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

Sandeep Shrestha*1, Kamal Prasad Thani1, Uttara Gautam1, Nishan Bhattarai1, Annie Shrestha1, Munna Keshari1, Akina Sainju1, Santosh Lamichhane1, Pooja Thapaliya2

1Department of Paediatrics & Neonatology, Karnali Academy of Health Sciences, Jumla
2Department 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 intrauterine 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,
amputations of the extremities, and pseudosyndactylies, among other pathognomonic symptoms.\(^\text{11}\)

ABS etiopathogenesis is still unknown, but there are two main theories.\(^\text{12}\) 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.\(^\text{13}\) 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.\(^\text{14}\) 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.\(^\text{15}\) Craniofacial abnormalities have been reported as encephalocele, anencephaly, cleft lip and cleft palate whereas Thoraco-abdominal complications include omphalocele and gastroschisis.\(^\text{16}\)

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.\(^\text{17}\) 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.\(^\text{18}\)

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.\(^\text{8}\)

**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)

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