AFRICAN JOURNAL OF FETOMATERNAL MEDICINE

Case Report

Complex Fetal Deformity with Placenta Morbidly Adherent to the Head Sequel to Amniotic Band Sequence in a Live-Term Birth: A Case Report

Ukwu A.E[1]., Akubo A[1]., Adeyi S., Legbo JN[3]

[1]Department of Obstetrics and Gynaecology Usmanu Danfodiyo Unuversity Teaching Hospital, Sokoto; [2]Neonatology Unit, Department of Paediatrics , Usmanu Danfodiyo University Teaching Hospital, Sokoto; [3]Plastic Surgery Unit, Department of Surgery, Usmanu Danfodiyo University Teaching Hospital, Sokoto.

Abstract

Amniotic band sequence, ABS, is a disorder of body deformities arising from the presence of amniotic bands that constrict the limbs, the body wall, the viscera, or other parts of the fetus including the head/neck region, and even the placenta. This results in simple or multiple constriction band deformities. Simple deformities are compatible with life and easily surgically correctible while complex deformities often result in abortions, stillbirth, or in live births with severe multiple malformations that die a few hours or few days. We present a very rare case of severe complex or multiple disorders involving the limbs, the body, the viscera, the head/neck region, and the placenta born to a 26-year-old primigravida who attended antenatal care and had an antenatal ultrasound diagnosis of omphalocele, delivered at term and survived several days up to 8 that presented very tasking management difficulties.

Keywords: Amniotic band sequence, complex fetal malformations, live birth.

Correspondence:

Dr Ukwu A Eze,
Department of Obstetrics and Gynaecology,
Usmanu Danfodiyo University Teaching Hospital, Sokoto.
Email: ukwuaeze@gmail.com

Introduction

Amniotic Band Sequence (ABS) is a terminology often used interchangeably with Amniotic Band Syndrome. However, the features of Amniotic Band Sequence Amniotic Band Syndrome are not consistently the same and the sequence is not of common single causative factor origin as in the syndrome. It is considered a sequence rather than a syndrome because the pattern of congenital anomalies is related to a single type of insult resulting from one or more amniotic bands that can result from a variety of different aetiologies for example rupture of membranes, vascular insult. By contrast, a syndrome refers to a pattern of congenital abnormalities that are known or at least assumed to result from a single aetiology for example Down syndrome is due to trisomy 21. Amniotic Band Sequence (ABS) refers to congenital abnormalities that occur in association with amniotic bands [1-6]. Heterogeneity in clinical manifestations, diagnosis, and recurrence risk of amniotic band-associated anomalies has contributed to confusion regarding nomenclature and so diverse terms have been used historically to refer to varieties of the same condition such as Amniotic Band Sequence, Amniotic Band Syndrome, Amniotic Band Disruption Complex, Body Wall Complex with Limb Deformities, Amniotic Deformity, Adhesion Mutilating (ADAM) Sequence [6-14]. The abnormalities in Amiotic Band Sequence occur after the affected parts of the body have formed normally. Intrinsic factors within the fetus or extrinsic factors

acting externally on the fetus or both the intrinsic and extrinsic factors may be the cause of ABS. The cause of the disorder in one infant may be different from the cause of the disorder in another infant.

Case Report

The index case was a born to a 26year old primigravida booked at an advanced gestational age of 32 weeks She had an early ultrasound scan at 10 weeks of gestation to confirm her pregnancy but booked late for antenatal care. She was regular at her antenatal visits and the pregnancy was largely uneventful with no report of any febrile illness. Routine investigations during pregnancy including urinalysis, PCV (48%), VDRL, HBsAg, RVS were all normal. The blood group was O-Rhesus positive, and the hemoglobin genotype was AA. Obstetric ultrasound at 36 weeks however revealed a congenital anomaly (omphalocele) and transverse lie. She was counseled for elective cesarean section at term if the transverse lie persisted. She developed labor pains at 38 weeks' gestation and so had an emergency cesarean section with difficult delivery on account of transverse lie and the complex multiple congenital abnormalities found.

