https://doi.org/10.36570/jduhs.2024.3

JDUHS

CASE REPORT

Amniotic Band Sequence in an Intrauterine Fetal Demise at 17 weeks of Pregnancy: A case report

Vladimir Bartos
Martin Biopsy Center, Ltd., Martin, Slovakia
Correspondence to: Dr. VladimIr Bartos , Email: vladim.bartos@gmail.com ORCiD: 0000-0002-7394-7241

ABSTRACT

Amniotic band sequence is a rare disorder characterised by various congenital abnormalities due to constrictions of certain parts of the fetal body by fibrous bands derived from the amniotic sac. A 29-year-old pregnant woman with a missed abortion at 17 weeks of gestation is reported. A routine prenatal 2D-ultrasound scanning done three weeks ago revealed a living fetus without any obvious structural abnormality. A medical abortion was induced. A macerated fetus was delivered whose legs were tightly wrapped by massive amniotic bands, in which part of the umbilical cord was also entangled. After their removal, the legs and feet were deformed with marked strangulation grooves. The left calf was edematous and thickened above that narrowing with histological feature of chronic myositis. This unique case demonstrates an aborted fetus with striking amniotic bands detected by fetal autopsy. Considering that even such a serious pathology may not be identified prenatally by ultrasound, this condition should be kept in mind while evaluating the cases of sudden intrauterine fetal death. It is important to thoroughly investigate all aborted fetuses for potential features of amniotic band sequence, even in early miscarriagies.

Keywords: Amniotic Band Sequence, Congenital Anomalies, Intrauterine Fetal Demise.

INTRODUCTION

Amniotic band sequence (ABS) is a rare disorder characterised by various congenital abnormalities due to constrictions of certain parts of the fetal body by fibrous bands derived from the amniotic sac.[1-3] This condition has an extremely variable manifestation that includes three types of anomalies: disruptions, deformations and malformations.[1,2] It can range from single ring constrictions of the digits which can be only aesthetic matter to complex anomalies of the extremities and other body sites which may be incompatible with life.[1-3] Because of obvious variation in its presentation, many synonyms for this entity have been used, e. g. amniotic deformity, adhesion, and mutilation (ADAM) complex , amniotic band syndrome, amniotic disruption complex, amnion rupture sequence, congenital constriction bands, Streeter bands, Streeter syndrome or Streeter dysplasia.[1] The prevalence of ABS ranges from 0.19 to 8.12 per 10,000 live births.[2] However, the disease probably occurs much more frequently because it is reported to be ten times more common in embryos and fetuses than that of neonates.[4] In one study the prevalence ratio of ABS was 1:56 in fetuses up to the 18th week of gestational age.[5] The diagnosis of ABS is most often made after birth when typical abnormalities of the newborn's limbs are discovered.[6-9] At that time, the amniotic bands are usually absent and visible somatic anomalies are indirect signs of their previous existence. Well-identifiable amniotic bands may be best documented by necroptic analysis of aborted fetuses in earlier stages of pregnancy. However, not many such studies have been published so far.[4,10-13] Herein, a case of spontaneously aborted fetus in early mid-trimester of gestation with histomorphologically confirmed massive amniotic bands wrapping and deforming the legs is described.

CASE REPORT

A 29-year-old pregnant woman, primigravida, with no particular medical history was referred to hospital at 17 weeks of gestation with a diagnosis of intrauterine fetal demise confirmed by recent ultrasound. It was a spontaneous pregnancy of a non-consanguineous partnership, the course of which had been uneventful until then. A routine prenatal 2D-ultrasound scanning done three weeks ago revealed a living fetus without any obvious structural abnormality (crown rump length 71 mm, nuchal translucency 1,8 mm, fetal heart rate 161 bmp). A medical abortion was induced per vias naturales and a macerated male fetus with placenta were delivered. Grossly, the lower limbs of the fetus were tied" together by strips of elastic tissue, in which

part of the umbilical cord was also entangled. No fusion of the fetus with the chorionic plate of the placental disc was seen. After cutting the umbilical cord, the fetus and the placenta were fixed in formalin and sent separately to the pathology laboratory. The pathologist performed a thorough macroscopic revision of the samples. The fetus weighed 200 g. and had a crown-rump lenght of 12 cm. On external examination, the distal parts of both lower extremities were tightly wrapped by fibrous bands that were fused with the skin and subcutaneous soft tissue (Figure 1, 2 and 3). The loops were firm and almost impossible to extract manually. After its removal using tweezers the legs and the feet were deformed and disrotated with marked strangulation grooves (Figure 4). The left calf was edematous and thickened above that narrowing. The placenta could not be reliable evaluated because it was artificially altered with torn placental membranes. The umbilical cord had a total lenght of 18 cm, which was normal when the length was adjusted to the developmental age of the fetus.

