Case Report

Acardiac twin - A case report

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Abstract

Acardiac twinning is a rare congenital anomaly of monozygotic twin pregnancy which often results from abnormal placental vascular anastomosis. It leads to twin reversal arterial perfusion with complex pathophysiology. Current information on early recognition and treatment for salvation of the normal twin is based on individual case reports in the literature. Mortality of Acardiac twin is 100%. Pump twin though structurally normal suffers due to heart failure and prematurity and has high morbidity and mortality all due to pumping blood to the acardiac twin. The acardiac twin receives all its blood supply from the pump twin through anastomotic channels, the term reversed perfusion is used to describe this condition because blood enters the acephalic twin through umbilical artery and exit through umbilical vein which is opposite to the normal blood supply. The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the pump twin. Here, we have presented a primigravida of 24-26 weeks of gestation with twin gestation of monochorionic diamniotic type in preterm labor. One of the twin was acardiac acephalous and another a pump twin with hydrops.

Key words

Acardiac twins, Twin pregnancy, Conservative management.

Introduction

The development of the acardiac anomaly is a rare complication of monozygotic multiple pregnancies. It was first described in the 16th century [1] and occurs in approximately 1% of monozygotic twin pregnancies and in 1 out of 35 000 deliveries [2]. The most widely accepted theory on the pathogenesis of acardius is the occurrence of a twin reversed arterial perfusion sequence (TRAP) [3]. TRAP sequence is an extremely rare anomaly with an overall incidence of 1 per 35,000 births, amounting to an average

risk of 1% among monozygotic twins [2]. TRAP sequence chiefly results from abnormal placental vascular anastomoses with consequent increase in arterial pressure in 1 twin leading to reversal of blood flow in the other.

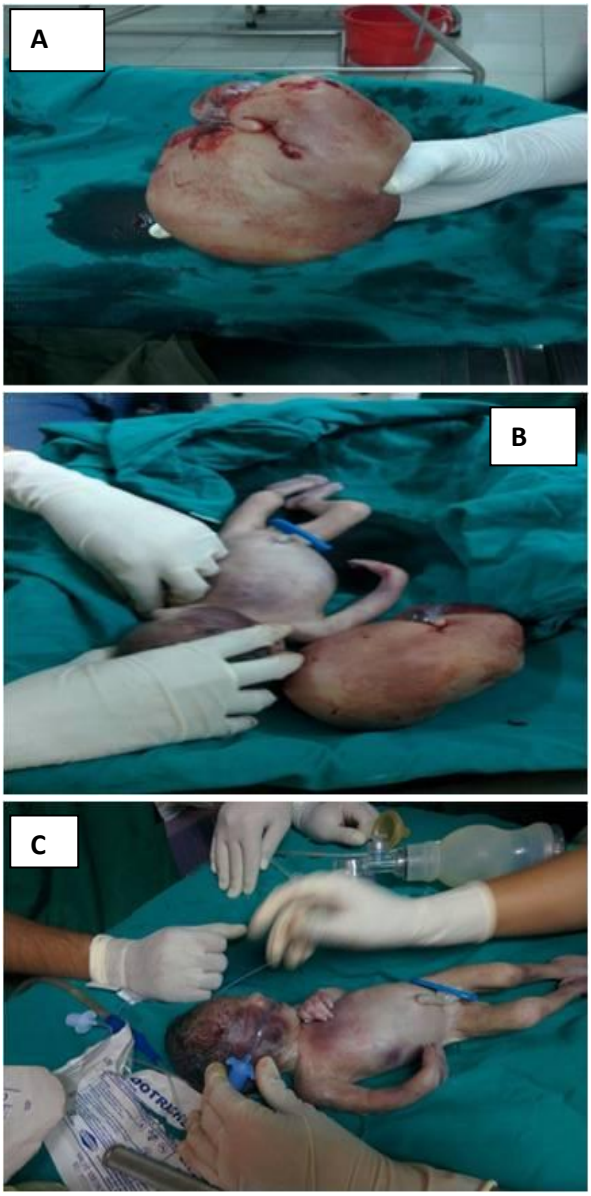
As a result, the "pump" twin perfuses deoxygenated blood into the "recipient" (acardiac) twin. A spectrum of anomalies due to reduced formation of body tissues, as a consequence of severe hypoxemia, results in acardiac-acephalus twins. We have reported here 2 cases of TRAP sequence, which created a considerable management dilemma with respect to salvation of the pump twin.

