CASE REPORT

Case Report on Conservative Management of Acardiac Acephalus

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Abstract

Background: Acephalus is a rare complication of multiple gestations with an incidence of 1 in 35,000 pregnancies and 1% of monochorionic twins.

Case description: A 21-year-old primigravida with a twin pregnancy with one acardiac acephalus, with ultrasonography (USG) s/o twin gestation approximately 25.3 weeks along with the presence of hydropic acardiac twin corresponding to 23 weeks and the presence of rudimentary spinal elements with no skull and upper extremities was managed conservatively at a tertiary care center with routine fetal monitoring using USG and Doppler ultrasonography.

Conclusion: Early and accurate antenatal diagnosis is very important in order to prevent the pump twin from developing its usual complications. Both conservative and interventional management approaches are available which varies from patient to patient depending on various factors.

Keywords: Acardiac acephalus, Monozygotic monochorionic, Twin reversed arterial perfusion.

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Background

Acephalus is an uncommon complication of multiple gestations with an incidence of 1 in 35,000.[1] Twin reversed arterial perfusion (TRAP) was previously known as an acardiac twin. The characteristic feature of this syndrome is that the pump twin is normally developed while the other is acardiac twin, i.e., a twin with absent cardiac structure, which is dependent on the normal pump twin[2] for its hemodynamic requirements. As a result of imbalance in the blood supply, the lower torso in the recipient twin is well perfused compared to the upper torso. As there is an increase cardiac workload in the pump twin, it is at risk of heart failure. Other complications such as cord entanglement and preterm birth have also been reported with a mortality rate of 50–75%. Other poor prognostic indicators include polyhydramnios, hydrops fetalis, and respiratory distress syndrome. Early antenatal diagnosis is very important in order to salvage the pump twin from its trivial complications. Both conservative and interventional management approaches are available which vary from patient to patient depending on various factors such as size of the acardiac twin morphology, cardiac status of the pump twin, and the relationship between pump and acardiac twin. Therefore, before selecting the most suitable therapy, all these factors must be taken into consideration. We are presenting a case of conservative management of acardiac twin pregnancy tertiary care center.

Case Report

A 21-year-old Hindu female, primigravida with monochorionic twin pregnancy with one acardiac acephalus, registered in our antenatal outpatient department (OPD) at 22 weeks of gestation. Her first ultrasonography (USG) was s/o twin gestation approximately 25.3 weeks with EFW 760 g with posterior placenta with changing presentation along with the presence of hydropic acardiac twin measuring 9.7 × 5 × 6.6 cm weighing approximately 205 cc with femur length corresponding to 23 weeks and the presence of rudimentary spinal elements with no skull and upper extremities.

Several antenatal ultrasonographic scans and Doppler were done at regular intervals to monitor the fetal growth throughout pregnancy. These routine USG scans showed adequate fetal growth with adequate uterine artery flow. Acardiac twin was also seen to increase in size. There were no signs of cardiovascular decompensation in her last Doppler study and fetal biometry corresponded to 33 weeks of gestation with EFW 2036 g and UA PI 0.6 with mild polyhydramnios and the presence of choroid plexus cyst in the left ventricle with acardiac twin measuring 880 g. Her other routine investigations and coagulation profiles were within the normal range. Elective lower segment Cesarean section was done at 37 weeks i/v/o first fetus with breech presentation. She delivered a healthy female child with no evidence of gross congenital anomaly and a birth weight of 2.9 kg (Fig. 1A) and a normal APGAR score. The other twin was a fleshy mass without any head-like structure weighing 884 g (Fig. 1B). The placenta was monochorionic with a normal gross appearance and placental weight of 500 g (Fig. 2). The mother had an uneventful postoperative period and was discharged on day 5 postoperatively day with a healthy female baby and was asked to follow-up after 7 days.

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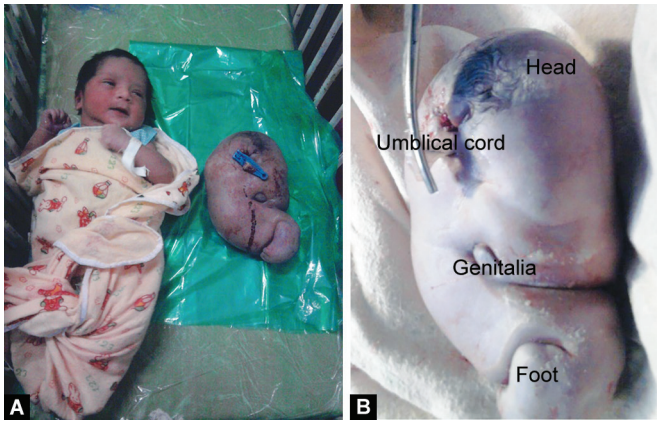

Figs 1A and B: (A) Left side: female fetus, right side: acardiac acephalus; (B) Acardiac acephalus

Fig. 2: Monochorionic placenta

Discussion

Acardiac acephalus is commonly referred to as TRAP sequence and one of the uncommon complications of monozygotic monochorionic twin pregnancies. It was first described by Grunewald in 1942.

