Acardiac Twin

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Abstract

Acardiac anomaly is a rare complication of multiple pregnancies. It is a hemodyamically disadvantaged nonviable twin which occurs in association with a twin reversed arterial perfusion sequence (TRAP). In TRAP, blood flows from an umbilical artery of the pump twin in reverse direction into umbilical artery of the perfused (or acardiac) twin via an arterial to arterial (AA) anastomosis. Its blood is poorly oxygenated and results in variable degrees of deficient development of the head, heart, and upper limb structures. The lower half of the body is usually better developed, which may be explained by the mechanism of perfusion. The pump twin is at risk of heart failure and problems related to preterm birth with a reported mortality of 50-75%. We present a case of multiple pregnancy with acardiac twin in a 26 year old lady with gestational diabetes mellitus. During ante natal check up several ultrasonographic (USG) screening showed twin pregnancy with one healthy fetus and the other fetus with inconspicuous anatomy and structure. Repeat USG at 36th week of gestation diagnosed twin pregnancy with acardiac twin. Strikingly the pump twin did not develop the usual complications and was delivered by cesarean section at 37th week of gestation.

Keywords: *Acardiac; twin.*

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Introduction

The development of the acardiac anomaly is a rare complication of monozygotic multiple pregnancies.[1] It was first described in the sixteenth century and occurs in 1% of monozygotic twin pregnancies and in one out of 35,000 deliveries.[2,3] The most widely accepted theory on the pathogenesis of acardius is the occurrence of a twin reversal arterial perfusion sequence (TRAP).[4] TRAP refers to a rare, unique complication of monochorionic twin pregnancies in which a twin with an absent or a non functioning heart (acardiac twin) is perfused by its co-twin (pump twin) via placental arterial to arterial (AA) anastomoses.[3,5] The acardiac twin usually has a poorly developed heart, upper body and head.[6] The pump twin is at risk of heart failure and problems related to preterm birth.[3,5] Two criteria must be fulfilled for the development of a TRAP sequence. The first one is an anastomosis and the second is discordant development or in utero demise of one of the

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monochorionic twins, allowing for the blood flow reversal. Not infrequently, chromosomal abnormalities are identified in acardiac twin, whereas the pump has a normal karyotype. The diagnosis can be reliably made on ultrasound scan in the first trimester.[7,8] TRAP sequence is characterized by a grossly abnormal fetus that grows, may even show movements, but has no functional cardiac activity of its own. There are four types of acardiac twins. In the acardiac-acephalus, the thoracic organs and the fetal head are absent. In the acardiac-acromas only the fetal head develops. The acardiac-amorphous consists a mass of tissue without recognizable human parts. Finally in the acardiac-myelacephalus the head and one or several extremities develop normally.[9]

Case Report

A 26 year old lady G2P1 was referred to Bangladesh Institute of Research & Rehabilitation in Diabetes, Endocrine and Metabolic Disorder (BIRDEM), Dhaka, Bangladesh at her 34th week of twin pregnancy with a history of caesarean section. Several (5 times) prenatal ultrasonographic (USG) screening showed one fetus with inconspicuous anatomy and structure. There were no signs of decompensation and biometry corresponded to 34 weeks of gestation. She was detected as a case of gestational diabetes mellitus (GDM) 7 days back. Her blood group was 'B' (+)ve. Routine investigations of pregnancy and coagulation profiles were done, which were within normal limits. She was treated with bed rest and close fetal monitoring. Her GDM was well controlled with diabetic pregnancy diet and insulin. A repeat USG was done on 36th week which showed one healthy fetus of 2.4 kg but chest and abdomen of the other fetus could not be identified. Head of the second fetus was deformed, bowel loops were floating in amniotic fluid and biparietal diameter (BPD) and abdominal circumference (AC) could not be measured. Placenta was single, large and fundal. Amniotic fluid was more than adequate (21cm). Caesarean section was done at her 37th week of gestation due to previous history of caesarean section. A healthy male baby with normal APGAR score[10] was delivered. The umbilical cord had three vessels, the birth weight was 2.5 kg, the length was 48 cm and the infant had no sign of decompensation. The acardiac twin with a single umbilical artery was delivered in a separate amniotic sac, together with the monochorionic placenta. The acardiac twin was 15 cm in length, showing the rudimentary head with cephalocele, omphalocele and its pelvis and the upper and lower limbs were poorly developed. The mother's postoperative period was uneventful. She was discharged on the 7th postoperative day with a healthy male baby and was advised to come for follow up after 7 days.

