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Acardiac Acephalus Twin Pregnancy with Poor Prenatal Outcomes: A Case Report

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Abstract

Introduction: Acardiac twin refers to a monochorionic pregnancy in which there is a twin with absent or nonfunctioning heart and a normal co-twin. This was a very rare disorder of monozygotic twin pregnancy. The aim of this report was to introduce a case of acardiac acephalus twin pregnancy.
Case Presentation: A 32- year- old woman (G3 P2 L2) referred to the hospital because of a decrease in fetal movement at 32 weeks of gestation. Ultrasonography showed a dead fetus. Her earlier sonography showed the possibility of acardiac twin pregnancy, but she did not have any appropriate or special prenatal care for this problem. After delivery, there was a normal dead fetus without any gross abnormality and another acardiac fetus.
Conclusions: Early diagnosis of acardiac twin pregnancy and appropriate prenatal care are of prime importance for appropriate intervention and best fetal outcomes.

Keywords: Acardiac Acephalus, Twin Pregnancy, Twin Reversed Arterial Perfusion

1. Introduction

Acardiac twin refers to a monochorionic twin pregnancy, in which there is a twin with absent, or nonfunctioning heart and a normal co-twin. This was a very rare disorder of monozygotic twin pregnancy (1). In monochorionic twin pregnancies, TRAP (twin reversal arterial perfusion) should be suspected; the definitive diagnosis of this syndrome is made when pulsative flow is seen in the umbilical artery goes towards the acardius (in Doppler Sonography) (1).

In this syndrome, the pump twin has a normal pattern of fetal circulation and a portion of its cardiac output travels through placental arterial-arterial anastomosis to the umbilical artery and finally to the systemic circulation of the recipient co-twin. Anastomosis may occur arteriovenous and veno-venous. The degree of this syndrome depends on the type of anastomosis and time of their development.

This anastomosis is common in monochorionic twin pregnancies and this is not the only etiologic factor of this syndrome (2).

This syndrome may be seen in fetuses between 18 and 27 weeks of gestation with signs of poor prognosis, including the presence of hydramnios, increase in the relative size of the acardiac twin, hydrops in the pump twin, mono amnioitic pregnancy and ratio of weight of the acardiac twin/weight of the pump twin greater than 0.7 (3).

In this syndrome, choice of treatment is dependent on the gestational age of the fetus. At the gestational ages of less than 16 weeks, the common intervention is intra- fetal laser treatment, and at the gestational ages greater than 16 weeks, the best treatment is radio frequency ablation (RFA) to ablate the anastomosis (4).

In TRAP without poor pregnancy criteria like hydramnios, weekly sonography is made for the early diagnosis of fetal hydrops. Some suggest the administration of a course of antenatal steroids and termination of these pregnancies at 34 to 36 weeks of gestation. Cesarean delivery is done for the usual obstetrical indications (1).

In this case report, we present a case of acardiac acephalus twin pregnancy.

2. Case Presentation

A 32-year- old woman (3rd gravid), who had two full term vaginal deliveries, para two, living two, referred to academic hospital of Mashhad University of Medical Sciences in August 2014 because of a decrease in fetal move-

ment at 32 weeks of gestation. Fetal heart rate was not detected by ultrasonography, so fetal death was confirmed.

She had a sonography that reported a single live fetus with normal amniotic fluid and normal placenta, the gestational age was 18 ± 1 w (based on bilateral diameter (BPD) and femur length (FL).

There was an abnormal and incomplete anatomical structure like edematous limbs near the live fetus which had no abnormality. Again, at 21st week of gestation, she underwent sonography, and there was a live fetus with normal amniotic fluid (AF), anterior placenta, 21 weeks, 4 days based on abdomen circumference (AC), BPD, FL, without any structural abnormality, plural effusion or ascites in this fetus. Moreover, there was an abnormal structure like edematous limb and elbow near the normal fetus, and there was no attachment between them (between the abnormal structure and the live fetus). The patient was not educated enough to seek more diagnostic or therapeutic medical procedure for this problem.

Finally, she was admitted because of decreased fetal movement and diagnosis of fetal death at 32nd weeks of gestation. At vaginal examination, the cervix was unripe. For termination of pregnancy, 50 μg sublingual misoprostol every four hours was administered. Normal vaginal delivery was done after five doses of misoprostol. A preterm dead male fetus, with 1700 grams weight and normal appearance was delivered. Then the second abnormal fetus was delivered without head, neck, chest and upper limb, but with only two edematous lower limbs with abnormal fingers. There was just the inferior part of the trunk with the cut umbilical cord and 520 grams weight (Figures 1 and 2).