Case report

A 25-year-old gravida 4 was referred to us with suspected twin pregnancy at 24 weeks of gestation. Examination revealed a term-sized gravid uterus with multiple fetal parts and an increased amount of amniotic fluid. Fetal heart sounds could not be localized. Investigations included complete blood count, urinalysis, syphilis screening, and blood glucose measurement. Ultrasonography revealed a monozygotic twin pregnancy with 1 acardiac fetus and 1 normal fetus with polyhydramnios (amniotic fluid index -20).

Acardiac twin had poorly formed upper limbs with absent head while the other twin had no visible malformation or features of congestive cardiac failure. Estimated fetal weight was 500 g for the pump twin and 700 g for the recipient twin. The mother was symptomatic because of polyhydramnios. A provisional plan of intra fetal ablation with amnionic fluid drainage was made but premature rupture of membranes occurred that day, followed by delivery of 2 stillborn female fetuses weighing 500 and 800 g (twin A and twin B, respectively), with a normal single placenta. Twin B was acardiac-acephalus, the head and neck were absent, and the trunk and upper extremities were poorly developed. The other twin was normal and had no features of hydrops, but could not be salvaged due to the extremely low birthweight. There was no obvious placental anomaly (**Figure – 1A, 1B, 1C**).

Figure – 1: Acardiac Twins.



Discussion

Prevalence of multiple pregnancy is 1.5% of all pregnancies with a perinatal morbidity and mortality of 10% [2]. Acardiac twinning or TRAP sequence is a rare congenital anomaly of monozygotic multiple pregnancy due to abnormal placental anastomosis characterized by formation of a malformed fetus with an absent or rudimentary heart (acardius) along with other structures. Acardiac fetuses were first described

by Benedetti in 1533 [1]. Most common type of acardiac fetus is Acardiac acephalic variety. It has been hypothesized that the TRAP sequence is caused by a large artery to artery placental shunt often accompanied by a vein to vein shunt. Within the single shared placenta, arterial perfusion pressure of the donor twin exceeds that of the recipient twin who thus receives reverse blood flow of deoxygenated arterial blood from its co-twin. This used blood reaches the recipient twin through its umbilical arteries and preferentially goes to its iliac vessels. Thus only the lower body is perfused and disrupted growth and development of the upper body results [4]. An acardiac twin should be suspected in all monochorionic, malformed fetuses with cystic hygroma, generalized edema and an absent cardiac pulsation with a non-functioning heart. Ultrasonography finding of twins revealing discordant or grotesque malformation along with reverse flow in the umbilical artery is usually diagnostic of an acardiac twin [5]. Vaginal scanning and color doppler sonography is useful to diagnose it. Such twin pregnancies can be accessed by serial ultrasonography. Acardius acephalus means failure of head growth; acardius myelocephalus means partially developed head with identifiable limbs and acardius amorphous means failure of any recognizable structure [6]. Acardiac twins can also be classified as mentioned blow.

Acardius anceps – when head is poorly formed,
Acardius acephalus – if the head is absent
Acardius acormus – when only head is present.

Based on the development of heart, acardiac twins can be classified as
Hemiacardius - when heart is incompletely formed and
holoacardius if the heart is absent [7].

Total or partial absence of cranial vault, holoprosencephaly, absent facial structures, anophthalmia, microphthalmia, cleft lip, cleft palate, absent or rudimentary limbs, lungs, heart, liver and gallbladder, diaphragmatic defects, esophageal atresia, ventral wall defects, ascites, edema of skin and single umbilical artery are prominent features of recipient twin.

Management of twin pregnancy with an acardiac fetus is a challenge as the continuous growth of the acardiac fetus is deleterious to the healthy pump twin. It can lead to cardiac insufficiency, polyhydramnios, prematurity and even death of the structurally intact twin in up to 50% of cases [8]. Moore, et al. [9] reviewed 49 cases of acardiac twins and reported that perinatal outcome was related to the ratio of weight of the acardiac twin to the weight of the normal twin. They stated that when twin weight ratio which is the acardiac twin weight divided by the normal twin weight was above 70%, preterm labor, hydramnios and congestive heart failure in the pump twin were found and when the weight of the acardiac fetus was less than 25% compared to the pump twin, the prognosis was better. Brassard, et al. [10] reported that low pulsatility indices in the umbilical artery perfusing the acardiac twin compared with the pump twin correlated with poor prognosis [9]. Optimal management is controversial. Expectant management versus prenatal intervention is to be debated. Many methods of management have been proposed including termination of pregnancy, serial ultrasound scans to monitor for signs of decompensation, medical management of polyhydramnios or by serial amniocentesis, hysterotomy to remove anomalous twin and invasive procedures. Goal of prenatal treatment is to stop blood flow to the acardiac twin without affecting the pump twin in order to improve its outcome. Platt, et al. [11] in 1983 were the first to suggest occlusion of the circulation to the acardiac twin as the definitive treatment to interrupt blood supply to it [10]. Minimally, invasive intervention methods are through cord occlusion techniques or intrafetal ablation. Cord occlusion has been attempted by embolization, cord ligation, laser coagulation, bipolar diathermy and monopolar diathermy while intrafetal ablation is performed with alcohol, monopolar diathermy, interstitial laser and radiofrequency [11]. Tan and Sepulveda recommended intra fetal ablation as the treatment

