Amniotic Band Syndromewith Unique Clinical Presentations: A Case Report

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Abstract

Amniotic band syndromeis a rare congenital disorder which associated with physical abnormalities such as disruption, deformation, and malformations of organs. The most common abnormalities usually involve the limbs that could range from simple constriction rings to complete amputation.In this case report, we report anamniotic band syndrome with unique clinical presentations. The case was a new born male baby who was normally delivered and presented with a ring-like constriction at over middle right lower limb and fusion (syndactyly) of left lower limb and right arm. In this report we also discuss different diagnostic modalities which could be used in diagnosingamniotic band syndrome, the risk factors, type of amniotic band syndrome, post-natal diagnose, as well as the therapy.

Keywords*: Amniotic band syndrome, ABS, constriction rings, syndactyly, case report*

Introduction

Amniotic band syndrome (ABS) is a group of congenital birth defects believed to be caused by entrapment of fetal parts (usually a limb or digits) in fibrous amniotic bands while in utero.[1,2]ABS should be called a sequence rather than a syndrome because ABS is related in wide range of physical abnormalities, which are significantly disabling and disfiguring in nature.[1-5]

Most of infants with ABS have multiple deformity of the limbs and arms and could range from simple constriction rings to complete amputation. ABS has known as amnion rupture sequence, amniotic deformities/adhesions/mutilations(ADAM complex), amniotic band disruption complex, congenital constricting bands, terminal transverse defects, or Streeter anomaly.[1,2] Various studies estimate the incidence of ABS to be between 1 in 1,200 to1 in 15,000 living births.[3-5,6]There is no known clear etiology of ABS.[3-15] In this case report we report an ABScase with unique clinical presentations.

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Case Report

A 22 years old female, 28-29-week gestation, presented to Arifin Achmad Hospital withfirst stage of labor and preterm premature rupture of membranes (PPROM) andoligohydramnios.The patient was referred from a clinic with placenta previa. The patient complained the rupture of membraneas wet in the cloths about one day before admitted to the hospital; however, the patient felt this about a month already. The patient

also had regular contraction followed by blood mucus.

At the hospital, the patient was thoroughly examined including general as well asobstetric and gynecologic examinations. Patient vital signs were stable and no associated comorbidities such as maternal trauma or drug intake. Physical examinationsuggested minimal fluxus from ostium urethra externa, nitrazine test was difficult to interpret due to blood mucus. Ultrasonography showed oligohydramnioswith maximum vertical pocket (MVP)of 0.75 cm.

Patient was managed conservatively and was planned for cerclage and amnioinfusion. However, the patient was in labor and delivered a single alive newborn male baby with 1.4 kg, APGAR score was 7/6 and amniotic fluid was clear. On gross examination, the baby had ring-like constriction at over middle right lower limb and fusion (syndactyly) of left lower limb and right arm (**Figure 1A-C**). The baby was admitted to Neonatology Unit as sepsis and asphyxia and was thoroughly evaluated. Karyotyping and histopathologyexamination were ordered and returned with normalresults (**Figure 1D**).

