JOURNAL OF RANGAMATI MEDICAL COLLEGE

Case Report

Volume 3, Issue 2, July 2020; 25-27

Acardiac Amorphous : A Unique Bizarre Congenital Anomaly

Nasima Begum[1*] Shoyela Shahnaz[2] Sharmin Nahar Bashar[3] Nargis Sultana[1]

ABSTRACT

Acardiac amorphous is a rare and bizarre complication of monochorionic twin pregnancy. We report a case of acardiac amorphous diagnosed during an emergency cesarean section. The condition results from abnormal placental vascular anastomoses termed as Twin Reversed Arterial Perfusion (TRAP) theory. A 26 years 2nd gravid woman presented at 33 weeks pregnancy with preterm labor pain. By cesarean section, a premature female baby was delivered weighing 1.7kg. A quadrangular big deformed mass, covered with normal skin with rudimentary lower limbs without cardiac pulsation and facial structures was connected to placenta with a thin- walled umbilical cord was delivered along with. A rare fetal monster without a functional heart with no resemblance to a human embryo. Timely diagnosis and appropriate management improve the survival of the normal twin.

Key words: Monozygotic twins; Congenital abnormality; Pregnancy; TRAP.

INTRODUCTION

Monozygotic twin comprises 20% of twin pregnancies and 75% of them share the same placenta. Most monozygotic monochorionic twins are characterized by varying degrees of circulatory anastomoses. Arterio-arterial anastomoses predominate in 75% of monochorionic placentas compared to vein-vein or artery-vein[1]. Consequently, the presence and persistence of hemodynamic imbalance in two fetal circulations may result in fetal-fetal transfusion disorders such as the Twin Reversed Arterial Perfusion (TRAP) sequence[1,2]. TRAP sequence or acardiac twin is the most extreme fetal-fetal transfusion disorder. One twin is called acardiac twin or TRAP fetus, lacks a heart. The other normal twin with a functioning heart pumps blood through both fetal circulations (Called pump twin). Pump twin is anatomically normal but may suffer from congestive cardiac failure. Due to the absence of the heart, acardiac twin does not send blood to any portion of the placenta, and all its blood supply is received from and goes back to the circulations of pump twin through a unique vascular connection on the surface of shared placenta. Artery usually carries blood away from the fetus and towards the placenta to receive oxygen from the mother's circulations[1-3]. When there is a TRAP sequence, the unique vascular connections allow blood in the artery to follow in reversed direction- toward the acardiac fetus rather than away from it. Thus the term Twin Reversed Arterial Perfusion (TRAP) sequence has been used to describe this condition. Development of acardiac twins is limited to the lower part of the body supplied from the pump twin and failure of development thoracic organs, upper limb, and facial structures[1,3].

CASE REPORT

Twenty-six years (2nd gravida , Para-1+0) an otherwise healthy woman- came on 10th April 2020 at her 33+ weeks of pregnancy for an antenatal check-up. She had only one ultrasonogram (USG) done at 3rd month of her pregnancy revealing, 13 weeks alive twin pregnancy. She had no other antenatal check-up afterward. On physical examination, the fetal size was large for the gestational duration without any apparent systemic disorder. She was advised for ultrasonography to see fetal wellbeing and any other pathology. The next day while doing her tests, preterm labor pain began, and she got admitted. USG revealed, 'Single alive fetus of 33 weeks of gestation with a breech presentation

1. Assistant Professor of Obstetics & Gynaecology
Rangamati Medical College, Rangamati, Bangladesh.
2. Assistant Professor of Obstetics & Gynaecology
Chittagong Medical College Hospital, Chattogram, Bangladesh.
3. Junior Consultant of Obstetics & Gynaecology
Chittagong General Hospital, Chattogram, Bangladesh.

*Correspondence to:
Dr. Nasima Begum
Mobile : +88 01715 02 73 50
Email: nasimacmc@yahoo.com

Date of Submission : 03-03-2020
Date of Acceptance : 09-04-2020

Volume 3, Issue 2, July 2020; 25-27

with another heterogeneous mass (17.9cm × 14.5cm) with multiple calcified structures without any heart suggestive of acardiac amorphous within a single amniotic cavity with single placenta'. An emergency caesarian section was done. At first, a premature normal-looking female baby was delivered. Amniotic fluid was more than average. After cutting the cord, placenta was separated and removed.

