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Case Report

A rare case report of twin reversed arterial perfusion sequence in monochorionic diamniotic twin and outcome of Dr. Pump and Mr. Acardiac acephalus in a tertiary care centre

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ABSTRACT

Twin reversed arterial perfusion (TRAP) sequence or Acardiac twinning is a rare complication seen in monozygotic gestation. The reported incidence of Acardiac twin is 1 in 35000 births and 1 in 100 monozygotic twin gestation. In monochorionic twin pregnancies, a twin with poorly developed heart and upper torso (the "Acardiac") is perfused by its co-twin (the "pump") via superficial arterio-arterial or veno-venous anastomoses located on the placental surface, through which blood pumped backwards. Due to poor prognosis of TRAP sequence in monochorionic pregnancies, 100% mortality was reported in Acardiac twin due to its severe malformations and also due to heart failure and prematurity in pump twin 50-55% mortality was reported. We hereby report a case of 22-year-old primigravida, monochorionic diamniotic (MCDA) twins presented with abruption at 24 weeks of gestation, resulted fetal demise of pump twin. Our study also reviewed necessity of thorough sonological evaluation and therapeutic opportunities in monochorionic multifetal pregnancies.

Keywords: Monochorionic twin gestation, TRAP sequence, Acardiac acephalus twin, Pump twin

INTRODUCTION

Acardiac twin also referred to as twin reversed arterial perfusion (TRAP) sequence in monochorionic twin pregnancy is an extremely rare complication with reported incidence of 1 in 35000 births and 1 in 100 monozygotic twin gestation.[1-3] TRAP sequence is the most severe form of twin-to-twin transfusion syndrome (TTTS) in monochorionic pregnancies, which is also known as chorioangiopagus parasiticus.

As per Quaas et al study report, the incidence of TRAP sequence could be higher due to early detection of first trimester scan and more use of assisted reproductive techniques in infertility management.[4] TRAP sequence in monochorionic gestation is commonly misinterpreted as fetal demise because of absent cardiac activity.

The pathophysiology of TRAP sequence in monochorionic gestation is abnormal placental vasculature leading to reversed arterial perfusion of Acardiac twin by pump twin through the umbilical arteries which is in opposite direction to what usually happens. Because of this reversed circulation, the normal pump twin not only support its circulation also it pumps thin oxygen-poor blood to Acardiac twin leading to development of lower extremity only. This may lead to cardiomegaly, high output congestive heart failure (CHF) in the pump twin eventually leads to polyhydramnios, hydrops fetalis and prematurity.

Acardiac anomalies are divided into four types based on placental blood flow disruption includes Acardiac acephalus, Acardiac amorphous, Acardiac acormus and Acardiac anceps. The fifth category of acardia called Acardia myelacephalus, was suggested by Simonds and Gowen.[5] Acardiac acephalus is the most common subtype,

which had well developed pelvis and lower limbs and poorly developed upper extremities and missing head and thoracic organs.

Our study was also aimed to provide an overview of literature data of Acardiac twin in monochorionic gestation about the diagnosis, prognosis and management which are still unclear and debated due to poor quality evidence.

CASE REPORT

A case of 22-year-old primigravida, who conceived spontaneously after 8 months of marriage, booked and Immunized in Government Villupuram Medical College, Tamil Nadu, India. During her routine Antenatal visit, she was advised ultrasonography at 13 weeks of gestation, revealed monochorionic diamniotic twin pregnancy with fetal demise of second twin. Counselling given to the parents regarding the prognosis of surviving twin and conservative, invasive treatment approach for dead fetus. Also, we counselled for invasive procedure for dead fetus in first trimester but patient opted to continue pregnancy without invasive intervention due to its high cost. After obtaining their consent, conservative treatment approach with regular follow-up and serial ultrasonographic (USG) examination was advised. The growth of the surviving fetus was satisfactory; she was periodically monitored and coagulation profile was checked once in a week.