Fig 1: The acardiac twin

Fig 2: Acardiac twin with its healthy co-twin

Discussion

In the TRAP sequence, the normal twin 'pumps' or 'donates' blood to the abnormal twin, which is called the 'recipient' or 'perfused' twin through abnormal artery-to-artery communications in the placenta.[5] There is a reversal of flow in the recipient twin, with relatively oxygenated blood flowing from the abnormal anastomosis to the umbilical artery; the flow then proceeds cranially, leaving the fetus via the umbilical vein; hence the term, TRAP sequence. This finding can be confirmed by pulsed Doppler of the umbilical artery of the recipient twin, which will reveal reversal of flow on the spectral waveform. In 75% of cases, the umbilical cord of the recipient twin contains a single umbilical artery[11], which was also seen in this case.

As a result of imbalance of the interfetal circulation, the caudal aspect of the perfused fetus receives blood with relatively more nutrients and oxygen than the upper torso, resulting in better development of the pelvis and lower extremities in the acardiac fetus. Fully desaturated blood then flows in a retrograde fashion to the upper body and head, leading to maldevelopment of the heart, head, and upper torso, which may be either completely absent or severely deficient.[5] This case was with these typical

features. Therefore, on USG it appears as a heterogeneous mass, simulating a teratoma or intrauterine fetal demise.[12] The acardiac twin usually has a dorsal cystic hygroma[13] but in this case it was not present.

The etiology of cytogenic discovery in TRAP twins is unclear. Polar body fertilization was described in an acardiac twin pregnancy, but zygosity testing excluded polar body fertilization as a likely cause of TRAP.[14,15] The development of acardiac fetuses in one of a pair of dizygotic twins in certain animal species who share anastomoses after fusion of two separated placenta has been described and these could be equivalent pathomechanism in humans.[16,17]

The principal perinatal problems associated with acardiac twinning such as pump twin congestive heart failure, maternal hydramnios and preterm delivery did not occur in our case. Damani reported a similar case with good perinatal outcome of the healthy co-twin without any antenatal intervention.[18] In a case series by Chandramouly and Namitha[3], they described two cases of acardiac twin with different outcomes. The first patient went into premature labor secondary to polyhydramnios at 27 weeks of gestation and delivered an amorphous mass and a normal-appearing fetus which died soon thereafter. The second patient was followed-up with serial USG examinations and she had a successful outcome, with the delivery of a normal fetus and an amorphous mass at term.

Chromosomal anomalies may be present in up to 50% of cases of acardiac fetus.[2] Chromosomal analysis of acardiac twin showed trisomy 2. Only a few abortion in the late first trimester have been reported and no case of trisomy 2 in which the fetus survived into the second trimester has been described.[19] Chromosomal analysis of the acardiac fetus was not done in our case, but to evaluate the impact of aneuploidy in acardiac twin pregnancies, cytogenic investigations should be carried out routinely.

The acardiac twin is non viable and the majority of efforts in management are focused on maintaining viability of the donor twin including close surveillance for development of hydrops. Several treatment modalities are practiced in these cases including palliative treatment which involve prolongation of pregnancy by serial amniodrainage and maternal administration of indomethacin for preterm labour.[7] Prenatal treatment involves occlusion of blood flow to the acardiac twin by endoscopic (fetoscopic) ligation or laser coagulation of the umbilical cord, bipolar cord cauterization, or intrafetal radiofrequency ablation.[11,20] The indications for prenatal treatment include polyhydramnios, cardiac dysfunction, hydrops of the pump twin, or a relatively large weight of the acardiac twin.[13]

Though none of these modalities were available or tried in our case, we could achieve a good outcome only by monitoring the patient with serial USG and termination at term.