Figure 1 : Acardiac twin without face & upper limb limblimb.

Figure 2 : Acardiac twin with small and narrow umbilical cord.

Figure 3 : Acardiac twin- no resemblance to human foetus.

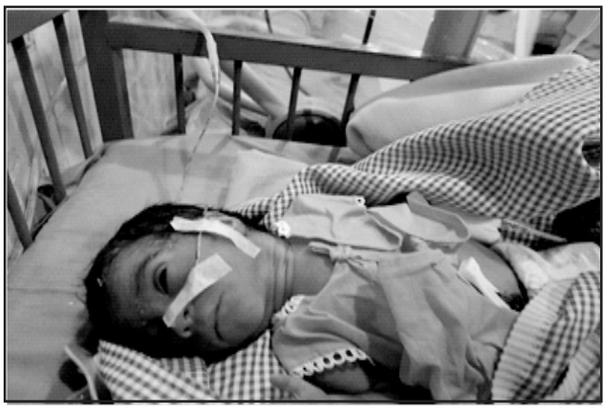

Figure 4 : Co-twin (Female) with all normal structures - after birth.

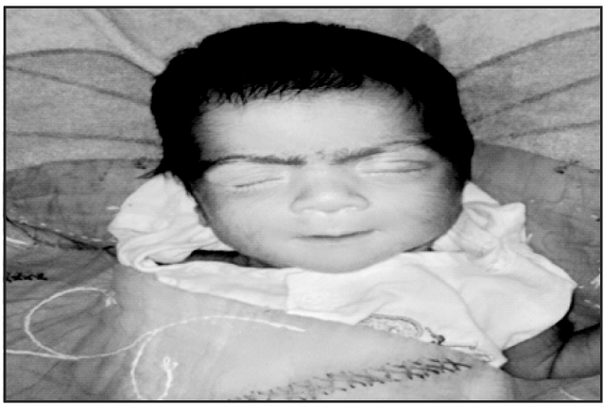

Figure 5 : Normal co-twin after one month.

A big irregular mass (20cm × 15cm) covered with skin- was found in the same amniotic cavity (Fig 1,2,3). It was delivered by application of pressure by the surgeon and assistant. The mass was misshapen with a narrow small umbilical cord attached in the middle, was separated beforehand spontaneously by itself. Rudimentary upper & lower limbs were present, head, face, neck were absent. The normal baby was weighing 1.7 kg & got admitted to neonatal intensive care unit. The newborn was doing well and found normal with X-ray & other investigations and discharged after 2 days (Fig 4,5).

DISCUSSION

The development of an acardiac twin is a rare entity with an unknown etiology. It occurs in 1 in 100 monozygotic twin pregnancies and 1 in 35,000 births[1-4]. Perinatal mortality of the normal twin may be as high as 50%[1,3,5-7]. Various type of acardiac twin ranges from the most developed acardiac anceps to the least developed acardiac amorphous[1,7,9,10]. The most widely accepted theory on the pathogenesis of occurrence of the acardiac twin is TRAP sequence. The less

Volume 3, Issue 2, July 2020; 25-27

popular theory is dysmorphogenesis which defines the underlying pathology as a defect in early organogenesis, resulting in a fatal malformed cardiac system[4-6]. Some researchers explain that compression of the cephalic pole of the embryo inhibiting curving and fusion of primitive heart tube is the basic pathology resulting in non-formation of heart and other organs[10].

However, whether this TRAP sequence is the cause or effect of acardia is still unknown. The acardiac twin essentially becomes a parasite, putting the surviving twin in high-output cardiac failure in most cases. TRAP holds perinatal mortality as high as 50-55% [5,6,8,9]. Though arterial anastomosis contributes to reversal blood, it is not responsible for acardia.