During her routine antenatal visit, at 23 weeks of gestation, detailed USG was advised due to discrepancy between gestational age according to fundal height. Detailed USG revealed, the normal fetus (pumping twin) was observed with polyhydramnios (single deepest pocket >12 cm), normal fetal anatomy and normal fetal heart and EFW 547 g, with single large placenta. The dead fetus (perfused twin) showed features of amorphous mass with rudimentary lower limbs, spine, omphalocele with extensive generalised subcutaneous oedema supplied by a short cord receiving the blood from pump twin with reversed perfusion measuring volume 271.61 cc. The Umbilical artery Doppler of PUMP twin was normal but MCA PSV falls 1.30 MoM for gestation but not indicative of fetal anaemia and Ductus venous Doppler also normal.

According to the International recommendations and protocols, Counselling was given to their parents which includes risk of chromosomal abnormalities (9%), risk of involution of Acardiac twin and haemodynamic complications which could lead to pregnancy involution. Also, risk of prematurity, polyhydramnios and haemodynamic changes were also explained. Possibility of interventions and its risks were explained. Active interventions like cord ablation by different energy sources like bipolar cord coagulation, intrafetal laser ablation, intrafetal radio-frequency ablation, microwave ablation of the umbilical cord has a technical and clinical success rate of 70%, 10% revascularization rate, procedure related miscarriage rate of 10%, decreases the co-twin residual neurological damage rate to 5%, preterm birth rate of 20%. In view of huge size of perfused fetus, the risk of demise or damage to pump twin are close to 90% if left untreated. Additionally, the perfused twin, is likely to grow into an amorphous monster, that would pose difficulty in delivery irrespective of the mode of delivery. Therefore, prenatal intervention is medically indicated to improve the maternal and perinatal outcome. Based on the size of the umbilical cord vessels, the anatomy at the hilus, extensive hydropic changes at the TRAP fetus, the option of Microwave cord coagulation was deemed appropriate for our case and hence we advised for the same. At this stage, patient was opted for invasive procedure and we admitted her for the same.

During her admission, she suddenly experienced severe abdominal pain and blood-stained liquor drained per vaginum at 24 weeks of gestation. On physical examination, she was afebrile, pallor, pulse rate-124/min, blood pressure-140/90 mmHg, and SpO_2-99% in RA. Per abdomen examination revealed uterus corresponds to 36 weeks' size, acting, tense and tender, FHS absent for both twins. Per vaginal examination showed cervical effacement of 25%, dilated os, membranes were absent, presenting part at brim, 250 grams of blood clots were removed. Grade 3 abruption was diagnosed and prognosis was explained to family. Labour was induced with intravenous oxytocin drip and she had vaginal delivery. During delivery 2 units of packed RBCs (blood) transfused. The first twin was female weighing 685 grams with distended abdomen and second twin weighed 460 grams, head and upper extremities were absent, a soft globular mass with poorly developed female external genitalia, the right foot had 3 toes and the left foot had 4 toes, lower leg bones and lower spinal column were apparently normal. Foetus was classified as Acardiac acephalus and autopsy revealed absent fetal heart. Placenta was monochorionic diamniotic with small thin cord attached to Acardiac twin and weighed 405 grams. Postpartum period was uneventful and discharged on 5th postnatal day.

Figure 1: Monochorionic diamniotic twin pregnancy-TRAP sequence- Dr. Pump and Mr. Acardiac acephalus- abruptio placentae (at 24 weeks of gestation) - 250 grams of retro placental clots.

DISCUSSION

Twin reversed arterial perfusion or chorioangiopagus parasiticus is scarce condition of monochorionic twin pregnancy with incidence of 1% monozygotic twins.[6] It is due to abnormally reversed umbilical arterial flow from the pump twin to the donor twin by placental arterio-arterial anastomosis. It has been divided into 5 types: Acardiac anceps-poorly formed head, Acardiac acephalus-absent head, Acardiac acormus-head only present, Acardiac amorphous-unrecognizable anatomy, and Acardiac myelacephalus refers to a partially developed head with identifiable upper limbs plus and/or minus some nervous tissue. It can also be classified as hemiacardius-incompletely formed head, and holoacardius-absent heart.[7] These classifications have no prognostic value and do not provide any information regarding management and used only for precise morphological description.

