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CASE REPORT

Acardiac Twin: A Rare Case Report

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ABSTRACT

The incidence of multifetal pregnancies has dramatically increased due to assisted reproductive technology. As a result, preterm labor, Preterm premature rupture of the membranes, congenital anomalies and fetal losses are commonly encountered by obstetricians. Despite advances in diagnostic modalities and prenatal care, many times the complications associated with multifetal pregnancies may remain undiagnosed. Complications are more with monochorionic twin gestation due to placental sharing. ''Twin – reversed arterial perfusion'' sequence or ''acardiac twin'' is one such complication. This case report presents an antenatally undiagnosed monochorionic twin pregnancy, resulting in preterm delivery of a healthy, normal twin along with an acardiac acephalus co-twin.

Keywords: Acardiac acephalus, Monochorionic, Twin – reversed arterial perfusion, Twins.

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INTRODUCTION

Monochorionic twinning often results in a number of serious complications due to sharing of a common placenta and abnormal placental vascular anastomoses between the twins. One such rare, serious complication is "twin reversed arterial perfusion" (TRAP) sequence, also known as "acardiac twin." Acardiac twins lack a heart and may have associated failure of head and limb growth along with a spectrum of various other anomalies. This case report presents an antenatally undiagnosed monochorionic twin pregnancy resulting in preterm delivery of a healthy, normal twin along with an acardiac acephalus cotwin, with maldeveloped limbs and omphalocele.

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CASE REPORT

Our patient was a 30-year-old gravida 3 para 2, all spontaneous conceptions, with both living issues delivered normally at term. She was registered at an outside hospital at 26 weeks of gestation and had a poor antenatal follow-up of only two visits. There was no early obstetric ultrasound or fetal anomaly scan. The only ultrasonography (USG) done was at 26 weeks, which reported a single live intrauterine pregnancy with all fetal and placental parameters within the normal limits.

The patient presented to us at 34 weeks of gestation in preterm labor. Per abdomen, the fundal height corresponded to 36 weeks; multiple fetal parts were felt with a single fetal heart sound audible. On per vaginal examination, a well-effaced, 4 cm dilated cervix with tense bulging bag of membranes and vertex presentation was noted. Within 2 hours following admission, the patient delivered a female weighing 1.6 kg appearing normal, with no external abnormality. While an attempt to deliver the placenta was made, fetal limb parts were felt, confirming the presence of another fetus *in utero*. Consequently, the second fetus was delivered by breech presentation. This was a female fetus weighing 1.1 kg, with anomalous appearance without head (acephalus), no cardiac activity, swollen malformed limbs, equinovarus deformity in both feet, an external sac containing abdominal viscera overlying the trunk (omphalocele). A single placenta was delivered weighing 620 gm with two umbilical cords (monochorionic diamniotic). An X-ray examination of this fetus was carried out, which confirmed our clinical findings (Figs 1 to 3).

DISCUSSION

Acardiac twin or TRAP sequence is a very rarely encountered consequence of monochorionic multifetal gestation. Its incidence is 1 in 35,000 births.[1] The TRAP sequence results mostly due to a large arterioarterial placental shunt and consists of a normally formed donor or "pump" twin that has an increased arterial perfusion pressure and a recipient twin congenitally lacking a heart (acardiac) which receives deoxygenated blood pumped by its cotwin due to reversal of flow in the umbilical artery. There is preferential perfusion of lower body of the acardiac fetus through iliac vessels. Thus, only the lower body is well developed, while there is maldevelopment or atrophy of upper body parts, such as head, neck, and upper limbs.

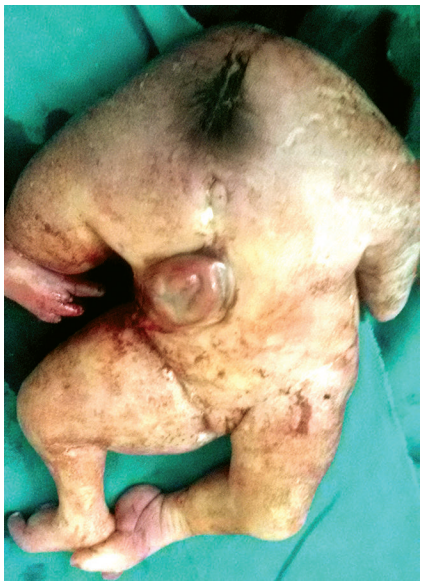

Fig. 1: Acardiac acephalus fetus

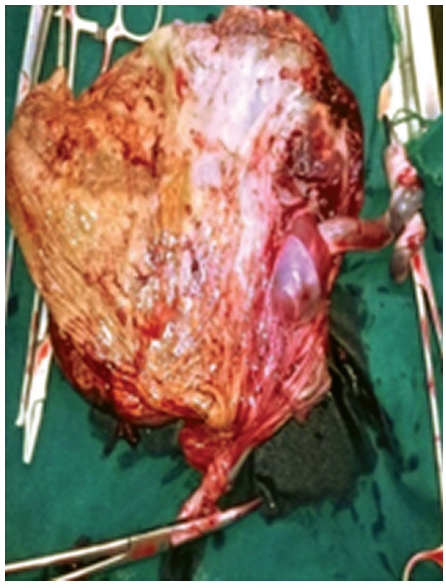

Fig. 2: Two-cord placenta

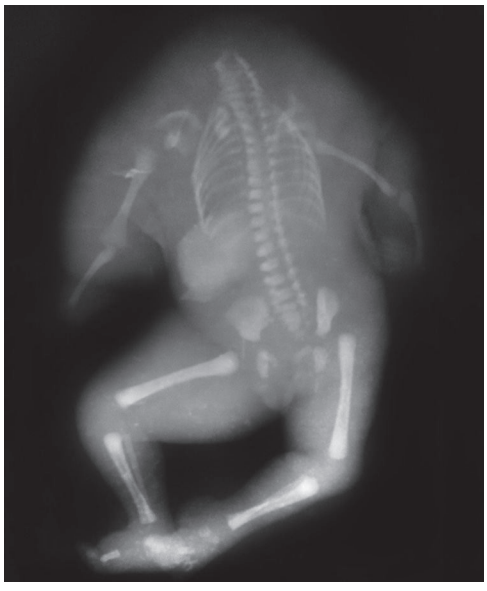

Fig. 3: Fetal X-ray

Very rarely, the twin may develop only a mass of tissue and no identifiable body structures. Acardiac anomaly is more common in monozygotic female twins and mortality is 100%.[2]

The acardiac twin may be of the following types based on the degree of cephalic and truncal maldevelopment:

- *Acardius acephalus*: Failure of head and upper limb growth.
- *Acardius anceps*: Partially developed head with identifiable limbs.
- *Acardius acormus*: The only developed structure is the head, and truncal structures are essentially absent. This is the rarest variety.
- *Acardius amorphous*: Failure of any recognizable cephalic or truncal structure. Least differentiated variety.[3]

The pump twin, though structurally normal, may develop congestive cardiac failure and hydramnios associated with preterm labor. Mortality is 50 to 70% in pump twin, and survival rate can be improved by early detection of monochorionic twinning with acardia on USG and Doppler and treatments like radiofrequency or fetoscopic laser ablation of anastomotic vessels.[4]

CONCLUSION

Acardiac twin is a rare, severe congenital malformation seen in monozygotic twin gestation, which is incompatible with life. Early detection of acardiac twin can be done in 1st trimester itself by USG and Doppler and is essential for initiating timely treatment and preventing complications, such as cardiac failure in pump twin, preterm labor; thereby improving the perinatal outcome.

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