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Acardiac Parabiotic Twin: A Case Report

The incidence of anomalies is high in multiple gestations especially in monochorionic twins. This paper reported a case of an acardiac parabiotic twin, a rare deformed fetus which occurs typically in monochorionic twin gestations. It is a severely deformed fetus without heart and limited development of the upper half of the body.

Keywords: acardiac twins, monochorionic

Introduction

Acardiac parabiotic twin is a bizarre phenomenon with an incidence of 1 in 35,000 pregnancies which occurs only in 1% of monochorionic twin pregnancies, through the arterial-arterial and venous-venous anastomoses within the placenta. Despite the relative rarity of the condition, it has a high mortality rate of 50%–75% and morbidity in pump twin.[1-5] In this condition, a severely malformed fetus is seen, most often with no formed heart (although occasionally with an inadequate very small two-chambered heart) and limited development of the upper half of the body characterized by anencephaly or small rudimentary head, holoprosencephaly, sever brain malformation or absent head and absent or hypoplastic upper torso and limbs (cervical spine, arms and ribs). Heart, lung and abdominal visceras and external genital organs may all be absent or severely malformed; pelvis and lower extremities usually have a better development. Multiloculated cystic hygroma is generally present and severe oligohydroamnious is also seen.[1,2,5-7]

Case Presentation

A 42-year-old pregnant woman in her 5th pregnancy ($G_5P_2A_2L_2$) with triplet pregnancy referred to the radiology clinic for routine pregnancy sonography. The mother did not suffer from any systemic illness. Her marriage was not consanguineous and she had no infection or drug intake during early pregnancy. No congenital anomalies were detected in maternal past family history. At sonographic evaluation, two dichorionic diamniotic normal 21-week female fetuses were seen. In the second twin, the amniotic sac had a normal amniotic fluid volume with a large soft tissue mass with deformed skeletal elements. It had an umbilical cord with a single artery, which was inserted to the pump twin shared placenta. Head and neck, heart, viscera and upper limbs were not detected; only a small lower lumbar vertebral column and a small hemipelvic with fully-formed right lower limb and a short femur on the left side were seen(Fig. 1). All these signs suggested acardiac twin.

The pump twin was normal, no complication was detectable until the 27th week of pregnancy when mother had preterm labor and two normal premature female

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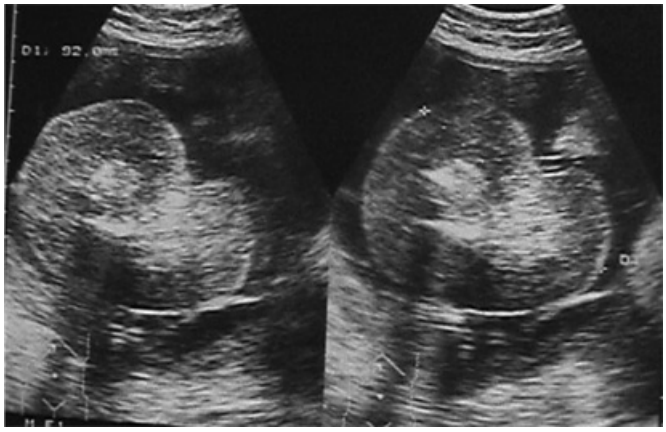

Fig. 1. Sonography of the acardia fetus that shows deformed lower limbs with no heart and upper half of body.

fetuses were born by cesarian section; a severely deformed third fetus was also born which had only two deformed lower limbs and a short single-artery umbilical cord. Head and neck and upper limbs and genital organs were not seen (Fig. 2). The pump twin had a weight of 1100 g; the acardiac twin was not weighed.In x-ray study, the acardiac twin had only a short lumbar vertebra with a small hemipelvis, a small right lower limb and only a left femur; no other skeletal bones were formed. In computed tomography, no viscera or heart was detected (Figs. 3 and 4).

In color Doppler study of the acardiac umbilical cord, the direction of blood flow was reversed which caused blood entering the body of the acardiac fetus from the umbilical artery instead of the vein.

Unfortunately, normal fetuses died of prematurity and respiratory distress syndrome (RDS). However, no other complication was found in the pump twin.