The main pathogenesis behind TRAP is due to flow of low oxygen blood from the pump twin passing through the placental arterioarterial anastomosis which ultimately end up in systemic blood flow of the recipient twin (Acardiac/perfused twin) resulting in TRAP sequence. Even though the pump twin has the usual circulatory pattern, the portion of its cardiac output has been filched by the recipient twin via vascular anastomosis. By this way the pumping twin has to maintain blood supply of its own and also for the Acardiac twin. This abnormal blood flow of fetus allows the low oxygen blood flow from the pumping twin to the Acardiac twin at the level of iliac arteries promoting raised blood flow to the lower part of the body with poor perfusion of upper body leading to the formation of inferior extremities with malformed or absent fetal heart.[8]

The pumping twin undergoes high cardiac output stress leading to polyhydraminos, cardiac failure, hydrops fetalis, preterm, fetal growth restriction. The recipient twin has various anomalies like absent cranial vault partially, malformed or absent facial growth, rudimentary heart or absent heart, rudimentary limbs development, poor thoracic development, abdominal wall defects, diaphragmatic defects, ascites.[9,10] Another proposed theory suggested that defective cardiac embryogenesis where due to absence of heart development, along with chromosomal abnormality or environmental factors the unique perfusion of anastomosis by placental vessels to Acardiac twin was developed. The cytogenetic evaluation of Acardiac twin in literature was finite, but difference in karyotyping noted in parasitic twins.[11] Hence altered embryogenesis leads to impaired development of Acardiac fetus with vascular anastomosis of placenta accompanied by pump twin.[12]

The management includes serial monitoring of pumping twin to look for early signs of hydrops development and any reversal of blood flow since salvage of pumping twin remains as primary goal. Lehr and DiRe made the first diagnosis of Acardiac twins.[13] Most of the time the diagnosis made in second or third trimester by trans abdominal ultrasound scan. Sometimes the diagnosis made during labour, after or prior to delivery of pumping twin by expulsion of amorphous ambiguous mass.[14]

In ultrasound, the pathognomic features like gross difference in fetal biometry particularly abdominal circumference and the Acardiac twin appears to have various malformations, absence of heart, soft tissue oedema. The pump twin appears to have fetal growth restrictions, Doppler changes, signs of heart failure like cardiomegaly, pleural and pericardial effusion, polyhydraminos, and signs of hydrops fetalis.[15] First trimester might be difficult but it can be made by transvaginal scan. 3D ultrasound also gives detailed anatomy of both twins.[16] Guimaraes advocates usage of magnetic resonance imaging in diagnosis of TRAP.[17]

Ishimatsu et al noted that umbilical artery reverse flow in doppler can be used in Acardiac twin.[18] The abnormal peak systolic to end diastolic velocity ratio from Doppler velocimetry of umbilical cord noted in recipient twin as per Shbire et al study.[19] As per Smith et al, Acardiac twin has poor trunk and upper limb formation with severe congenital anomalies and anencephaly.[20] Pinet et al reported that Acardiac twin has various congenital anomalies like cardiogenic abnormalities, abdominal wall defects, skeletal abnormalities.[21]

The goal of treatment is to deliver pump twin at near term without any hydrops or heart failure and early diagnosis to explain TRAP sequence to parents and opt for termination of pregnancy or to continue the pregnancy with serial ultrasound and Doppler follow up. The perinatal mortality of pump twin was found to be 55%.[22]