Histologically, the fibrous bands consisted of amniotic tissue with well preserved, partly hyperplastic amniotic epithelium (Figure 5). In skeletal muscle taken from the left calf a diffuse lymphocytic inflammatory infiltration was seen, reflecting a long term irritation. There were no other identifiable congenital anomalies in the fetus. As the rest of the autopsy including internal investigation was negative, the cause of death was determined to be amniotic band sequence.

DISCUSSION

The ABS comprises a heterogeneous scale of congenital abnormalities that has been divided into three groups based on the body location affected: a) extremities, b) craniofacial region, c) other body areas.[3] Typical limb anomalies include constrictions with distal edematous swelling, various deformities, syndactilies, and partial or total digit or limb amputations.[3,8] Craniofacial defects include distortion of the face, cleft lip or palate, eye/ear/nose deformities or various anomalies of the brain.[3] Other somatic malformations contain thoraco-abdominal wall defects, and spinal column abnormalities.[3] The proportions of the limb and the non-limb involvement have greatly varied depending on given studies. For example, Kalousek et al.[4] studied eighteen affected fetuses, of which eleven individuals (61%) had limb defects only and 7 fetuses (39%) also had additional somatic anomalies. In contrast, Guzmán-Huerta et al.[3] analyzed 50 patients with prenatally diagnosed ABS. They found a single case (2%) with limb involvement only, while the vast majority showed associated malformations in other parts of the body. They proposed[3] a new classification of the ABS phenotypes depending on the anatomic region which are involved. These include: I. Craniofacial defect + limb defect, II. Craniofacial defect + limb defect + abdominal wall, spinal column, and/or thoracic defects, III. Limb defect + abdominal wall, spinal column, and/or thoracic defects; and IV. Isolated defect (craniofacial, limb, or thoraco-abdominal wall). According to this classification scheme our case falls under the category of isolated defect (only limb involvement).

The pathogenesis of ABS has been debated for a long time and is still not clear, but two basic theories were put forward: the intrinsic and extrinsic models.[1-3] The intrinsic theory supposes that the fibrotic bands and fetal abnormalities harbor a common origin, which is due to imperfect histogenesis and defective development of the germinal embryonic disc during the first weeks of gestation. It follows that the amniotic bands as such are not direct consequences of the fetal malformations observed.[1-3] The extrinsic theory

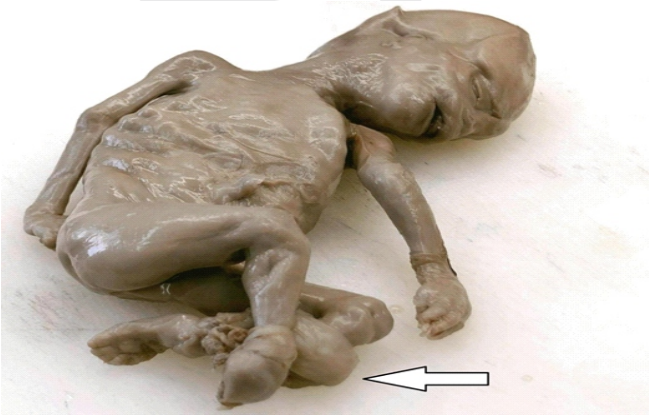

Figure 1: Whole-body picture of the macerated fetus showing lower extremities „tied“ together (arrow). (formalin-fixed sample)

