Twin reversed arterial perfusion sequence in monochorionic diamniotic pregnancy: A rare occurrence

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ABSTRACT

Twin reversed arterial perfusion is a rare complication occurring in monochorionic multifetal pregnancies. This condition is characterized by a malformed fetus being perfused by a normal twin through an artery-to-artery anastomosis in the reverse direction. Herein, we report a case of a multigravida female with a twin pregnancy at 27 weeks 5 days of gestation referred in view of twin pregnancy with the intrauterine demise of one twin. Gray scale and color Doppler imaging revealed a monochorionic diamniotic pregnancy with a viable, normal twin, and an amorphously developed acardiac twin. The patient was monitored with weekly ultrasonography, echocardiography, and Doppler ultrasound examination to ascertain the well-being of the pump twin. She delivered a normal live baby at term and an acardius acephalus fetus. The perinatal mortality of the pump twin is significantly high. Therefore, it is necessary to diagnose this entity at an early gestational age through improved imaging techniques, so that timely intervention can be planned.

Key words: Acardiac twin, Color Doppler, Monochorionic diamniotic pregnancy, Twin reversed arterial perfusion

We report the case of a multigravida female with monochorionic diamniotic (MCDA) pregnancy who has been referred to our institute at 27 weeks 5 days of gestation in view of twin pregnancy with the intrauterine fetal demise of one twin which was later diagnosed as TRAP sequence at our institute.

CASE REPORT

A 22-year-old G2 P1 L1 A0 female was referred to our department in view of twin pregnancy with intrauterine fetal demise of one twin.

The patient had previous scans at 9-week POG which showed the absence of cardiac activity in one twin at that time. Gray scale imaging performed at our institute on the Philips EPIQ-5 machine revealed a MCDA twin pregnancy with a viable fetus showing normal cardiac activity and another amorphously developed acardiac twin. The patient was noted to have a normal live baby at term with a normal head circumference. The perinatal mortality of the pump twin is significantly high. Therefore, it is necessary to diagnose this entity at an early gestational age through improved imaging techniques, so that timely intervention can be planned.

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corresponded to 24 weeks 2 days. The recipient twin showed dysmorphic growth and defective organogenesis with multiple anomalies. Skull vault was absent with severely dysmorphic brain parenchyma and unidentifiable facial features (Fig. 1b). The fetal thorax was underdeveloped with hypoplastic ribs and without any recognizable cardiac tissue indicating acardia (Fig. 2a). Stomach bubble, gallbladder, kidneys, and urinary bladder were not identified and the spine was severely deformed (Fig. 2b). Upper limbs were absent. However, the lower extremities were well developed. There was evidence of generalized subcutaneous edema in the recipient twin (Fig. 3) with multiple well-defined cystic lesions in the abdominal wall in subcutaneous tissue.

On further evaluation with color Doppler imaging, the first twin showed normal fetal Doppler parameters for the middle cerebral artery and umbilical arteries (Fig. 4). However, there was a single umbilical artery in the acardiac twin which showed the reversal of flow to the acardiac fetus (Fig. 5). This finding was inconsistent with the normal fetal physiology. Hence, the diagnosis of TRAP sequence was made and the mother was informed that the second twin was non-viable and acardiac which was supplied by its co-twin. She was also informed and counseled about the prognosis of the acardiac twin, risks to the pump twin, and in addition, the risk of preterm labor.

Fetal echocardiography was then performed for the pump twin at our department which was normal. Four chamber views, outflow tracts, three-vessel views, three-vessel trachea views, and bi-caval views were normal. There was no evidence of tricuspid or mitral regurgitation or pericardial effusion. The patient was then followed up weekly with routine fetal scans, color Doppler imaging, and interval fetal echocardiography to assess for any complications in the pump twin. Planned cesarean delivery was attempted at term and she delivered a normal healthy twin and other dysmorphic non-viable twin. The acardiac twin showed abnormal morphology with a poorly developed head, absent facial structures, and upper extremities (Fig. 6). The mother, however, refused the autopsy of the acardiac twin.

**DISCUSSION**

The TRAP sequence or chorioangiopagus parasiticus is a rare and unique anomaly that is seen in monochorionic multifetal pregnancies with a reported incidence of 1 in 100 monozygotic twin pregnancies and 1 in 35,000 overall births [3,10,11]. One of the important and popular theories regarding the pathophysiology is that there is abnormal vascular anastomosis within the placenta,
i.e., aberrant arterio-arterial anastomosis which leads to the perfusion of the deoxygenated blood from the pump twin in a retrograde fashion into the acardiac twin [1,2,10]. Therefore, the blood supply to the acardiac twin helps in maintaining the growth of the lower limbs and nearby structures supplied by the iliac vessels and distal abdominal aorta but flow to upper limbs and other distant structures may be inappropriate to sustain growth [7,11]. Hence, acardiac twins may develop abnormalities such as encephaly, microcephaly, absent/hypoplastic upper half of the body and upper limbs, dysmorphic heart, and cystic hygroma [11]. However, the other proposed hypothesis was that cardiac dysmorphogenesis during early embryogenesis leads to the development of acardiac twins in multifetal pregnancies [2,10].