Treatment options remain debatable to choose between expectant management or prenatal intervention. Various options like termination of pregnancy, serial ultrasound monitoring of pump twin for fetal growth restriction, signs of heart failure, treating polyhydraminos by medically or amniocentesis, removal of Acardiac twin by hysterotomy and invasive procedures. According to Sullivan et al, expectant management in 10 cases of TRAP results in 90% survival rate of pump twin hence warning against aggressive intervention and promoting close surveillance.[23] Stamatian et al described that conservative management was advisable in case of small Acardiac twin and pump twin without any signs of heart failure.[24] Jelin et al and Mann et al also prefer expectant management.[25,26] Platt et al suggested occlusion of blood flow to Acardiac twin is the definitive management.[27] Various minimally invasive techniques like cord occlusion via embolization, ligation, laser ablation, usage of bipolar cautery can be used along with fetoscopy or ultrasound guidance. Intrafetal ablation by interstitial laser, radiofrequency on pelvic vessels or abdominal aorta can also be tried using ultrasound doppler.[28] Lee et al described that 80% pump twin survived after radiofrequency ablation of Acardiac twin.[29] According to Tan and Supelveda, intrafetal ablation

has low technical failure rate and low preterm delivery with high clinical success rate.[30] Cabassa et al also reported 85% of neonatal survival rate after radiofrequency ablation treatment in TRAP.[31] Chaveeva et al, meta-analysis along with case series update revealed 76% nenonatal survival rate in intrafetal laser therapy.[32]

Won et al described treatment options based on classification where abdominal circumference used to evaluate Acardiac twin size on antenatal ultrasound and signs of cardiac failure in pump twin on antenatal ultrasound. According to this classification, type 1- small or medium size Acardiac fetus with abdominal circumference ratio less than 50%, type 2- large Acardiac twin with abdominal circumference more than 50%. Here "subtype a" includes pump twin without cardiac failure and "subtype b" includes pump twin with cardiac failure. Type 1a can be managed by conservative expectant management, type 1b requires serial ultrasound follow up look for spontaneous resolution of anastomosis or invasive treatment on worsening state. Type 2a needs prenatal treatment due to large Acardiac twin to prevent preterm birth. Type 2b needs urge treatment.[33]

The time of intervention in TRAP also remains controversial, whether to wait for pump twin to develop ultrasound features of cardiac failure and to intervene at 16-18 weeks of gestational age to avoid early intervention causing abortion. Otherwise, to intervene at 12 weeks to avoid pump twin death between 12-16 weeks. As per Pagani et al, intervention before 16 weeks had low adverse pregnancy outcome.[34] Roethlisberger et al revealed that fetal loss rate is significant in cases treated before 14 weeks of gestational age.[35]

Moore et al studied 49 cases of Acardiac twin and reported that when the ratio of weight of Acardiac twin to the weight of pump twin was more than 0.7 (70:100), the incidence of preterm labour, polyhydraminos, cardiac failure of pump twin noted to be in higher percentiles as 90%, 40%, and 30%.[36] When this ratio falls below 25% prognosis of pump twin was found to be better. Here the weight was derived after delivery. In our study this prognosis ratio value was 0.6 explaining the favourable outcome if intervention was done. Second order regression equation was used to predict antenatal weight of Acardiac twin as the following.

$$Weight\ in\ grams = 1.2 \times (Longest\ dimension\ of\ acardiac\ twin\ in\ cm^2) - 1.7 \times (Longest\ dimension\ of\ acardiac\ twin)$$

The prognosis during early weeks of gestation can also be evaluated using ultrasound indicators such as pump twin Crown rump length (CRL) and Acardiac twin upper pole rump length (URL) by CRL-URL/CRL and URL/CRL.[35] According to Tang et al, 21 cases of TRAP evaluated and early pregnancy prognosis identified using this ratio.[37] In this study CRLURL/CRL ratio >0.43 and URL/CRL ratio <0.57 used to identify pump twin survivors by using multiple regression analysis.

CONCLUSION

TRAP should be suspected in all cases of monochorionic twins with one twin presenting with absent cardiac structure or as fetal demise. Early diagnosis and serial monitoring for fetal growth, fetal heart failure is implicated. For optimal management and accurate diagnosis of Acardiac twin, a high index of suspicion is required. Prompt early diagnosis, Mode of treatment by intrafetal laser ablation and time of intervention before 16 weeks might improve pump twin outcome of TRAP. Even though we had a favourable outcome according to the prognosis criteria, in our case both twins expired due to unexpected placental abruption.

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