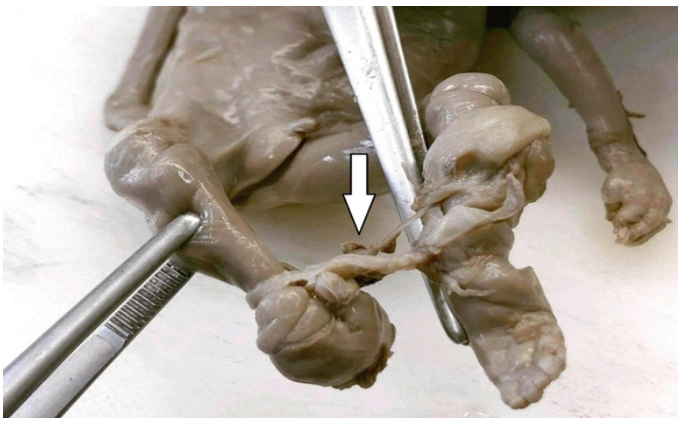

Figure 2: Detail on lower limbs entangled with massive amniotic bands (arrow).
(formalin-fixed sample)

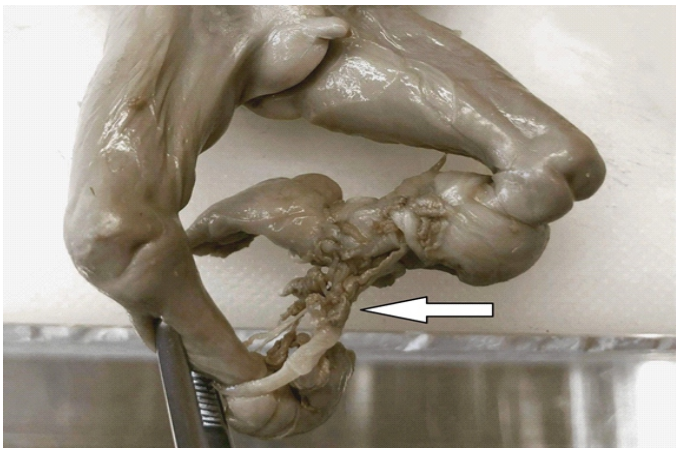

Figure 3: Amniotic bands (arrow) fused with the skin and subcutaneous soft tissue of the legs.
(formalin-fixed sample)

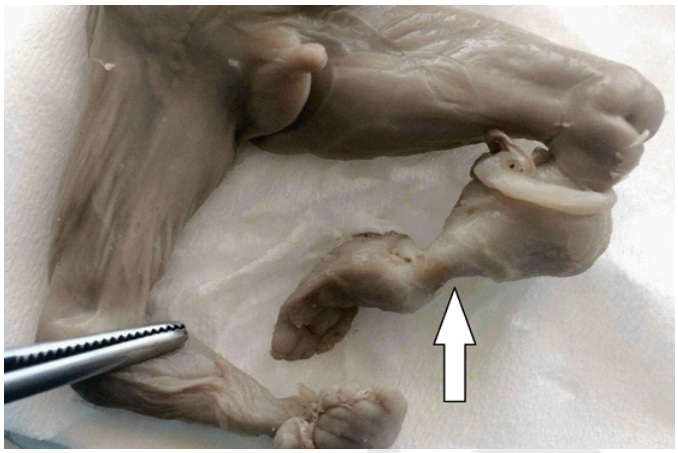

Figure 4: Severe deformations and strangulation grooves (arrow) are visible in the legs after removal of the amniotic bands. (formalin-fixed sample)

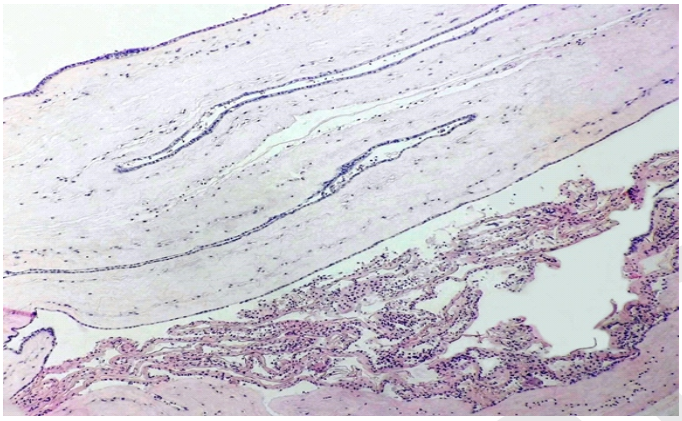

Figure 5: Amniotic band with well preserved, partly hyperplastic (lower part) amniotic epithelium. (hematoxylin & eosin, 20x)