In literature, four morphologic types of acardiac twins have been described: acardius acephalus, acardius ances, acardius amorphous, and acardius acorum. In acardius acephalus (60–75% of cases), the head and thorax are not well developed but the lower limbs are developed. In acardius amorphous (20% of cases), there is an amorphous mass of tissue with no recognizable structures. In acardius ances (10% of cases), there is a partly developed head, thorax, and abdomen. Only cephalic structures are developed in acardius acorum (5% of cases) [4,7,9,12]. Acardius myelacephalus, another category of acardia was introduced in 1925 by Simonds and Gowen. In this category, the acardiac fetus shows only a partially developed head with recognizable upper limbs with or without some nervous tissue [9]. The mortality rate of the acardiac twin is 100% [13,14]. The overall mortality rate of a normal fetus varies from 33% to 55% [7,13,14].

Based on the routine ultrasound (USG) scans, fetal Doppler, and fetal assessment scans, our fetus belongs to the acardius acephalus category because the lower limbs were developed in our case with the absence of upper limbs and partially developed head and thorax. We should always consider this entity in our differential diagnosis if we see an abnormally developing fetus with a normal fetus in a monochorionic twin pregnancy. Other differential diagnoses for this entity are intrauterine fetal demise, twin-twin transfusion syndrome, vanishing twin, and fetus papyraceous [9]. However, the constant increase in the growth of the “presumed dead” fetus on follow-up scans should make the examiner reconsider the diagnosis of acardiac twins [5]. As in our case, the diagnosis of TRAP sequence was missed on the first-trimester scan. However, on further scan at our institute, the discordant growth, abnormal morphology, and reversal of flow in the umbilical artery of the dysmorphic fetus on color Doppler examination led us toward the final diagnosis of TRAP sequence. It is crucial to know the entity’s pathophysiology and related imaging features to reach the appropriate diagnosis of the TRAP sequence at an early gestational age. The correct documentation of reversal of flow in the umbilical arteries on color Doppler examination is a pathognomonic feature of this rare entity [7,9].

Khanduri et al. also reported a similar case of acardiac twinning in which they monitored the patient with weekly ultrasonography, echocardiography, and color Doppler examination to observe the well-being of the pump twin [1]. Dhanji and Bredadam also reported a case of TRAP sequence in which the diagnosis was missed in the first-trimester US. After the diagnosis was made, radiofrequency ablation (RFA) was performed for the survival of the pump twin, but the prognosis was abysmal [9]. Vitucci et al. reported that the typical features of USG are discrepancies in biometrical measurements of twins, especially the abdominal circumference, non-visualization of a structurally normal heart in one twin with other malformations of head, trunk, upper and lower limb, and the presence of subcutaneous edema [15]. In a similar case reported by Behura et al., the patient was diagnosed as a case of MCDA twin pregnancy with a normal live fetus and another as a vanishing twin which was deformed and showed absent cardiac activity. On further follow-up, the vanishing twin showed serial interval growth and color Doppler revealed blood flow toward the acardiac fetus; hence, the diagnosis of TRAP sequence was made [16]. Nanthakomon et al. reported a similar case where a twin pregnancy with a single intrauterine fetal demise was suspected. USG showed that the surviving twin was structural whereas the other twin was grossly malformed at the cephalic part with a large septate cystic hygroma at the thoracic area and well-developed lower extremities with club feet. Cardiac structure could not be identified in this fetus. Doppler study revealed that the direction of blood flow in the umbilical arteries of the acardiac fetus was in the opposite direction whereas the pump twin had a normal flow pattern [17].

Magnetic resonance imaging can be a useful adjunct in some cases to reach a confident diagnosis. Several studies emphasized the early diagnosis of this entity for effective management and to reduce the risk of complications in pump twins, such as high-output cardiac failure, preterm delivery, and intrauterine fetal demise [3,5,7,9]. Signs of high-output cardiac failure include cardiomegaly, pleural effusion, ascites, generalized edema, tricuspid regurgitation, and polyhydramnios. Therefore, fetal echocardiography is of utmost importance for the evaluation and surveillance of cardiac function in normal twins [4]. The ultimate goal of management in the TRAP sequence is toward the survival of the pump twin and to be able to deliver the normal pump twin at term. Many studies recommend that conservative management.
with close follow-up/monitoring is the right option when the weight of the acardiac twin is <50% of the weight of the pump twin [18]. Evidence of high-output cardiac failure in pump twins is a poor prognostic indicator and these can be further managed with interventions such as target occlusion of the umbilical cord of an acardiac fetus using laser ablation, bipolar cord coagulation, diathermy, cord embolization, or RFA [1].

CONCLUSION

TRAP sequence is a rare obstetrical complication of twin pregnancy which includes a normally developed viable pump fetus and an abnormal non-viable acardiac fetus. The pump fetus has a high risk of high-output cardiac failure and congenital anomalies with an increased mortality rate. Serial USG scans, fetal echocardiography, and Doppler assessment are useful in following up on patients affected by the TRAP sequence to assess the circulatory well-being of the pump fetus. Therefore, it is crucial to diagnose this condition with due diligence for the timely management of such patients, as most often the normal twin can be saved.

AUTHORS’ CONTRIBUTIONS

1. Dr. Aarti – She was responsible for doing the ultrasound of the patient firsthand and came up with the correct diagnosis. She also advised and closely followed up the patient by serial gray-scale ultrasound and color Doppler. She wrote the manuscript with keen interest and performed an extensive review of the literature.

2. Dr. Aayush Bansal – He was responsible for the finalization of the manuscript.

3. Dr. Rajesh K. Badhan – He supervised the ultrasound of the patient and helped come up with the correct diagnosis.

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Batoy A, Bansal A, Badhan RK. Twin reversed arterial perfusion sequence in monochorionic diamniotic pregnancy: A rare occurrence. Indian J Case Reports. 2024; April 19 [Epub ahead of print].