Discussion

Acardiac parabiotic twin is a rare and bizarre phenomenon, which occurs only in monochorionic twin pregnancies through the arterial to arterial and venous to venous anastomoses within the placenta.[1] The flow direction in the umbilical arteries and vein of the recipient fetus is reversed and tissue perfusion of this non-viable fetus is accomplished by the artery to artery and vein to vein communications in the placenta from the circulatory system of the co-twin referred to as the "pump twin" [1,2]

Because the acardiac twin receives poorly oxygenated blood through the umbilical arteries, the structures supplied by the iliac arteries and distal abdominal aorta are relatively well perfused whereas the upper body and head receive essentially no oxygenated blood. Thereby, acardiac twin had limited development of the upper half of the body.[1,2,6]

Mortality of the acardiac twin is universally either *in utero* or at the time of delivery. The normal pump twin is at increased risk because of high cardiac out put and congestive heart failure, hydrops and polyhydramnios. The overall perinatal mortality of pump twin is approximately 50%–75%. The occurrence of preterm delivery, polyhydramnios and heart failure are all strongly associated with ratio of the weight of acardiac to the pump twin. If the weight of acardiac twin is >70% of the pump twin, preterm labor occurs in 9%, polyhydramnios in 40% and congestive heart failure in 30% of instances. If the weight ratio is <70%, the rates are 75%, 30% and 10%, respectively.

Management strategies for pump twin only include tocolytic agents and volume reduction amniocentesis to treat preterm labor and frequent interval monitoring studies for detection of signs of congestive heart failure. If fetal distress develops, invasive procedures, such as ligation of the cord of acardiac twin or radiofrequency ablation, are done.[1-3,5,6,8,]

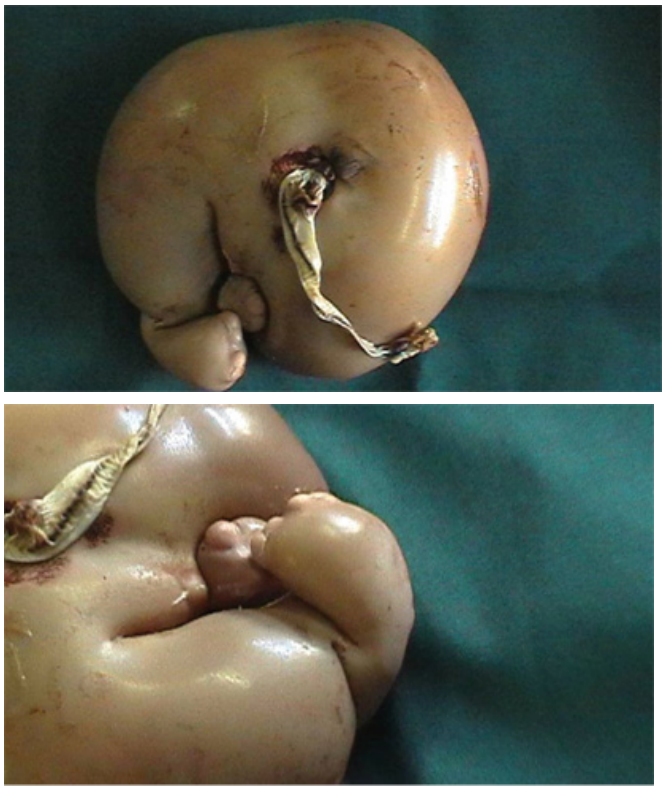

Fig. 2. A and B. Gross anatomy of the acardiac fetus with single arterial umbilical cord.

Fig. 3. X-ray of the acardiac fetus that shows deformed lower limbs, pelvic and lumbar vertebral skeletal structure.

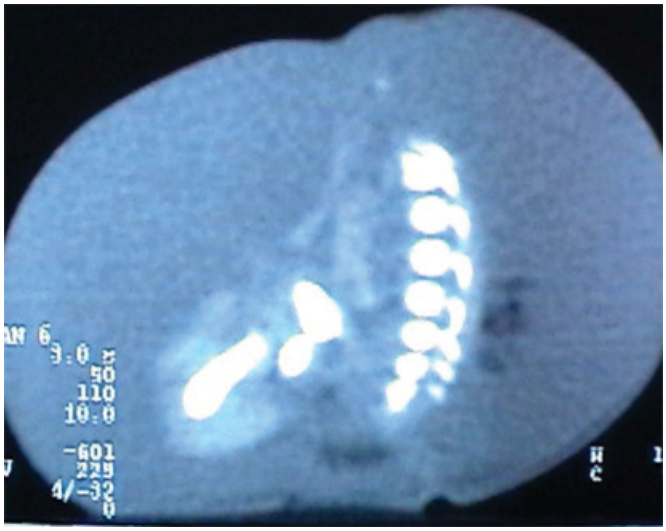

Fig. 4. Computed tomographies of the acardiac fetus, no viseral structures are detected.

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