suggests that the body anomalies have a secondary origin and are caused by bands of amniotic tissue as a result of amnion rupture. The limbs or other parts of the fetal body are entrapped by these bands causing compression, tissue necrosis and eventually amputation of fingers or extremities, followed by their discharge into the chorionic cavity.[1-3] Our case rather supports the extrinsic theory as the primary pathological mechanism.

The overall prognosis of affected babies mainly depends on which body part gets entrapped and at what time of gestation.[3,9,10,13] Clinical manifestation may range from a cosmetic imperfection only to malformations incompatible with postnatal life.[3] More severe phenotype of the fetus usually results in intrauterine demise. Umbilical cord strangulation by amniotic bands causing fetal asphyxia is probably a common cause of intrauterine death in ABS. Kalousek *et al*.[4] discovered it in 33.3% of all dead fetuses. However, that percentage can be higher because in the remaining cases the umbilical cord was not available for complete analysis. Even in our case the fetal demise seemed to be directly linked to umbilical cord constriction by amniotic bands. Two similar cases have also been published in recent literature.[12,13] Shah *et al*.[13] describe a fetus that spontaneously died at 16 weeks of pregnancy. It has an amniotic band between lower limbs, entangling right foot and left ancle, involving the umbilical cord. Elayedatt *et al*.[12] report a case of intrauterine fetal demise at 17 weeks of gestation. The fetus showed bands of tissue running across the dorsal surface of right palm, above the level of both ankle joints and one string was seen constricting the umbilical cord. From a gynecological point of view, a prenatal diagnosis of ABS by routine ultrasound is intricating and challenging. It should be taken into account when characteristic asymmetric anomalies of the fetus are seen ultrasonographically, regardless the presence or absence of fibrous streaks.[1-3,14-16] It is almost impossible to detect the amniotic bands in the first trimester of pregnancy. In the second and third trimesters, there is accessible to identify the major abnormalities suggestive of ABS, i.e. absent digits or distal portions of the limbs, or swollen deformed parts of the upper or lower limbs secondary to constrictive bands.[1-3,14,15] The visualization of amniotic bands on ultrasound is not required to make the diagnosis.[16] The disease is therefore more often established in utero by the effects that the strings give to fetal morphology. Anyway, an antenatal diagnosis is carried out in only 29-50% of cases.[1] Even in our case the prenatal ultrasound scan did not suggest any pathology and the disease was diagnosed incidentally on post mortem examination. Recently, a few similar case reports of intrauterine fetal demise have been described,[10-12] in which the ABS remained undetected antenatally but was definitively revealed by fetal autopsy.

The current trends have attempted to treat selective cases of ABS prenatally by fetoscopy.[17-19] The fetoscopic laser ablation and mechanical lysis may release the constriction bands and thus prevent the amputation or severe damage of affected structures.[17-19] Although this procedure is technically feasible, it is a highly specialized method, the outcomes of which have been controversial so far.

CONCLUSION

This unique case demonstrates an aborted fetus with massive amniotic bands detected by fetal autopsy. Considering that even such a serious pathology may not be identified prenatally by ultrasound, this condition should be kept in mind while evaluating the cases of sudden intrauterine fetal death. It is important to thoroughly investigate all aborted fetuses for potential features of amniotic band sequence, even in early miscarriagies.

