

Acardiac Fetus in a Triplet Pregnancy: A Case Report

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Abstract

Background: Acardiac twinning is a rare complication of monozygotic multifetal gestation that is thought to be the consequence of twin reversed arterial perfusion syndrome. In our case, an acardiac fetus in a triplet is presented and expectant management in the follow-up of the pregnancy is discussed.

Case: He the patient, 25 years old, with 16 weeks of pregnancy for spontaneous triplets admitted to our hospital for her first antenatal obstetric visit. In the ultrasonographic examination; two fetuses with cardiac activities and measurements in concordance with the gestational weeks were observed besides a mass with no cardiac activity in another amniotic sac. After the Doppler sonography revealed reverse flow in the umbilical cord of the mass, the diagnosis was made as acardiac fetus. The fetuses were followed up with expectant management. In the 34th gestational week, she had preterm premature rupture of membranes and delivered by cesarean section. Two healthy babies and a 1610 gr. acardiac fetus were born.

Conclusion: Acardiac fetus, which is as rare as one in 35000 births and usually observed in monozygotic twins, is reported in a spontaneous triplet pregnancy in our case and healthy twins were delivered successfully with expectant management.

Keywords: Triplet pregnancy, acardiac fetus, expectant management.

Üçüz gebelikte akardiyak fetus: olgu sunumu

Amaç: Akardiyak fetus monozygotik multifetal gestasyonun çok nadir bir komplikasyonudur ve ikizden ikize ters arteriel perfüzyonun sonucu olduğu düşünülmektedir. Bu yazıda çok daha nadir görülen üçüz gebelikte akardiyak fetus ve bu gebeliğin beklentisel yaklaşımla izlemi sunulmuştur.

Olgu: Hastanemiz gebelik polikliniğine gebelik kontrolü için ilk kez başvuran 25 yaşındaki, 16 gestasyonel haftalık spontan üçüz gebeliği olan olguya rutin ultrasonografik tarama yapıldı. Ultrasonografik değerlendirmede gestasyonel yaşla uyumlu ölçümleri olan ve kardiyak aktiviteleri bulunan iki fetus ve ayrı amniotik kese içinde kardiyak aktivitesi olmayan, organları ayırt edilemeyen kitle saptandı. Doppler ultrasonografide kitlenin umbilikal kordundan ters akım izlendi ve akardiyak fetus tanısı kondu. Beklentisel yaklaşımla yakından izlenen gebe, 34. haftada erken membran rüptürü nedeniyle sezaryene alındı. Sağlıklı ikiz bebeklerin yanı sıra 1610 gr. ağırlığında akardiyak fetus doğurtuldu.

Sonuç: Otuzbeşbin doğumda bir kadar nadir ve genellikle monozygotik ikizlerde gözlenen akardiyak fetus, olgumuzda spontan üçüz gebelikte saptanmış ve beklentisel yaklaşımla sağlıklı ikizler doğurtulmuştur.

Anahtar Sözcükler: Üçüz gebelik, akardiyak fetus, beklentisel yaklaşım.

Introduction

Acardiac twinning, known also as twin reverse arterial perfusion (TRAP) sequence, is a rare complication of monochorionic placenta-

tion occurring in 1% of monozygotic twins and in 1:35.000 births.¹ In this case, a constitutionally normal “pump” twin and a parasitic acardiac twin is in question. Twin to twin transfusion, hinders the organogenesis in the acardiac

receiver twin. Even though the acardiac twin is a fetus with serious malformations, lacking most of the organs, especially the heart, during pregnancy it grows via the arterioarterial and venovenous placental anastomosis from the pump twin. In most of the cases, the continuous growth of the acardiac twin and the “vascular stealing” phenomenon causes cardiac failure, polyhydramnios and prematurity in the pump twin and in 50% of the cases perinatal death of the constitutionally normal twin.²

Acardiac fetus may be diagnosed in early monochorionic twin pregnancy when a deformed twin with no cardiac activity is seen in ultrasonographic examination or may be recognized during the ultrasound scans in the further weeks of pregnancy after clinical symptoms like polyhydramnios occur. The reversed blood flow in the umbilical artery (TRAP sequence) may be shown in color Doppler sonographic examination.

