World Journal of Surgical Research

Case Report

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A Rare Case of Acardiac Twin in Postterm Pregnancy

[1]Rakesh Ranjan, [2]Jaya Kumari, [2]Sangeeta Pankaj, [2]Syed Nazneen , [2]Anita Kumari, [2]Anjili Kumari, [2]Simi Kumari

[1]Department of Pediatrics, Indira Gandhi Institute of Medical Sciences, Patna, India

[2]Department of Gynecological Oncology, Indira Gandhi Institute of Medical Sciences, Patna, India

Abstract

Introduction: Acardiac twinning is a rare congenital anomaly of monozygotic twin pregnancy. It results from abnormal placental vascular anastomoses. This leads to twin reversal arterial perfusion with complex pathophysiology. Here we present a case report of acardiac twin with late presentation. This case report shows the need for early ultrasonographic diagnosis of chorionicity and detailed ultrasonographic follow up of twin pregnancies and treatment for salvation of the normal twin.

Case Report: Here we present a case of acardiac twin with late presentation. This case report shows the need for correct and early ultrasonographic diagnosis of chorionicity and detailed ultrasonographic follow up of twin pregnancies.

Conclusions: Early and accurate antenatal diagnosis is essential to improve the prognosis.

Key Words –Acardiac twin, Twin Reversed Arterial Transfusion, Monozygotic twin, Ultrasound.

Introduction

Multiple pregnancy accounts for 1.5% of all pregnancies, with approximate perinatal morbidity and mortality of 10%. Multiple pregnancies are complicated by congenital malformations twice as often as with singletons. Certain malformations, such as conjoined twins and chorioangiopagus parasiticus due to twin reversed-arterial-perfusion (TRAP) sequence, are unique to monochorionic twin pregnancy. Incidence of acardiac twin is 1 per 35000 live births and occurs in approximately 1% of monochorionic twins [1, 2]. In this pregnancy, one twin has no cardiac structures or placental circulation and they receive the blood supply from the healthy twin by means of deep artery to artery (A - A) anastomosis or vein to vein (V-V) anastomosis. The blood of donor twin enters the acardiac twin via the umbilical artery and the iliac vessels. Thus, the arterial system of the acardiac twin is perfused in reverse order with deoxygenated blood from the normal twin (twin reversed arterial perfusion, or TRAP). As a consequence of severe hypoxia, a spectrum of anomalies occurs due to reduced formation of body tissue and dramatic alteration of fetal physiology, resulting in acardiac – acephalus twins. Complications in normal twin are congestive heart failure, preterm labor,

Address for correspondence and reprint requests to: Dr. Sangeeta Pankaj, Additional Professor, Gynecological Oncology, IGIMS, Patna drsangeetapankaj@gmail.com

Submitted Friday, August 18, 2017; Accepted: Saturday, October 21, 2017; Published Thursday, November 23, 2017

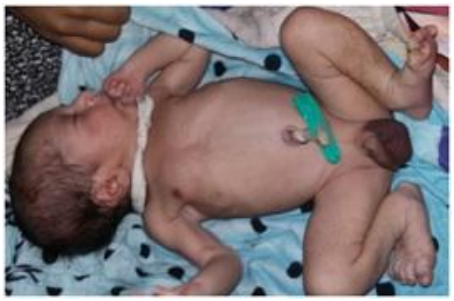

Figure 1 Normal Male Ist Twin

Fig2, Second baby acardiac twin

polyhydramnios and death in 50-70% of the cases. Acardiac twin has 100% mortality. We present a case of acardius acephalus twin.

Case Presentation

A 25 year old, belonging to low socioeconomic status, gravida 1 at 41 weeks of gestation came to our institute with complain of pain in abdomen and leaking per vagina for last 6 hours. She had not taken any antenatal check up earlier and had not undergone any ultrasonographic examination. On clinical examination uterus was term size with cephalic presentation, liquor seems to be adequate, head was 3/5 th palpable abdominally. Single Fetal Heart Sound (FHS) was heard in the middle of left spino-umblical line. She was admitted for institutional delivery. Subsequently after few hours she delivered 1st twin vaginally which was a normal male child of 2.5kg (Figure 1).

After 15 minutes 2nd twin delivered vaginally which was amorphous soft, globular mass. Mass had no any recognizable structure and it weighed 480 gram. Picture of cardiac twin shown in figure 2.

On gross examination by pediatrician first baby was male child of 2.5 kg without any obvious congenital anomalies. Apgar score was 8 after 1minute and 9 after 5 minutes, umbilical artery PH 7.23. Second baby which was acardiac twin had partially developed head with hair but no recognizable facial structure. Small cord present just below the malformed head. Head was attached with globular mass which resemble trunk (figure 3). Upper and lower extremity was not formed.

A single placenta was delivered with single amniotic membrane and two cords inserted side by side, one of them was very small and attenuated which belonged to the acardiac twin. Postnatal period was uneventful and the patient was discharged after 48 hrs of delivery.

Discussion

Prevalence of multiple pregnancies is 1.5% of all pregnancies with a perinatal morbidity and mortality of 10% [3]. Acardiac twining 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 Beneditti in 1533 [4].