Received: August 23, 2023
Accepted: October 30, 2024

REFERENCES

1. RodrIguez GonzAlez ZI, Soriano Padilla F. Complex of amniotic deformities, adhesions, mutilations: endless debate. Bol Med Hosp Infant Mex 2015; 72:159-68. doi:10.1016/j.bmhimx.2015.06.006
2. Lopez-Munoz E, Becerra-Solano LE. An update on amniotic bands sequence. Arch Argent Pediatr 2018; 116:409-42. doi:10.5546/aap.2018.eng.e409
3. Guzman-Huerta ME, Muro-Barragan SA, Acevedo-Gallegos S, Velazquez-Torres B, Gallardo-Gaona JM, Ramirez-Calvo JA, et al. Amniotic band sequence: prenatal diagnosis, phenotype descriptions, and a proposal of a new classification based on morphologic findings. Rev Invest Clin 2013; 65:300-6.
4. Kalousek DK, Bamforth S. Amnion rupture sequence in previable fetuses. Am J Med Genet 1988; 31:63-73. doi:10.1002/ajmg.1320310110
5. MbodjiA, WadeM, GueyeM, NdiayeMD, DiaAD, SeneM, et al. Amniotic band syndrome: a new case report. Int J Reprod Contracept Obstet Gynecol 2021;10:2864-6. doi:10.18203/2320-1770.ijrcog20212682
6. Jaanbasha M, Nagnur MI, Mohiuddin MS, Mohiuddin MJ, Salman S, Sunder CS. Streeter's syndrome of lower limb associated with CTEV. J Orthopaedic Case Reports 2022; 12:50-3. doi:10.13107/jocr.2022.v12.i12.3460
7. Phan TH, Thi Nguyen PT, Nguyen PN, Pham HH, Ngo QD, La Nguyen PT. Amniotic band syndrome leading to severe malformations of the newborn: a case report at Tu Du Hospital, Vietnam, and literature review. Ann Med Surg (Lond) 2023; 85:592-7. doi:10.1097/MS9.0000000000000263
8. Demetri LR, Starcevich AG, Manske MC, James MA. Clinical manifestations of constriction band syndrome. J Bone Joint Surg Am 2022; 104:1301-10. doi:10.2106/JBJS.21.01286
9. Neupane S, Shrestha S, Jha MK, Shakya N, Neupane P. Amniotic band sequence: A rare case seen in Birat Medical College. BJHS 2023; 8:1995-8. doi:10.3126/bjhs.v8i1.57294
10. Mishra VV, Solanki SB, Aggarwal R, Shah K, Anusha M. Amniotic band syndrome: a diagnostic dilemma. Int J Reprod Contracept Obstet Gynecol 2022; 11:2049-51.
11. Sahu KK, Rao R, Chaithra GV, Philipose CS, Sneha Rao AR, Vatsala KB. Amniotic band syndrome- A rare case report. Biomedicine 2023; 43:1056-8. doi:10.51248/.v43i3.2495
12. Elayedatt RA, Krishnan V, Chandraprabha V. Fetal autopsy-a game changer. J Fetal Med 2023; 10:99-104. doi: 10.1055/s-0043-1776056
13. Shah K, Sangha S, Ashwini AP, Bhat S, Vinod V. Amniotic band destruction syndrome causing umbilical cord strangulation in a pregnancy with diabetes: a case report. Obstel Gynaecol Forum 2021; 31:28-30.
14. Sujetha R, Palanivelraja T, Kumaravel KS, Gobinathan S, Sampathkumar P. Amniotic band syndrome: A case report and review of literature. Pediatr Oncall J 2022; 19:134-5. doi:10.7199/ped.oncall.2022.49
15. Lamrisi A, Mourabbih M, Ouajih O, Jalal M, Fichtali K, Bouhya S. Amniotic band syndrome: A case report. Int J Surg Case Rep 2022; 95:107096. doi:10.1016/j.ijscr.2022.107096
16. Munisteri MK, Gonzales-Brown V, Villazana-Kretzer D. Amniotic band syndrome: A case series. J Case Rep Images Obstet Gynecol 2022; 8:33-38. doi:10.5348/100122Z08MM2022CS
17. Minella C, Costantino B, Ruano R, Koch A, Weingertner AS, Favre R, et al. Fetoscopic release of amniotic band syndrome: an update. J Ultrasound Med 2021; 40:1039-48. doi:10.1002/jum.15480
18. Kumru S, Kaya S, Atim A, Aslan A. Treatment of an amniotic band syndrome with fetoscopic surgery: a case report. Perinat 2022; 30:195-9. doi:10.2399/prn.22.0302006
19. Dogru S, Acar A. Fetoscopic surgery for amniotic band syndrome: Case series. Congenit Anom (Kyoto) 2023; 63:4-8. doi:10.1111/cga.12494