The findings at birth were a male baby with complex multiple congenital abnormalities. It had anencephaly, protruding brain matter, Placenta tissue firmly attached to the head, disfigured facial features, absent nostrils with bilateral cleft lips and palate, contractures, anophthalmia of the right side, rocker bottom feet, polysyndactyly, multiple tail-like soft tissues -5 in number attached to the back, and hypoplasia of the right-hand 3rd and 4thdigits. The baby did not cry at birth but responded to minimal resuscitative efforts. The APGAR Scores were 1 in 3 minutes, 6 in 5 minutes and 9 in 10 minutes. SPO2 was between 84 -87%. The birth weight was 3.5kg. The blood loss was about 800mls.
The mother was discharged from the postnatal ward on the 4th day postoperatively for a review at the post-natal clinic in two weeks. She was counseled on expressed breast milk, EBM, for her baby, childhood immunization, and contraception.

The baby was admitted to the special care baby unit, SCBU, and neonatologists co-managed him with a team of neurosurgeons, ortho-rhino-laryngologists, and plastic surgeons until death on the 8th day of life.

Trans-fontanelle Ultrasound scan done on the 5th day of life demonstrated cerebral parenchyma herniating into anterior cranial swellings more marked on the right side, and CSF with mobile internal echoes surrounding the herniated brain parenchyma. The cerebellum showed normal echogenicity. The interhemispheric fissure was demonstrated. However,

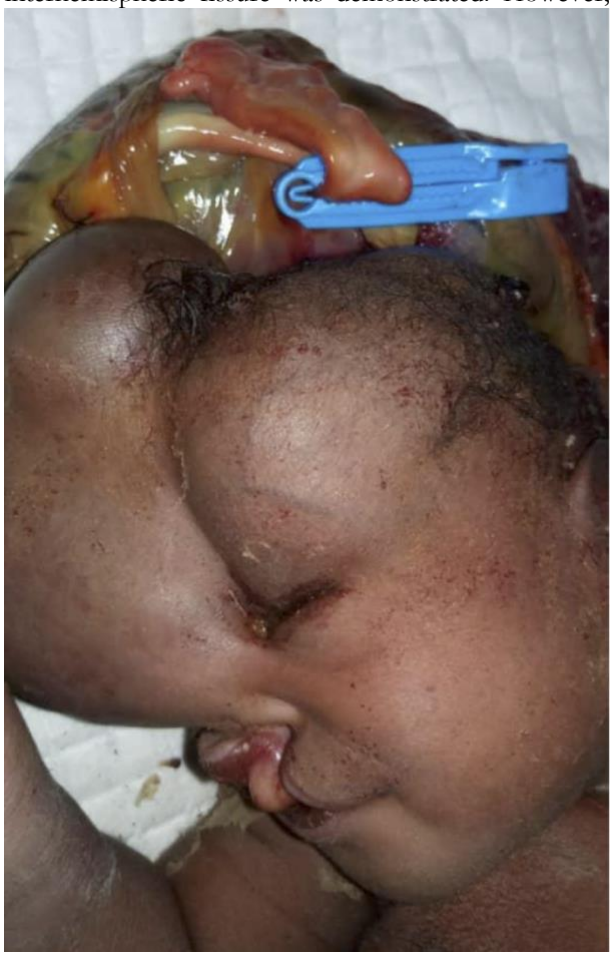

Figure1: Head Morbidly adherent to Placenta

the corpus callosum and the thalami were not demonstrated. The right lateral ventricles, the third and fourth ventricles were not demonstrated.

The ocular ultrasound scan demonstrated a normal-sized left eyeball 17x17mm in size. The left lens and the posterior segments were within normal limits. No focal mass lesion was seen. The left retro-bulbar space, the extraocular muscles and the optic nerve were within normal limits. The right was orbit was not demonstrable.

The Abdominal ultrasound scan done demonstrated normal liver, normal intra and extra hepatic bile ducts, normal gall bladder, normal pancreas, spleen and kidneys. The bowel loops were normal and had good peristalsis. An omphalocele was demonstrated. The urinary bladder was not demonstrable, presumably empty. The conclusion was essentially an omphalocele and otherwise normal internal abdominal organs on abdominal ultrasound scan.