It has been hypothesized that the TRAP sequence is caused by a deep artery to artery placental shunt often accompanied by a vein to vein shunt. Within single

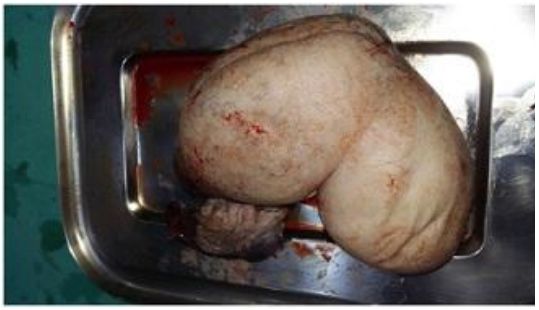

Fig-3, Second baby acardiac twin

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 results in disrupted growth and development of the upper body [5]. An acardiac twin should be suspected in all monochorionic twin with malformed fetuses with cystic hygroma , generalized edema and an absent cardiac pulsation with a non-functioning heart. Ultra sonographic finding of twin revealing discordant or grotesque malformation along with reverse flow in the umbilical artery is usually diagnostic of an acardiac twin [6]. This can be diagnosed in first trimester by vaginal scanning and colour Doppler sonography. Serial USG is indicated to assess such twin pregnancy.

Failure of head growth is called acardia acephalus, partially developed head with identifiable limbs is acardius myelocephalus , and failure of any recognizable structure to form is acardius amorphous [6].

Acardiac twin is classified according to the degree of cephalic and truncal maldevelopment [7]

The first type is acardius – acephalus. It is the most common variety. In this cephalic structure not formed.

The second is acardius – anceps. It is highly developed form. In this some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed.

The third is acardius –acromus. It is the rarest one. In this onlycephalic structure is present. The umbilical cord is attached to the head.

The fourth type is acardius amorphous. It is least developed and not recognizable as a human form with minimal development. It has no any distinguishable cephalic or truncal structure. This differs from teratoma only by its attachment to an umbilical cord.

Based on the development of heart, acardia twin can be classified as Hemiacardius – when heart is incompletely formed and holoacardius –if heart is absent [8].

The pathogenesis in TRAP sequence include [7, 9] deep placental anastomoses in early embryogenesis. It causes malformation of the acardiac twin. The early pressure flow in one twin exceeds that of other leading to the reversal of flow in the umbilical artery of the co – twin.

A primary defect in the embryogenesis in one twin which leads to failure of cardiac development in other. The normal twin then perfuses the acardiac twin via artery – artery anastomoses. The anastomoses are not responsible for the cardiac anomaly but are established as a result of defective embryogenesis.

The prominent feature of recipient twin are total or partial absence of cranial vault, holoporencephaly, absent or rudimentary

limb, lungs, liver, gallbladder, absent facial structure or rudimentary limb anophthalmia or microophthalmia, diaphragmatic defect, esophageal atresia, ventral wall defects , ascites , edema of skin cleft lip, cleft palate, and single umbilical artery. Management of twin pregnancy with acardiac fetus is essential in that twin in which continuous growth of acardiac fetus is deleterious to healthy pumped twin. It can lead to cardiac insufficiency, prematurity and even death of pumped twin in upto 50 % of cases [10].

Invasive treatment should be restricted to those pregnancies in which the pump twin is at a significant risk of prematurity, cardiac insufficiency or death .Treatment should be considered in presence of poor prognostic factors like polyhydramnios, ultrasound markers of cardiac insufficiency, large acardiac twin and rapid growth of acardiac twin or evidence of substantial blood flow transfusion through the umbilical vessel to the acardic twin [11]. Minimal invasive procedures like percutaneous insertion of helical metal coil to induce thrombogenesis in single umbilical artery of acardiac twin can be done. Blocking the vessels by coagulation using Nd:Yag laser and radiofrequency ablation under ultrasound guidance are now the first line of treatment [12].

Conclusion

Early and accurate antenatal diagnosis is essential to improve the prognosis of this rare entity of trap sequence. Early diagnosis may reduce the risk of complications in healthy twin by early intervention in complicated cases. Improved imaging techniques like 2D ultrasonography, 3D ultrasonography and transvaginal Doppler ulrasonography have made the diagnosis of acardiac possible in the first trimester of pregnancy by detecting inversion of vascular flow in the recipient acardiac fetus. There could be 95% survival in the pump twin with an average age at delivery between 36 and 37 weeks. No significant neurological abnormalities have been identified in these children so far.

Authors' contribution

RR - carried out the experiments and interpreted the results,

JK - carried out the literature search and prepared the draft manuscript,

SP - designed the study and performed the analysis,

SN - conceived the study, participated in design and edited the final manuscript.

AK - conceived the study, participated in design and edited the final manuscript.

AK - conceived the study, participated in design and edited the final manuscript.

SK - conceived the study, participated in design and edited the final manuscript.

All authors read and approved the final manuscript.

Conflict of Interests

The authors declare that there are no conflicts of interests.

Ethical consideration

Written informed consent was obtained from the patient and a copy of consent is available with the authors

Funding

None declared

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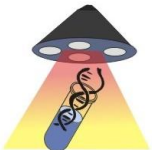

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