of choice than cord occlusion [11]. They claimed ultrasound guided intra fetal approach to be easier, less invasive and with a higher rate of success than ultrasound and fetoscopy guided cord occlusion procedures. Invasive treatment should be restricted to those pregnancies which would benefit from prenatal intervention like those where the pump twin is at a significant risk of prematurity, cardiac insufficiency or death and should be considered in presence of poor prognostic factors like polyhydramnios, ultrasound markers of cardiac insufficiency, large acardiac twin and rapid growth of or evidence of substantial blood flow perfusion through the umbilical vessel supplying the parasitic mass [12]. Sepulveda W, et al. advocated expectant management in all cases [13]. They reported 90% survival in pump twin in 10 pregnancies with an acardiac twin managed expectantly. They cautioned against aggressive intervention and recommended expectant management with close fetal surveillance. Sepulveda W, et al. gave an opinion that conservative management is indicated in cases where the acardiac twin is small and when there are no signs of cardiovascular impairment in the pump twin. Serial ultrasound surveillance is important for detecting any worsening of the condition, which may suggest the need for interventions to optimize the pump-twin's chance for survival.

Conclusion

Expectant management with close antepartum surveillance deserves consideration in cases of monozygotic twins with Twin Reversed Arterial Perfusion sequence. Neonatal mortality of the pump twin diagnosed antenatally may be considerably less than reported. The interruption of vascular communication between the twins is difficult to accomplish. It needs expensive equipment and trained personnel for the procedures. Hence invasive treatment should be considered only in cases with poor prognostic factors. Such pregnancies should be managed by fetal medicine specialists familiar with invasive procedures. Intra fetal ablation procedures are better than cord occlusion techniques. In our cases, all the three pump twins survived with expectant management.

References

1. Benedetti A. De morborum a capitae ad pedis signis. Habes Lector Studiose Hoc Voluminae. Lucae Antonij Juntae, Venice, 1533.
2. James W.H. A note on the epidemiology of acardiac monsters. Teratology, 1977; 16: 211–216.
3. Haring D.A.J.P., Cornell M.C., Van der Linden J.C., et al. Acardius acephalus after induced ovulation: a case report. Teratology, 1993; 47: 257–262.
4. Blaicher W, Repa C, Schaller A. Acardiac twin pregnancy: associated with trisomy 2: case report. Hum Reprod., 2000; 15(2): 474–5.
5. Cunningham FG, Leveno KJ, Bloom SL, Hauth JC, Rouse DJ, Spong CY. Williams Obstetrics, 23rd edition, McGraw Hill, New York, 2010, p. 872.
6. Ishimatsu J, Nakanami H, Hamada T, Yakushiji M. Color and pulsed Doppler ultrasonography of reversed umbilical blood flow in an acardiac twin. Asia Oceania J Obstet Gynaecol., 1993; 19(3): 271–5.
7. Faye-Petersen OM, Heller DS, Joshi VV. Handbook of Placental Pathology, 2nd edition, London, Taylor and Francis, 2006.
8. Chandramouly M, Namitha. Case series: TRAP sequence. Indian J Radiol Imaging, 2009; 19(1): 81–3.
9. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. Am J Obstet Gynecol., 1990; 163(3): 907–12.
10. Brassard M, Fouron JC, Leduc L, Grignon A, Proulx F. Prognostic markers in twin pregnancies with an acardiac fetus. Obstet Gynecol., 1999; 94(3): 409–14.

11. Platt LD, DeVore GR, Bieniarz A, Benner P, Rao R. Antenatal diagnosis of acephalus acardia: a proposed management scheme. Am J Obstet Gynecol., 1983; 146(7): 857–9.
12. Tan TY, Sepulveda W. Acardiac twin: A systematic review of minimally invasive treatment modalities. Ultrasound Obstet Gynecol., 2003; 22(4): 409–19.
13. Sepulveda W, Sebire NJ. Acardiac twin: too many invasive treatment options – the problem and not the solution. Ultrasound Obstet Gynecol., 2004; 24(4): 387–9.