A case report of twin reversed arterial perfusion sequence with expectant management

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Keywords: Monochorionic twin pregnancy, acardiac twin, TRAP sequence

Abstract

Acardiac twin or TRAP (twin reversed arterial perfusion) sequence is a rare complication of monochorionic pregnancies. In these cases, the heart is either absent or non-functional. It's controversial whether conservative management or therapeutic treatment is better in TRAP-sequence.

In this case, we present a 19-years old, primigravida diagnosed with spontaneous monochorionic monoamniotic twin pregnancy at 7th week of gestation. One of the fetuses had a crown-rump length of 8 mm and fetal heart rate 122/minute while the other one had a crown-rump of 7 mm with no detected fetal heartbeat. At the 11th week of gestation, the acardiac twin continued growing despite the absence of the fetal heart beat and fetal extremities. The cranium could not be evaluated clearly. The diagnosis of TRAP sequence was confirmed by the reversed direction of flow observed in the umbilical artery. Since the patient and her husband did not want any intervention, no interventional diagnostic and treatment modalities were applied. Preterm labor started at the 32th week. Because the presentations were transverse and breech, pump and acardiac fetus, respectively, a cesarean delivery was performed. A healthy female baby, weighing 1650 gr with APGAR scores of 9-10, first and fifth minutes, respectively, was delivered along with the acardiac fetus which was 1550 gram in weight, fetal heart beat negative, with upper and lower extremity deformities. The uniqueness of the present case is that there was no significant difference in the weight of both twins. The acardiac twin was as large as the pump twin. Except for twenty days hospitalization because of neonatal respiratory distress syndrome, which was a consequence of preterm labor, there were no problems with the pump twin even though managed conservatively.

In monochorionic twin pregnancies, when one of the twins is found to be fetal heartbeat negative and it continues to grow with concomitant structural abnormalities, the TRAP sequence should come to mind. If the acardiac fetus is small and the pump fetus has no complications, the conservative approach can be considered. Our goal should be to deliver the pump twin with minimal complications. To achieve this, follow-ups and treatment should be individualized for each patient, the pump fetus should be monitored closely.

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Introduction

Acardiac twin or TRAP (twin reversed arterial perfusion) sequence is a rare complication of monochorionic pregnancies. In these cases, the heart is either absent or non-functional. It is seen in one percent of monozygotic twins, the incidence in all births is approximately 1/35,000.[1] The absence of one fetal heartbeat in twin pregnancies may be diagnosed in the first trimester. Rarely the acardiac twin is detected to continue growing despite the absence of a fetal heartbeat and concomitant fetal anomalies. The deformities can be in a wide range from mild to severe. Acardius acephalus, is the most common form which has no cranial structures and is composed of only lower limbs and a mass-like body. Acardius anceps is formed partly by the head and face, acardius amorphous is only a mass, and the rarest form acardius acormus is composed of the only head can be detected also.[2]

The nonviable twin is perfused with retrograde blood flow from the other twin that flows through arterioarterial anastomoses; the blood returns to the normal twin by venovenous anastomoses that bypass the placenta. Because these vessels carry less oxygenated blood and the flow is asymmetrical, the result is malformations mostly seen in the cranium and upper extremities.[3] In our case we present an acardius anceps that reached the same size as the pump fetus and was followed conservatively.