Management options in acardiac fetus include elective termination; expectant management (with serial cardiotocography, ultrasonography and ecocardiography); non-invasive symptomatic procedures like indomethacine, digitalization and tocolysis; and in appropriate cases invasive surgical procedures. In many cases invasive procedures like the selective delivery of the acardiac twin or umbilical cord ligation through a hysterotomy; the embolization of the umbilical cord using alcohol platinum coils or thrombogenic coils via ultrasound or by endoscopy; or the laser ablation of the umbilical cord have led to successful conclusions.

Case Report

A 25 year old primigravid with a spontaneous triplet pregnancy admitted to the obstetric outpatient clinics of S.B. Aegean Maternity and Women’s Health Teaching Hospital for her first antenatal visit at 16th gestational week. In

the ultrasonographic examination; a mass with no cardiac activity and no organs other than an extremity was observed in a separate amniotic sac besides two fetuses with cardiac activities and biometric measurements concordant with the gestational age. The triplets observed in three amniotic sacs were triamniotic mono-chorionic and amnion fluid was normal. The Doppler ultrasonography revealed venous blood flow with no pulsation in the anatomic location of the heart. The mass was diagnosed as acardiac fetus when reversed blood flow from the umbilical cord of the mass was observed. The ultrasonographic examination revealed no fetal anomaly in the twin fetuses. Since the frequent periodic evaluations, suggested normal growth patterns of the twins with normal amnion fluid indexes, normal Doppler flow measurements and reactive non-stress tests; the case was followed with expectant management.

In the repeat ultrasonographic scan healthy twins of 32 weeks of gestation were observed and in another amniotic sac, a 131x168 mm. viscous mass showing 52 mm. long echo of a probable femur and smaller echoes of bone density of a probable tibia and fibula was noted. In the 34th gestational week, the patient was admitted to the hospital with preterm premature rupture of membranes and she delivered via cesarean section. Healthy twins weighing 1800gr. and 1700gr., and a 1610gr. acardiac triplet were delivered. In the pathological examination, the acardiac triplet was a 20x15x15cm. ovoid mass covered with skin and had on one side 13cm. long, 0.5cm. in diameter umbilical cord, and two feet like structures (Figure 1). When it was dissected near the umbilical pole; cartilage and muscle structures in a lipid gelatinous matrix were observed besides lung tissue but no organ was distinguished. The case was performed an emergency cesarean section and due to technical error during transportation, the placenta could not be examined pathologically.



Figure 1. Acardiac fetus.

Discussion

Acardiac fetus is the most serious complication seen in monozygotic multiple pregnancies with twin to twin transfusion syndrome (TTTS). Early diagnosis is important for proper management of the case so detailed studies have been done on the clinical and anatomical features of acardiac twins. In the study of Chanoufi et al. including six acardiac twins, severe agenesis or hypoplasia of thoracoabdominal organs were frequently seen and arms were noted to be more affected among extremities.³ The weight ratio of the acardiac fetus to the “pump” twin changed between 50-142%. The researchers reported that a deformed fetus with no cardiac activity encountered in the ultrasonography of a monochorionic twin would lead to the diagnosis of acardiac fetus; and cardiac insufficiency, polyhydramnios and a weight ratio of greater than 50% were prognostic factors for the “pump” twin.