The arterio-arterial anastomosis concept describes a reversed umbilical artery blood flow as the principal cause of this condition. Anastomosis causes deoxygenated arterial blood to flow from the donor to the acardiac twin. Unfortunately, circumventing the placenta results in a lack of perfusion for vital organ development, leading to the recipient twin appearing a heterogeneous mass rather than a properly developing fetus[3-5]. Four types of acardia are classified based on the presence or absence of various limbs and organs. The most common type acardiac acephalus is the absence of the head, upper extremities, and trunk but maintenance of lower limbs, genitalia, and abdominal organs[6,7]. In acardiac anceps, the most developed type, facial features, and cranial structures are present but incomplete rudimentary limbs and organs. The third type described in this case is the least differentiated one-acardiac amorphous, the recipient twin-an ill-defined mass of tissue with no head and organs with rudimentary extremities, the fourth type, and least type acardiac acormus the head is the only formed structure with no limb or organs[5,7-9].

An antenatal intervention involves fetal surgery that obstructs blood flow to the acardiac mass via endoscopic ligation, or Laser coagulation of the umbilical cord, bipolar cord coagulation or intrafetal radiofrequency ablation. Intrafetal Laser treatment is recommended at less than 16th of weeks pregnancy, whereas radiofrequency ablation is preferred if greater than 16th weeks due to higher blood flow[4,7,10]. All methods can be performed under local anesthesia. Survival of donor twin in utero after treatment is 80-90%.

Chance of maternal complications is very less, there is still risk of hemorrhage and chorio-amnionities[7,8].

CONCLUSION

As a unique and extreme complication, TRAP sequence needs a thorough clinical and sonographic evaluation as well as high index of suspicion in monochorionic multi-fetal gestation. Despite the rarity of TRAP sequence with proper surveillance and intervention morbidity and mortality can be significantly reduced to yield a normally developed child.

DISCLOSURE

Al the authors declared no competing interest.

REFERENCES

1. Otieno PO, Kosgei W, Keitany KK. Acardiac Twin: The Overlooked Diagnosis. A Recent Case Report and Review of Literature. Gyne Obs Case Rep. 2020; 6 (2): 90.

2. Ray S, Basak S, Ghosh PC, Pal M, Laga M, Pradhan M. Acardius Acephalus: "Headless- Heartless Nine-month Life". J Obstet & Gynaecol India. 2012; 62 (Suppl- 1):16-18.

3. Bhat DM, Butale PR, kumbhalkar DT, Raut WK. Fetus acardiac amorphous: a rare case report. Int J of Reprod Contracept Obstet and Gyneco1. 2017; 6(10) :4686-46-89.

4. Mann S, Chauhan MB, Malik R, Nanda S, Malhotra V. Acardiac Monster at Term-A rare Entity. Journal of Gynecologic Surgery. 2010. 26(4): 275-276.

5. Blaicher W, Repa C, Schaller A. Acardic twin pregnancy: associated with trisomy 2: Case report. Human Reproduction.2000; 15(2) : 474-475.

6. Krishna LG, Uppoor R, Shankar JS. Amorphous acardiac parabiotic twin. Indian J Radiol Imaging. 2005; 15(2) : 267-270.

7. Joseph R, Anderson S,. The Case of the Vampire Baby Case Report: Twin Reversed Arterial Perfusion Sequence. J Fam Med. 2016; 3(7): 1076.

8. Athwal S, Millard K, lakhoo K. Twin reversed arterial perfusion(TRAP)sequence in association with VACTERL association: a case report. J Med Case Reports. https://doi.org/10.1186/1752-1947-4-411.

9. Nigam A, Agarwal R, Saxena P, Barla J. Acardiac anceps: A rare congenital Anomaly. BMJ Case Rep. 2014. doi: 10.1136/bcr-2013-202583.

10. Cleary-Goldman J, D'Alton ME. Physiological Effects of Multiple Pregnancy on Mother and Fetus. In. Polin RA, Fox WW, Abman SH, eds. Fetal and Neonatal Physiology. 3rd ed. Philadelphia: Saunders, 2004. 165-178.