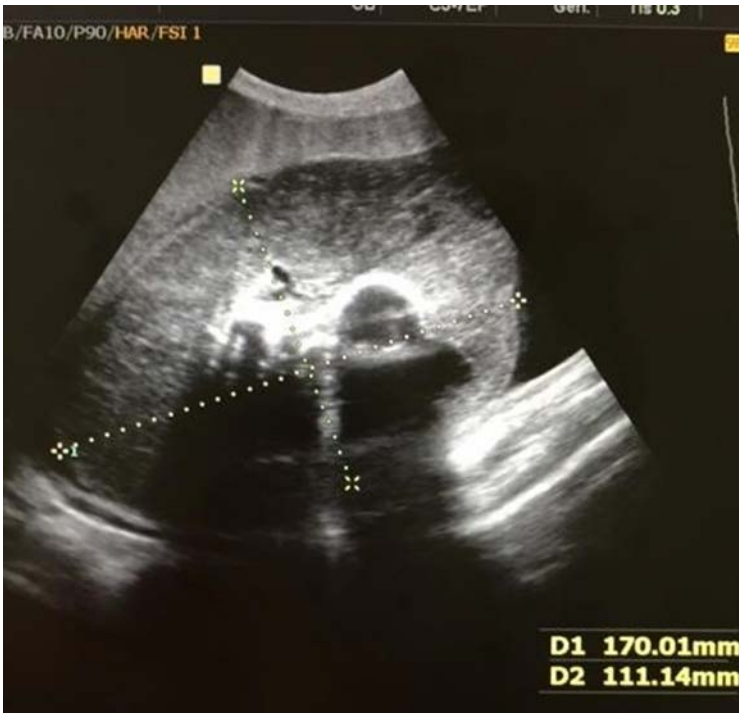

Figure 1. 28th week ultrasonographic view of TRAP sequence

Case

We present a 19-year-old, primigravida with a spontaneous twin pregnancy, with no family history of twin pregnancies and no systemic disease. The first examination by us was performed on the seventh gestational week according to the last menstrual period, a monochorionic monoamniotic twin pregnancy was diagnosed. One of the fetuses was found to be fetal heartbeat negative. At her second ultrasound screening at 11th weeks of gestation the 1st fetus crown-rump length (CRL) was 43 mm, nuchal translucency (NT) was 1.2 mm, nasal bone was present and fetal heartbeat was positive without any congenital malformations. At the same examination, the second fetus CRL was 39 mm, fetal heartbeat was negative, cranial structures and upper limbs could not be evaluated clearly. The next examination which was carried out two weeks later at 13th week revealed that the acardiac twin had continued growing and TRAP sequence diagnosis was confirmed when arteria-arterial anastomoses was observed by doppler ultrasonography. The patient was informed in detail about complications and interventional treatment, the patient and her husband refused all interventions and stated their rejection verbally and written. In the strict follow-up of the patient, a detailed sonography was perfomred at the 20th week. The acardiac fetus continued to grow at the same rate as the pump fetus, lower extremities were observed, upper structures and head were not evaluated clearly. (Figure 1) Anti-D immunoglobulin for RH incompatibility at 28th gestational week and betamethamethasone for fetal lung development before cesarean delivery were administered. Preterm labor started at the 32th week. Because the presentations were transverse and breech, pump and acardiac fetus, respectively, a cesarean delivery was performed. The first delivered was a live female via finding and pulling a foot, with Apgar scores of 9-10 at one and five minutes. Birth weight was 1650 gr. The acardiac twin's birth weigth was 1550 gr and 30 cm. A thin hairy scalp was seen on the structure which was thought to be the head. Depressions for the eyes on the front face of the head were monitored. The acardiac twin appeared as a trunk like structure. The lower limbs were clearly visible up to the three centimeters below the knee. Gender was not clearly seen. The stumps that were thought to be the upper extremities were stuck to the lower part of the head. (Figure 2) The placenta was unique and relatively large, was completely removed and there were no complications at birth. The female twin was discharged after 20 days in the intensive care unit because of neonatal reespiratory distress syndrome. There was no developmental or neurologic pathology observed at her 6th month control examination by pediatrists.