Figure 2: Anterior Abdominal Wall Defect Covered

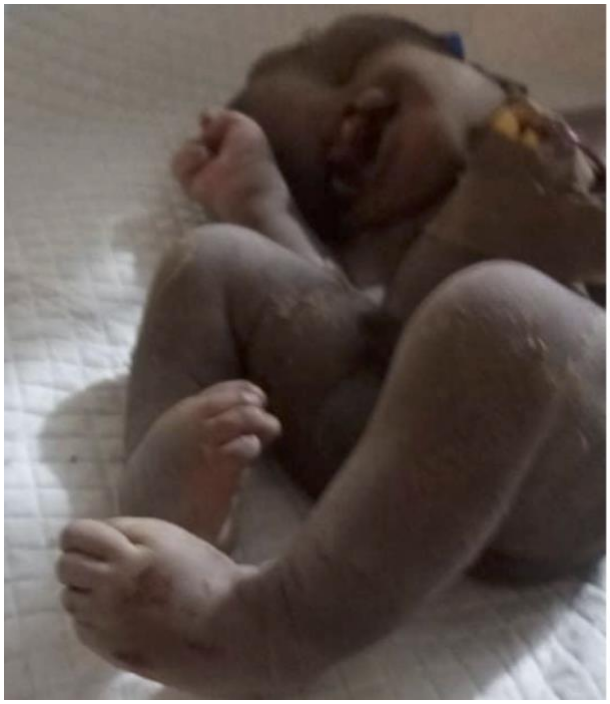

Figure 3: Shows Facial Anomalies including cleft lips

Figure 4: Upper Limb Anomalies

Figure 5: Showing Mouth and Nasal Anomalies

Figure 6: Showing Upper Limb Anomalies

A diagnosis of Amniotic Band Sequence with Complex Multiple Congenital Anomaly and morbidly adherent placenta to the head, not compatible with life was made.

Discussion

Cases of multiple complex anomaly due to Amniotic Band Sequence, ABS, end in lifeless fetal abortions in most cases or stillbirths at advanced gestational age or at term. When born alive at advanced gestational age or at term they die within a few hours to a few days (10).

The prevalence of ABS varies across regions of the world from 1: 11,200 to 1:18000 births[6]. A much higher prevalence 1:1,200 found by Ossipof and Hall was related to miscarriages, stillborn and newborns inclusion. Geographic differences in prevalence could be a pointer to specific genetic interactions with environmental factors of affected populations and so should be further studied (12).

The risk factors associated with ABS include living at higher altitudes which might involve tissue hypoxia causing amniotic membrane damage (20,21). Familial occurrence with the risk higher in the first- and second-degree relatives have been observed and the firstborns are more likely to be affected (11,12). Medication drugs used in pregnancy such as such as misoprostol, and vaginal bleeding in the first trimester of pregnancy have also been implicated with higher prevalence of ABS. Low birth weights >2.5kg, intrauterine growth restriction and prematurity and non-cephalic presentations are more frequent with ABS cases than controls. Some studies from Boston, USA have associated cigarette smoking, use amphetaminophine, low socioeconomic class, low educational status, young maternal age, nonwhite, non-Hispanic ethnicity and unplanned pregnancies with higher prevalence of ABS. However, studies from Hungary found that multiparous women have greater risk association with age and reported positive association for low socioeconomic status, unplanned pregnancy and smoking during pregnancy in some variants of amniotic band sequence abnormality manifestation like ARS-L though this study has limitations of being medical records data entry rather than direct observation of cases in American study (12). This is the first of such complex multiple congenital abnormality due to ABS delivered alive at term and managed in our facility. The management in the 8 days of life was extremely challenging with both parents rejecting the baby and the complexities of multidisciplinary care with the team of experts struggling with limited resources for investigations and basic life support for the baby.