Bannykh and colleagues underlined that on normal genetic ground, the severe restriction in the brain perfusion might also lead to the neuronal migration defects caused by failure of prosencephalic unfolding and deterioration of the glia-pial border.⁴ Spencer claimed that some

cases may be defective fetuses developing from genetically defective embryos since chromosomal abnormalities are encountered and these cases are more frequent in familial twin pregnancy history and in girls.⁵ He also noted that taking into consideration the lack of brain and heart in acardiac fetus; a primary cardiac malformation yielding to secondary brain development failure may play a role in etiology. Shih et al. investigated the interfetal hemodynamics in acardiac twins and reported that acardiac twins could be the reason of placental vascular anastomosis involving artery to artery or artery to vein anastomosis as well as the result of them.⁶

Defined high risk factors for bad obstetric outcome are acardiac aneups, polyhydramnios, acardiac twin with ears and pump twin with cardiac failure. At the moment, there are no defined intervention methods for different clinical presentations since there are no long term follow-up data on living pump twins.

Ultrasound guided percutaneous laser coagulation of intraabdominal vessels have been proposed as an effective and minimally invasive method.⁷ Sepulveda and colleagues tried percutaneous laser coagulation via epidural anesthesia in an acardiac twin at 25th gestational week which had been followed up with expectant management since 12th gestational week but showed cardiomegaly and polyhydramnios in the pump twin and serious overgrowth in acardiac fetus in the second trimester.⁸ Concurrently 2200 ml. of amniocentesis was performed. During the follow-up the acardiac mass got smaller and the pump twin showed normal fetal growth. The authors reported that the procedure was technically hard and it should only be tried when the overgrowing acardiac fetus starts to decompensate the pump twin in previable gestation.

The fact that the umbilical cord of the acardiac fetus is abnormally short, thin and easily squashed; the possible cord accidents because of the proximity of single umbilical artery and vein; and the fact that the healthy twin may die because of the possibility of ablative material mixing up in the blood stream of the pump twin have made trials involving the umbilical cord of the acardiac twin incredibly hard.

Rodeck and colleagues evaluated thermocoagulation with Doppler sonography which they performed in four cases in the second trimester as safe and effective.⁹ In a literature review including a total of 22 invasive procedures in acardiac twins with seven of them being endoscopic laser coagulation, Arias et al. stated that the mortality in the pump twin was 13,6% with fetal surgery and 50% with expectant management.¹⁰

Hirose and colleagues performed radiofrequency ablation to a hydropic pump twin at 27th gestational week and reported that the hydrops dissolved at 32nd gestational week.¹¹ In the literature review of Tan and Sepulveda consisting of 32 articles about minimally invasive techniques performed on 74 acardiac twins; the mean procedure week was the 21st gestational week (range 14-28th gestational week) and mean delivery week was 36th gestational week (range 19-42nd gestational week)¹² In the review reporting 76% as mean survival rate of the pump twin; intrafetal ablation (alcohol, monopolar diathermia, interstitial laser, radiofrequency) was correlated with a later delivery week, lower technical error rate, lower premature birth rate and higher clinical success rate compared to cord occlusion techniques (embolization, cord ligation, laser coagulation, bipolar diathermia, monopolar diathermia).

Sepulveda and colleagues reported 63% of pump twin survival rate as a result of intrafetal absolute alcohol chemosclerosis performed on

8 hydropic acardiac twins at five different centers.¹³ Three pump twins were lost as a result of the intravascular transfer of alcohol. The authors stated that the procedure was simple but insisted that it should be done in pregnancies with bad prognostic factors or in countries where more sophisticated therapy methods are unavailable.

In our case when the acardiac fetus was diagnosed, detailed fetal anomaly scanning of the other fetuses was done and since they showed healthy fetal growth with no anomaly expectant management with ultrasonographic examination and Doppler sonography in every two weeks and periodic non-stress tests in the later weeks was decided on. No invasive procedure was necessary since the pump twins showed no cardiac failure or polyhydramnios and the acardiac mass did not overgrow. As Sepulveda stated, prenatal procedures should only be used as a therapy method in complicated cases which would benefit from the procedure but not as prophylaxis in inappropriate cases.⁸ The most important factor in deciding on the correct choice of management is the early diagnosis of acardiac twin.

Kaynaklar

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