of ABS is difficult to estimate because of inadequate antenatal facilities.

A study conducted by the National Center on Birth Defects and Development Disabilities observed that maternal cigarette smoking increased the risk for amniotic band sequence. The mother of the patient however, claimed to be a non-smoker [11].

A very weak association has been observed between ABS and disruption of internal organs therefore the absence of any deformity of internal organs in the patient was no surprise. The outcome of this disease however, depends on the degree of severity of the malformations that occur in-utero. In one case, strangulation of umbilical chord by amniotic band had occurred which caused death of the fetus in the third trimester. This indicated the unpredictability of the manifestations of this syndrome [12].

This disease is the most difficult to detect in underdeveloped countries, especially those of Sub-Saharan Africa, the birthplace of this particular patient. This difficulty arises from the lack of antenatal ultrasound screening and the inefficient monitoring of pregnancies, therefore preventing early diagnosis of the disease. Illiteracy, poverty and less medicalization cause a delayed diagnosis of ABS and therefore increase the mortality rate [13].

4. CONCLUSION

A case of a 6-month old girl, hailing from Tanzania, which presented with amniotic band sequence has been reported. The girl has undergone a Z-plasty surgery to improve the cosmetic appearance of lower limbs. The surgery was a success as the bands seem to dissipate and the swelling is slowly reducing. The patient may undergo another surgery on the left leg to further reduce the scarring. In this case, no pre-natal diagnosis of amniotic band sequence was made in Africa, due to insufficient pregnancy

monitoring and inefficient pre-natal care. This case highlights the importance of general availability of pre-natal care such as ultrasonography to diagnose such congenital diseases at an earlier stage in the pregnancy. In this case however, the timing worked in the child's favor and she did not face the worst this disease had to offer.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this paper and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Seeds JW, Cefalo RC, Herbert WN. Amniotic band syndrome. *Am J Obstet Gynecol.* 1982;144(3):243-8. Epub 1982/10/01. PubMed PMID: 7124837.
2. Ross MG. Pathogenesis of amniotic band syndrome. *Am J Obstet Gynecol.* 197. United States. 2007;219-20:author reply 20.
3. Martinez-Frias 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. Epub 1997/12/31 23:48. PubMed PMID: 9409868.
4. Ossipoff V, Hall BD. Etiologic factors in the amniotic band syndrome: A study of 24 patients. *Birth Defects Orig Artic Ser.* 1977;13(3d):117-32. Epub 1977/01/01. PubMed PMID: 922131.
5. GL S. Focal deficiencies in fetal tissues and their relation to intrauterine

- amputations. *Contrib Embryol Carnegie Institute*. 1930;1-44.
6. Torpin R. Amniochorionic mesoblastic fibrous strings and amnionic bands: Associated constricting fetal malformations or fetal death. *Am J Obstet Gynecol*. 1965;91:65-75. Epub 1965/01/01. PubMed PMID: 14245093.
 7. Adeosun OO, James O, Akinmoladun VI, Owobu T. Amniotic band syndrome associated with orofacial clefts: A report of two cases.
 8. Mahmood N, Board A. Amniotic bands: Are they all threat for the fetus? *Bahrain Medical Bulletin*. 2008;30(2).
 9. Kroes H, Pals G, Van Essen A. Ehlers–danlos syndrome type IV: Unusual congenital anomalies in a mother and son with a COL3A1 mutation and a normal collagen III protein profile. *Clinical Genetics*. 2003;63(3):224-7.
 10. Poeuf B, Samson P, Magalon G. Amniotic band syndrome. *Chirurgie de la Main*. 2008;27:S136-47.
 11. Werler MM, Bosco JL, Shapira SK. Maternal vasoactive exposures, amniotic bands and terminal transverse limb defects. *Birth Defects Res A Clin Mol Teratol*. 2009;85(1):52-7. Epub 2008/12/11. DOI: 10.1002/bdra.20524 PubMed PMID: 19067400; PubMed Central PMCID: PMC2741326
 12. Chatzigeorgiou K, Theodoridis T, Efstratiou I, Athanasiadis A, Zepiridis L, Tzevelekis F, et al. Strangulation of the umbilical cord by an amnion band - a rare cause of intrauterine demise: A case report. *Cases J*. 2009;2:9108. Epub 2010/01/12. DOI: 10.1186/1757-1626-2-9108 PubMed PMID: 20062685; PubMed Central PMCID: PMC2803905.
 13. Nagalo K, Badiel R, Koueta F, Tall FH, Ye D. Amniotic bands syndrome and its diagnostic difficulties and management in Burkina Faso. *Pan Afr Med J*. 2015;20:208. Epub 2015/06/27. DOI: 10.11604/pamj.2015.20.208.6129 PubMed PMID: 26113939; PubMed Central PMCID: PMC4470413.

© 2016 Agrawal et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:

The peer review history for this paper can be accessed here:
<http://sciencedomain.org/review-history/15985>