AMNIOTIC BAND SYNDROME WITH HOLOPROSENCEPHALY IN PALESTINIAN NEONATE: A CASE REPORT

Allam FM Abuhamda

"consultant paediatrician-neonatologist, the consultant of Palestinian ministry of health for neonatal intensive care units affairs in Gaza Strip, Gaza, Palestine. Qatar Red Crescent senior consultant neonatologist in Gaza Strip.

ABSTRACT Background: A newborn was diagnosed with a rare case of amniotic band syndrome which associated with the disfigured face, amputated limbs and holoprosencephaly. Case summary: He was full term and male. After birth, on physical examination, the baby found to have multiple constriction bands, amputated fingers, and toes. He had cleft lip and palate, malformed nose, hypertelorism, exophthalmos, red-sided absent eyelids, and low set ears and brain CT showed holoprosencephaly. Chromosomal karyotype was 46,xy: the cases were diagnosed as amniotic band syndrome with alobar and semi-lobar holoprosencephaly. Baby was kept on nasogastric Ryle feeding. He died at the age of 7 months. Conclusion: A new form between alobar and semi-lobar holoprosencephaly is the 1st time to be described in association with amniotic band syndrome.

KEYWORDS amniotic band syndrome, amputation, holoprosencephaly, disfigured face.

Introduction

The incidence of ABS is 1 in 1,200 to 15,000 live births worldwide [1]. Female and male are equally affected [2]. The mechanism of ABS is still unknown. There is two theory of this syndrome [3]. The extrinsic theory; amniotic band syndrome is that strands of tissue separate from the inner layer [amnion] of the amniotic sac. These floating bands in amniotic fluid could entangle different part of the fetus, arms, legs, fingers, and toes. If amniotic strands are still attached to amnion, it will affect the movement and development of the fetus. The other intrinsic [vascular] theory came to explain internal organ anomalies and ABS in case of intact amnion. The ABS in intrinsic theory is related to impaired blood flow to specific organs [4,5].

Case report

The parents were nonconsanguineous, they had no family history of congenital diseases, the mother was G1P1, the baby was full term male, and the delivery was by cesarean section.

His Birth weight was 2700 grams, and Apgar score was 7, nine at 1, 5 minutes respectively. After delivery on physical examination, the baby found to have: hypertelorism, malformed nose, right-sided eye exophthalmia and absent eye led, left-sided eye enophthalmos, cleft lip, cleft palate, prominent forehead [Figure 1], low set ears [Figure 2], macrocephaly, left hand had constriction rings around the thumb and ring fingers and amputation of the rest of fingers [Figure 3, 4].

The right hand had three fingers; there was tight constriction ring around the right arm [Figure 5]. The left foot had amputated 2nd toe and constriction ring around the fourth toe [Figure 6].

The other parts of physical examination were normal, and the baby had normal external male genitalia.

The baby was breathing comfortably in room air. Also hemodynamic system was stable. Baby tolerated orogastric feedings and passed stool.

Abdominal ultrasound and echocardiography studies were normal.

Brain CT showed a form between alobar [absence of brain lobe] and semi-lobar holoprosencephaly [Figure 7, 8].
Karyotyping was normal 46 XY chromosomes  
The baby died at the age of 7 months old in Shifa neonatal intensive care unit.  
(Figure 7 and eight brain CT showed a form between alobar and semi-lobar holoprosencephaly)

**Discussion**

This is a severe form of amniotic band syndrome; the baby had disfigured face, holoprosencephaly, amputated fingers, constriction rings and malformed hand. Holoprosencephaly is a cephalic disorder in which the prosencephalon fails to develop into two hemispheres. A rare form of holoprosencephaly is known to be associated with ABS [6], but this is the first time to describe a form between alobar and semi-lobar holoprosencephaly associated with ABS. The baby had significant congenital anomalies related to brain, face and limbs which make him incompatible with life. The multidisciplinary team; neonatologist, pediatric surgeon, pediatric neurologist and pediatric neurosurgeon decided after the discussion with family that surgical intervention
Figure 4: Left hand had constriction band around the thumb and ring fingers and amputated the rest fingers.

Figure 5: The right hand had three fingers; there was tight constriction ring around the right arm.

Figure 6: The left foot had amputated 2nd toe and constriction ring on the fourth toe.

Figure 7: Brain CT.
and invasive management are not advisable DNR (do not resuscitate) status was agreed with family.

The baby till the age of 7 months old, he was active, he had severe hydrocephalus (head circumference 80 cm), he was hemodynamically stable, he breathed comfortably in room air, he tolerated orogastric feeds, and he passed stool. The baby at the age of 7 months old suddenly arrested and died.

The pathophysiological mechanism of ABS is still controversial between extrinsic and intrinsic theory. Extrinsic theory in this baby could explain the separated amniotic strands, and it was the responsible for constriction rings, and it’s a complication. In same time holoprosencephaly and malformed right hand were unexplained by extrinsic theory.

**Conclusion:**

A new form between alobar and semi-lobar holoprosencephaly is the 1st time to be described in association with ABS. More studies are needed to understand the pathophysiological mechanism of the amniotic band syndrome.

**Acknowledgement**

To Dr. Wajdy Jarbou, consultant radiologist, the head of CT department in Shifa hospital, MOH, Gaza, Palestine.

**Authors’ Statements**

**Competing Interests**

The authors declare no conflict of interest.

**References**

1. Amrutha, K. V., et al. "AMNIOTIC BAND SYNDROME—A HOSPITAL BASED FETAL STUDY." Int J Biol Med Res 7.1 (2016): 5421-5425.

2. Richardson, Sunil, Rakshit Vijay Khandeparker, and Philippe Pellerin. "Amniotic constriction band: a report of two cases with unique clinical presentations." Journal of the Korean Association of Oral and Maxillofacial Surgeons 43.3 (2017): 171-177.

3. Cortez-Ortega, Carolina et al. “Management of the Amniotic Band Syndrome with Cleft Palate: Literature Review and Report of a Case.” Case Reports in Dentistry 2017 (2017): 7620416. PMC. Web. 27 May 2018.

4. Bora, Bhagyashri, R. Ravishankar, and Priti Mhatre. "Amniotic band syndrome-A Case Report." Pravara Medical Review 8.2 (2016).

5. Abdel-Rahim, Shady, and Kevin Chung. "Amniotic Band Syndrome 22." Congenital Anomalies of the Upper Extremity: Etiology and Management (2014): 295.

6. Das G, Gayen S, Bandyopadhyay S, Das D. Ethmocephaly with Amniotic Band Syndrome. Middle East African Journal of Ophthalmology. 2012;19(4):429-431. doi:10.4103/0974-9233.102769.