Oliemen Peterside, and colleagues had reported 2 cases of amniotic band sequence seen in Bayelsa State, Nigeria to highlight the existence of the disorder in that region and to highlight the difficulties in antenatal diagnosis and postnatal management[15]. Similarly in the index case highlights the existence of this pathology in Sokoto. We could not make the accurate diagnosis in the antenatal period and the postnatal management was equally challenging.

Ibrahim Aliyu et al, reported a case of upper limb constriction due to Amniotic band disruption syndrome in Kano, Nigeria to document the presence of the rare disorder in that region of Northern Nigeria and acknowledged more reports of the disorder in Southern Nigeria[16].

This index case was characterized by extreme complexity and severity of amniotic band sequence seen in a live birth. It is the first case of amniotic band sequence to be documented in Sokoto. It is also likely the first of its kind documented in the literature in Nigeria.

Conclusion

We have presented the first case of ABS managed in our institution. This index case was diagnosed at the antenatal period as exomphalos and only confirmed as ABS with very severe complex abnormalities at term delivery, considered not compatible with life but survived for 8 days in our care.

References

1. Seeds JW, Cefalo RC. Amniotic band syndrome. Am J Obstet Gynecol; 144:243
2. Human Malformations and Related Anomalies, 2nd ed, Stephenson RE, Ha II JG(Eds), Oxford University Press, Oxford 2006. P.871
3. Torpin R. Amniotic Mesoblasticfibrous Strings And Amniotic Bands: Associated Constricting Fetal Malformations Or Fetal Death. Am J Obstet Gynaecol 1965; 91:65
4. Hunter AG, Carpinter BF. Implications of malformations not due to amniotic band in the amniotic band sequence. Am J Med Genet1986; 24:691
5. Kalousek DK, Bamforth S. Amnion rupture sequence in previable fetuses. Am J Med Genet 1997:73:176.
6. Martinez-Frias ML. Epidemiological Characteristics of amniotic band sequence (ABS) and body wall complex(BWC):are they two different entities? AMm J Med Genet 1997; 73:176
7. Chemke J, Graff G, Hurwitz N. Liban E. The amniotic band syndrome. Obstet Gynecol
8. Higginbottom MC,Jones KL, Hall BD, Smith DW. The amniotic band rupture and variable spectra of consequent defects. J Paeditr 1979; 95544.
9. Van Allen MI, Curry C, Gallaher L. Limb body wall complex:1. Pathogenesis. Am j Med Genet 1987;28: 549
10. Van Allen MI, Curry C, Walden CE, et al. Limb and spine defects. Am J Med Genet 1987; 28 :249.
11. Pietro Cignini et al J Prenatal Med.2012 Oct-Dec; 6(4):59-63
12. Ossipoff V, Hall BD. Aetiologic factors in amniotic band syndrome: A study of 24 patients. March of Dimes Birth Defects: Orig Artic Ser xiil 1977; 3D:17117-132. [PubMed] [Google Scholar]
13. Castilla EE, Lopez-CameloJS.The surveillance ofbirth defects in South America. The search for time clusters: Epidemics. Adv Mutagens Res. 1990; 2:191-210. [Google Scholar]
14. . Czeizel AE, Vitez M Kodaj I, Lenz I. Study of associated apparent limb deficiency in Hungary,1975-1984. Am J Med Genet. 1993;46: 3732-378 [Pub Med] [Google Scholar]
15. Oliemen Peterside,James E Omietimi, Oyedeji O Adeyemi Amniotic band syndrome; a reportof two case sand a review of the literature.January 2013. IOSR Journalof Dental and Medical Sciences (IOSR-JDMS0) 5(3): 30-34.DOI:10.9790/0853-0533034
16. Ibrahim Aliyu, Umma Idris, Zainab Fumilayo Ibrahim. Amniotic band disruption syndrome: Our experience. LETTER TO THE EDITOR, Medical Journal of DR. D.Y. Patil Vidyapeeth. Year2018/Volume11/Issue 2/Page196-197 DOI: 10.4103/MJDRDYPU.MJDRDYPU_52_17