Aetiopathogenesis

There are several hypotheses proposed to explain the development of this anomaly. Firstly, during embryonic development, there is a delayed separation of the inner cell mass into two which is followed by the development of anastomosis in the vessels of the placental bed, such that the two circulatory systems of the twin remain interconnected.[3] Thus, as a result of the difference in the pressure of the blood flow there is development of retrograde perfusion, leading to a developmental arrest in one of the twins which is completely depended on the pump twin for its survival[3] resulting in the formation of acardiac cephalus. Secondly, Fusi et al. reported that, due to thrombotic phenomenon occurring in the thoracoabdominal vessels, there is resorption of the affected tissues that result in the development of acardiac twin. In addition, chromosomal aberrations have also been associated with the development of acardiac fetus. TRAP sequence can be diagnosed earliest by the 9th week of gestation using grayscale USG and Doppler ultrasonography. Serial sonograms show the presence of grossly malformed fetus with an amorphous fetal pole with absent cardiac pulsation and poorly defined fetal trunk as in the present case. Arterial blood flow toward the affected fetus on color Doppler is diagnostic of TRAP sequence. The key factors in the management of the pregnancy depend on the degree of cardiac involvement in the pump fetus, duration of pregnancy, and the survival of the normal co-twin. Majority of the patients can be conservatively managed, but in some, minimally invasive techniques to interrupt the vascular anastomosis,[4] in order to ameliorate the outcome in the pump twin, can be used.

Conservative management is recommended in patients where there is no sign of cardiovascular compromise in the pump twin and acardiac twin is small in size. The main aim of the management here includes monitoring for the development of complications in the pump twin. The patient should be regularly followed up at 2 weekly intervals with USG and Doppler ultrasonography. Fetal 2D ECHO, biophysical score (BPP) and non-stress test (NST) should be done in all cases.

In utero treatment modalities include inotropic drugs, e.g., digoxin, to treat heart failure; while amniotic fluid drainages or indomethacin treatment can be performed to improve the prognosis in the surviving twin by reducing the amount of amniotic fluid volume which in turn is beneficial in preventing premature labor. Treatment modalities to occlude the circulation to the acardiac twin such as endoscopic ligation of anastomotic vessels, coagulation of anastomotic vessels with neodymium-doped yttrium aluminum garnet (Nd: YAG) laser, bipolar cord cauterization, radio frequency ablation, and with absolute alcohol have also been advised by few.

Conclusion

Acardiac acephalus is one of the rare complications of monozygotic monochorionic twin gestation. The present case of acardiac acephalic twin is the most common form of all acardiac twins where a lower trunk was partially developed with a totally absent upper half of the body. Early antenatal diagnosis is very important in order to salvage the pump twin from its trivial complications. Imaging techniques such as 3D ultrasonography and Doppler ultrasonography can be used in the first trimester to confirm the diagnosis. In some cases, a minimally invasive technique to interrupt the vascular anastomosis, in order to ameliorate the outcome in the pump twin can be used.

References

1. Barth RA, Crow HC. Ultrasound evaluation of multifetal gestations. In: Callen PW, editor. Callen ultrasonography in obstetrics and gynaecology. 4th ed. Pennsylvania: WB Saunders; 2000. p. 196–198.
2. Prasad RH, Prasad TR, Kumar KD. TRAP sequence-an interesting entity in twins. J Clin Imaging Sci 2012;2:56. DOI: 10.4103/2156-7514.100997.
3. Saritha S, Sumedha S, Anjankar VP. Twin reversed arterial perfusion sequence (TRAP Sequence). The acardiac/acephalic twin. Int J Anat Res 2013;03:140–144. ISSN: 2321-4287.
4. Stamatian F, Muresan D, Caracostea G, et al. Advances in ultrasonic assessment of acardiac twin. Donald School J Ultrasound Obstet Gynecol 2011;5(3):213–218. DOI: 10.5005/jp-journals-10009-1198.