References

1. Søgaard K, Skibsted L, Brocks V. Acardiac Twins: Pathophysiology, Diagnosis, Outcome and Treatment. Six Cases and Review of the Literature. Fetal Diagn Ther. 1999;14(1):53-59.
2. Barth RA, Crow HC. Ultrasound Evaluation of Multifetal Gestations. In: Callen PW, editor. Callen Ultrasonography in Obstetrics and Gynaecology. 4th ed. Pennsylvania: W B Saunders; 2000.p.196-98.
3. Chandramouly M, Namitha. Case Series: TRAP Sequence. Indian J Radiol Imaging. 2009;19(1):81-83.
4. Haring DAJP, Cornell MC, van der Linden JC, van Vugt JM, Kwee ML. Acardius Acephalus after Induced Ovulation: A Case Report. Teratology. 1993;47(4):257-62.
5. Levi CS, Lyons EA, Martel MJ. Sonography of Multifetal Pregnancy. In: Carol M, Rumack, editors. Diagnostic Ultrasound. Vol 2. 3rd ed. Missouri: Elsivier Mosby; 2005.p.1207-1209.
6. Saxena R. Evidence Based Color Atlas of Obstetrics & Gynaecology: Diagnosis and Management. New Delhi: Jaypee Brothers Medical Publishers; 2013.p.189.
7. Lewi L, Deprest F. Fetal Problems in Multiple Pregnancy. In: James DK, Steer PJ, Weiner CP, Gonik B, editors. High Risk Pregnancy Management Options. 3rd ed. New Delhi: Saunders; 2006.p.524-60.
8. Coulam CB, Wright G. First Trimester Diagnosis of Acardiac Twins. Early Pregnancy. 2000;4(4):261-70.

9. Echeverry CJ. Multiple Gestation. In: Arias F, Daftary SN, Bhide AG, editors. Practical Guide to High Risk Pregnancy & Delivery A South Asian Perspective. 3rd ed. New Delhi: Elsevier; 2008.p.293-322.
10. Haddad GG, Green TP. Diagnostic Approach to Respiratory Disease. In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF, editors. Nelson Textbook of Pediatrics. 19th ed. Philadelphia: Saunders Elsevier; 2011.p.366.
11. Malone FD, D'Alton ME. Anomalies Peculiar to Multiple Gestations. Clin Perinatol. 2000;27:1033-46.
12. Monteagudo A, Roman AS. Ultrasound in Multiple Gestations: Twins and Other Multifetal Pregnancies. Clin Perinatol. 2005;32:329-54.
13. Egan JF, Borgida AF. Ultrasound Evaluation of Multiple Pregnancies. In: Callen PW, editor. Ultrasonography in Obstetrics and Gynecology. 5th ed. Pennsylvania: W B Saunders; 2008.p.86-88.
14. Chaliha C, Schwarzler P, Booker M, Battash MA, Ville Y. Trisomy 2 in an Acardiac Twin in a Triplet in-Vitro Fertilization Pregnancy. Hum Reprod. 1999;14(5):1378-80.
15. Fisk NM, Ware M, Stanier P, Moore G, Bennett P. Molecular Genetic Etiology of Twin Reversed Arterial Perfusion Sequence. Am J Obstet Gynecol. 1996;174(3):891-94.
16. Bieber FR, Nance WE, Morton CC, Brown JA, Redwine FO, Jordan RL, Mohanakumar T. Genetic Studies of an Acardiac Monster: Evidence of Polar Body Twinning in Man. Science. 1981;213(4509):775-77.
17. Benirschke K. The Biology of the Twinning Process: How Placentation Influences Outcome. Semin Perinatol. 1995;19:342-50.
18. Damani M. A Rare Case of Acardiac Twin. Journal of South Asian Federation of Obstetrics and Gynaecology. 2011;3(1):46-48.
19. Benirschke K, Kaufmann P. Pathology of the Human Placenta. 2nd ed. New York: Springer; 1995.p.702-10.
20. Wong AE, Sepulveda W. Acardiac Anomaly: Current Issues in Prenatal Assessment and Treatment. Prenat Diagn. 2005;25(9):796-806.