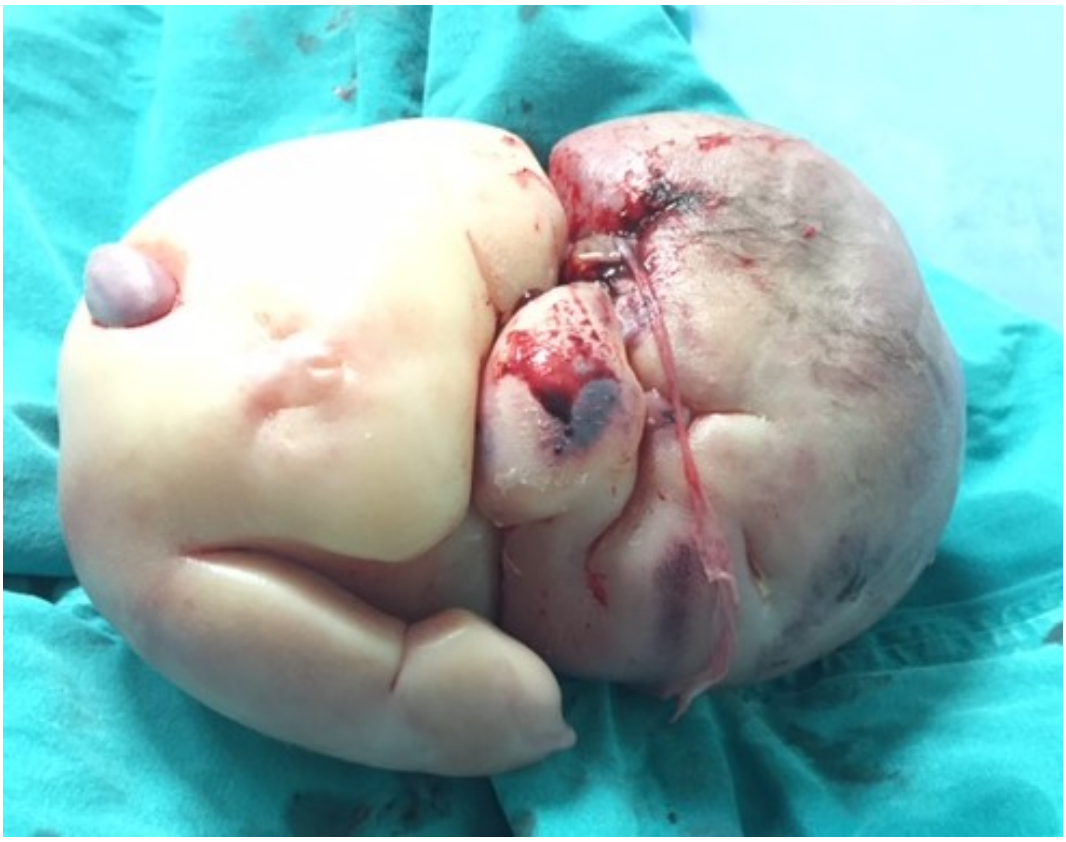

Figure 2. Twin 2 acardiac twin baby after birth. Birth weigth was 1550 gr and 30 cm.

Discussion

TRAP sequence is an unusual complication of monochorionic twin pregnancy with an incidence of 2.6%.[4] In monochorionic twin pregnancies, if one of the twins is detected to have structural deformities and it continues to grow in follow-ups despite the absence of fetal heart beat, TRAP sequence should be suspected. The diagnosis is confirmed by visualizing anastomoses between the twins with doppler sonography. With a detailed ultrasonographic examination it can be diagnosed in the first trimester of pregnancy, as early as the 11th gestational week.[5] Once the diagnosis is finalized as TRAP sequence, the ratio of the weight of the acardiac twin to the pump twin can predict fetal outcome. Moore et al. showed that when the ratio of the weight of acardiac twin to that of the pump twin is >0.70, the risk of preterm delivery and hydramnios increases. The perinatal mortality rate is 50% or more in these cases.[6] In the present case there is a minimal difference between twins' weight. The pump twin should be evaluated with frequent doppler studies of the middle cerebral artery for the risk of anemia and other possible complications. Umblical artery and ductus venosus blood flows can be also indicators of the pump fetus' prognosis.

There are two types of approaches in the literature; the conservative approach and the antenatal interventional approach such as laser coagulation or radiofrequency ablation. Patients should be informed about the interventions and benefits and risks should be discussed together. Complications such as maternal bleeding, thermal injury, chorioamnionitis, and disseminated intravascular coagulation have been reported after these procedures.[7] Before intervention, karyotype analysis of the pump twin should be done because the probability of an abnormal karyotye is up to 9%. Chromosomal abnormalities have been detected in one third of the acardiac cases.[2] The conservative approach can be considered if the acardiac fetus is small, its growth is rather slower and the pump fetus has no complications.[8,9] Yet expectant management can yield to perinatal mortality due to the development of heart failure or preterm delivery as shown in a case series of Pagani et al.[10] It's controversial whether conservative management or therapeutic treatment is better in TRAP-sequence. It seems appropriate to choose an interventional approach in the cases of hydrops, polyhidroamnios and abnormal vascular anastomosis.

Our goal should be to improve the outcome of the pump twin by minimalizing complications with close fetal monitoring with weekly ultrasound examinations or more frequently if necessary. As there is a risk of preterm labor, antenatal corticosteroids should be administered before 34 weeks of gestation. We have to approach individually each patient and consider the most beneficial treatment modality.

The uniqueness of this case, was that there was no significant difference in the weight of both twins. The acardiac twin was as large as the pump twin. Except for twenty days hospitalization of the female twin, because of neonatal respiratory distress syndrome, there were no complications due to conservative management.

Conclusion

Monochorionic pregnancies should be carefully evaluated with first trimester ultrasound examination and TRAP sequence should be kept in mind as a complication of it. When TRAP sequence is diagnosed fetuses that are suitable for conservative management and fetuses which are candidates for interventional treatment should be selected carefully. Considerations should include fetal weigth differences, amniotic fluid measure, and pump fetus well-being.

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