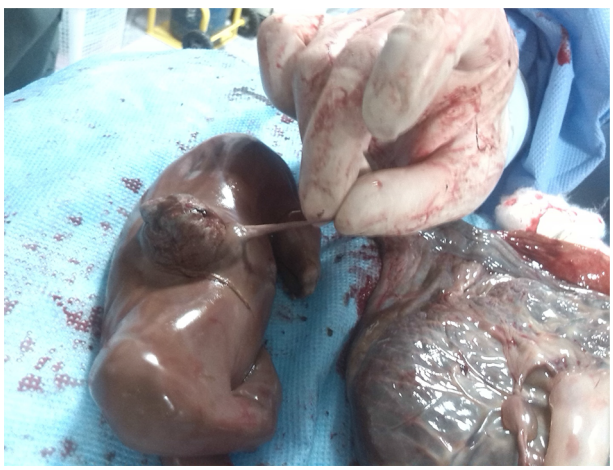

Figure 1. The Inferior Part of the Trunk Within the Cut Umbilical Cord

The placenta was delivered spontaneously and there was just one normal placenta. There was about 60-centimeter length of very narrow umbilical cord of the second anomalous twin that was tightly twisted around the normal fetus umbilical cord two times, and this could have been responsible for narrowing the normal umbilical cord and even occluded blood circulation in the normal fetus. After examining the placenta, a monoamniotic monochrionic placenta with one normal umbilical cord that had two arteries and one vein and another abnormal cord that had two vessels with thrombosis were found. The mother was discharged one day after delivery with good general condition.

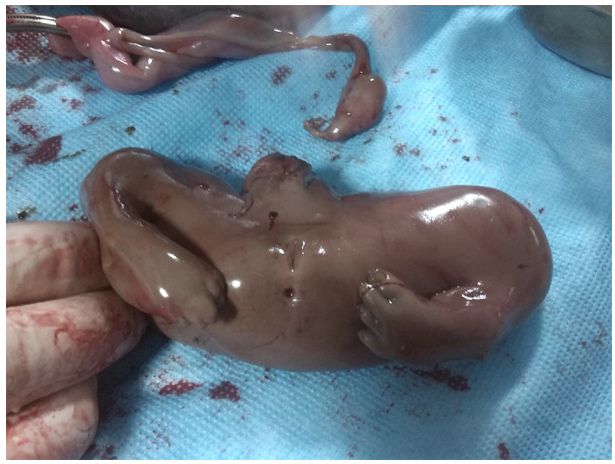

Figure 2. The Second Abnormal Fetus Delivered without Head, Neck, Chest and Upper Limb, but with Only Two Edematous Lower Limb and Abnormal Fingers

3. Discussion

Acardiac twin pregnancy is a syndrome with vascular anastomosis and absent or malfunctioning heart in a twin. The degree of anomalies is variable in this syndrome. The acardiac phenotype ranges from developed lower extremities, abdomen or pelvic to a tissue mass which is not recognizable as fetal parts (5). This syndrome is common in monochorionic twins, and the umbilical cord contains two vessels in 70% of the acardiac fetuses (6).

In a study by Healey (7) in 1994, five cases of acardiac twins were reported. Acardiac twin was more common in primigravid women with monoamniotic monochrionic twin pregnancy. Umbilical cord consisting of two vessels, and preterm labor before 32 weeks of gestation was common.

This syndrome is more common in female twins, and because the disorder is monozygotic, the twins are usually of the same gender (8).Our patient had a monoamniotic monochrionic twin pregnancy, but she was multiparous.

Acardiac twin pregnancy is classified into four forms: 1) Acardiac acephalus: The fetal head and thoracic organs are absent (this is the most common type); 2) Acardic acormous only: Just the fetal head develops; 3) Acardius amorphous, which consists of different shapeless tissues; 4) Acardius myelocephlous in which head and one or more extremities are partially developed (5).

In this syndrome, the pump twin has a normal pattern of fetal circulation and a portion of its cardiac output travels through the placental anastomosis to the umbilical artery, and finally to the systemic circulation of the recipient co-twin, creating reverse circulation (2).

The pump twin may have signs of cardiac failure, ascites, pleural and pericardial effusion, cardiomegaly, hydramnios and tricuspid regurgitation. Degree of heart failure depends on the ratio of the weight of acardiac twin to the pump twin, and if it exceeds 0.7, the risk of heart failure is increased (9).

In the study of Aggarwal in 2002, he reported a case of acardiac twin pregnancy that was terminated in 36 weeks of gestation. The ratio of the size of acardiac twin to that of the pump twin was 0.67. There was no congestive heart failure in the normal fetus (5).

Our case was an acardiac twin pregnancy in which intrauterine fetal death occurred and there was no sign of ascites or poly hydramnios in the normal fetus and the weight's ratio was 0.32.

Diagnosis of this syndrome was made with ultrasonography that is usually helpful from 11 weeks of gestation. Nevertheless, in this case, no diagnosis of acardiac twin was made by ultrasonography until 18 weeks.

In the study of Bornstein in 2008 on the diagnosis of twin reversed arterial perfusion, it was reported that three-dimensional and Color Doppler Ultrasonography are useful for diagnosis of TRAP (9). Early sonographic diagnosis, intervention via cord occlusion and laser ablation, bipolar cord coagulation and radiofrequency ablation under local anesthesia and appropriate follow-up will improve the pregnancy outcomes (10, 11).

3.1. Conclusion

Early diagnosis of acardiac twin pregnancy by ultrasonography and regular follow-up by serial sonography or Color Doppler Ultrasonography for the early diagnosis of poor conditions and appropriate intervention are crucial to achieve optimal acardiac pregnancy outcome.

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