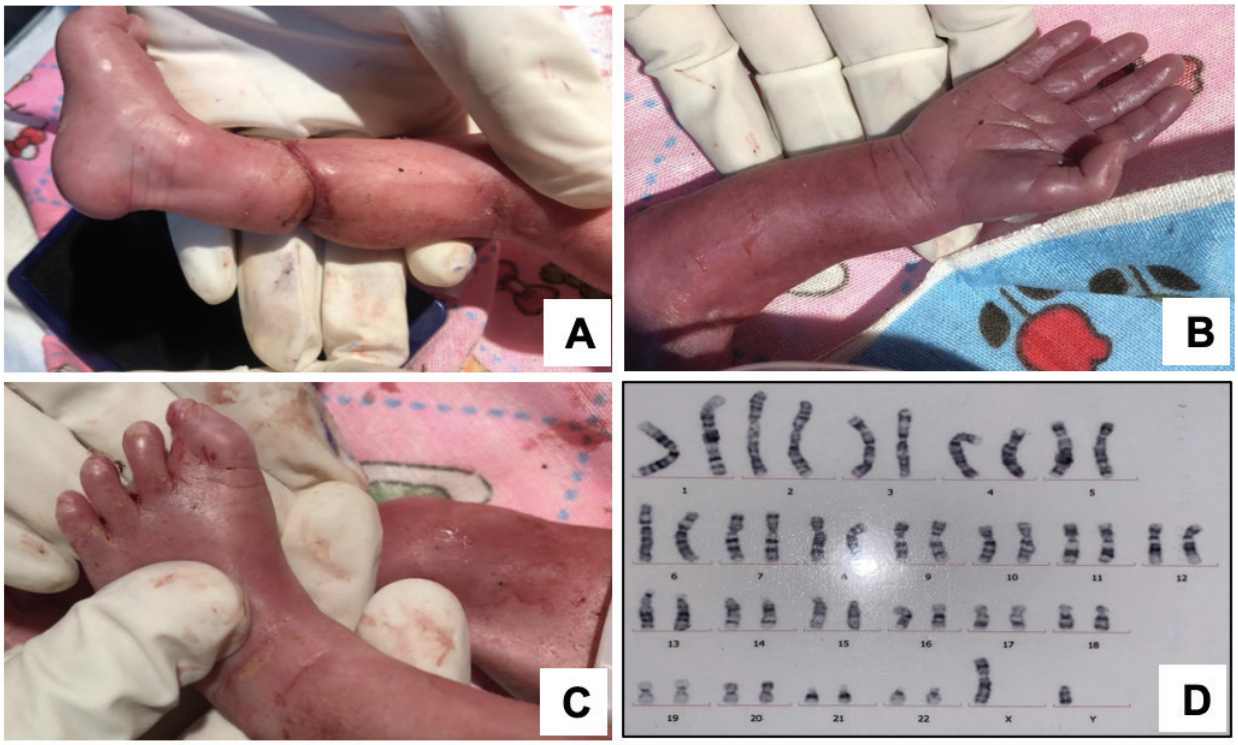

Figure 1. Clinical manifestation of the ABS case and itskaryotyping. (A) Constriction rings at over middle right lower limb. (B)Syndactyly of the right arm. (C) Syndactyly of left lower limb. (D) Karyotypingshowing normal result.

Discussion

ABS is a rare disorder with the estimated incidence between 1 in 1,200 to 1 in 15,000 living births.[11]ABS can be diagnosed prenatally by ultrasound and the most important ultrasound diagnostic features arevisible amniotic bands, constriction rings, irregular amputations of fingers and/or toes with a terminal syndactyly, or deformation of major anatomic. In a rare case, a strand of amniotic fibrous tissue could be seen attached to tissues and restricting the free movement of fetus in-utero. In this case, patient ultrasoundshowedno significant deformity since it was difficult to figure the extremities. Some studies found mild defects during ultrasound

examination;however,ABS is less likely to be diagnosed prenatally and most of defects were seen after birth.[6,16-20]

ABS etiopathogenesis is still unknown, but there are four main theories.[12]In this case, karyotyping result was normal (46 XY), the possibility cause is oligohydramnios. One of the well-known theories is rupture of the amnion in early pregnancy, with formation of amniotic band and liquid loss, or multiple loose strands (amniotic bands), followed by extrusion of all or parts of the fetus into the chorionic cavity. Bands entrap the parts of the growing fetus or limbs or other body parts become entangled and are subject to compression.

The abnormalities such as constriction rings and in some severe cases leading to vascular disruption andcould potentially result in amputation of the involved anatomic structures of the body parts.Adherence, even without constriction can have adverse mechanical effect that result in malformation or deformation. In this case, the baby was classified as group IV of ABS (isolated defect).[13] Other theories suggest that the amniotic band or vascular disruption due to genetic mutation.[8-10,13-17]

In such condition, all patients should receive counseling about the fetal abnormalities detected. Consulting with subspecialists is ideally recommended. There is no standard guideline of management for pregnancy complication with fetal ABS.Recently, some treatments for prenatal with ABS have been tried such as fetoscopic laser cuttingof amniotic bandsbefore their compression on the fetus.[19-20]However, the efficacy of the procedure is not well-documented.

Conclusion

ABS is a rare disorder caused by entrapment of fetal parts usually limbs or arms in fibrous amniotic bands while in utero leading to complex multisystem anomalies. The prenatal diagnosis is difficult although could be diagnosed by ultrasound which indirectly showing the ABS. The treatment and follow-up of ABS children require a team of specialists accordingly.

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Ethical Clearance: Written informed consent for publication was sought from the parent.

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