CASE REPORT

Acardiac aniceps: a rare congenital anomaly

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SUMMARY

Acardiac twin is a rare congenital anomaly and is exclusively associated with monochorionic twin pregnancies. The abnormalities occur due to abnormal communication between the two fetuses in the form of arterioarterial and venovenous communications, resulting in a grossly abnormal acardiac twin with reduction anomalies mainly of the upper body and gross oedema. Since no two acardiac twins are alike, this case report will add to the acardiac twin anomaly spectrum.

BACKGROUND

Acardiac twin is a rare congenital anomaly of monozygotic twin pregnancy, which occurs due to twin reversed arterial perfusion (TRAP) sequence. The incidence of an acardiac twin is 1 in 35 000 pregnancies¹ and it affects approximately 1% of monochorionic twin pregnancies. TRAP sequence is caused by arterioarterial and venovenous anastomosis between both cord insertions and this may lead to reverse perfusion via the umbilical artery of one twin. This condition is characterised by a normally formed donor twin and a severely malformed recipient twin who lacks the heart.

CASE PRESENTATION

A 30-year-old woman, gravida 2 with one live issue and 33 weeks of pregnancy from her last menstrual period, presented with preterm labour pains. She had a twin pregnancy with intrauterine death of one fetus. There is no previous history of antenatal check-up. Her general physical and systemic examination was within normal limits. Abdominal examination revealed a 34-week size uterus, with fetus in longitudinal lie and cephalic presentation. Her vaginal examination revealed cervical os 2 cm, 50% effaced, vertex high up.

INVESTIGATIONS

The patient's obstetric ultrasonography revealed diamniotic, monochorionic twin pregnancy. One of the twin fetuses had normal cardiac activity with an average maturity corresponding to 30 weeks 4 days and with no gross congenital anomaly. The other twin was an amorphous mass with some skeletal, soft tissue and cystic components without any cardiac activity (figure 1). The lungs, brain, abdominal viscera and upper limbs were not seen in the obstetric ultrasonography but a single lower limb was identified. On Doppler ultrasound, an occasional trickle of flow was seen in the mass.

OUTCOME AND FOLLOW-UP

The patient delivered vaginally a normal pump twin weighing 2000 g and an acardiac twin whose

Figure 1 Antenatal ultrasound showing (A) the empty and partially formed skull, (B) subcutaneous oedema and (C) deformed skeletal structures with no identifiable organ in the acardiac twin.

weight was 1500 g and length 29 cm. The normal twin showed no signs of cardiac failure and had no obvious congenital malformations. The acardiac twin was a round mass of soft tissue with few hairs at one end and a small limb-like structure at the other end (figure 2), and was covered with thick, oedematous skin. Postmortem radiograph (figure 3) of the acardiac twin showed a disproportionate large soft tissue mass with deformed, partially formed skeletal structures. There was a rudimentary vertebral column and few skull and facial bones. Few deformed bilateral upper ribs were also seen. A hemipelvis with unilateral femur, tibia, fibula and few bones of the foot was noted. Autopsy revealed an amorphous mass filled with serous fluid and fat. No organ structure could be identified apart from one small kidney and an empty partially formed skull (figure 4). The placenta was monochorionic and diamniotic, and it

Figure 2 Acardiac twin after delivery, arrow (A) depicting cephalic end with few hairs, arrow (B) showing limb bud, arrow and (C) showing thin atrophic cord.

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Figure 3 Postmortem skiagram of the fetus showing (A) skull bones, (B) ribs, (C) vertebral column, (D) hemipelvis, (E) femur, (F) tibia, (G) fibula and (H) soft tissue.

weighed 450 g. Normal umbilical cord was 22 cm with central attachment whereas the abnormal cord measured 16 cm and was thin.

DISCUSSION

Acardiac twin is a rare entity with unknown aetiology. Compression of the cephalic pole of the embryo inhibiting curving and fusion of the primitive heart tube is the basic cause resulting in non-formation of the heart and other organs. However, whether this TRAP sequence is the cause or effect of acardia is still unknown. The presence of vascular anastomosis on the surface of the placenta is a common feature of acardiac twins. The artery-to-artery anastomosis with perfusion in reverse direction leads to the arterial perfusion pressure of the donor twin more than that of the recipient twin, resulting in deoxygenated blood reaching the umbilical arteries of the recipient twin. This preferentially goes into the iliac vessels resulting in the development of only the lower extremities. Owing to the variation in the time of disruption of this developmental process, no two acardiac fetuses are alike. The presence of kidney as the only visceral organ in the abdominal cavity leaves an unanswered question that why only this organ has developed preferentially.

Acardiac twins are classified according to the degree of cephalic and truncal maldevelopment.² The first type is acephalus

with no cephalic structures. The second type is aniceps, where some cranial structures and/or neural tissues are present as seen in this case. The third type is acornus with a cephalic structure but no truncal structures are present. The fourth type is amorphous with no distinguishable cephalic or truncal structure. Development of the upper extremities varies.

TRAP sequence can result in a number of complications. Death of the acardiac twin either in utero or at the time of delivery is the rule. The normal twin is at a high risk of cardiac failure due to high cardiac output, hydrops and polyhydramnios. The perinatal mortality is about 50–70% without intervention.³ All these complications are strongly associated with the ratio of weight of the acardiac twin to the pump twin. If weight of the acardiac twin is more than 70% of the normal twin, preterm labour occurs in 90% as in this case, polyhydramnios in 40% and cardiac failure in 30% cases.⁴ If the ratio is less than 70%, preterm labour occurs in 75%, polyhydramnios in 30% and cardiac failure in 10% of these patients. In our case even though the weight ratio was more than 70%, there was no polyhydramnios or cardiac failure.

Early diagnosis of this condition may allow measures to be taken that may help in reducing complications. Diagnosis can be made by Doppler sonography as early as in the first trimester of pregnancy by detecting inversion of vascular flow in the recipient acardiac fetus.⁵ The second trimester scan displays the morphology of the fetuses well and allows a definitive diagnosis. Many cases may be managed conservatively by regular ultrasound follow-up to detect any signs of cardiac failure in the normal twin. But this involves risk of sudden fetal demise or devastating sequelae of premature delivery as was seen in the present case.

Management strategies may include ultrasonography-guided volume reduction amniocentesis and frequent interval monitoring studies. Invasive procedures, such as ligation of the cord of the acardiac twin or radiofrequency ablation may be opted under ultrasonographic guidance if the normal twin shows any signs of failure. Intrafetal ablation and fetoscopic laser coagulation have been tried by different authors with comparable success rates.^{1 6} Hecher *et al* reported a better success rate in early second trimester and less success in the third trimester because the umbilical cord is more oedematous with higher content of Wharton's jelly.⁷

Figure 4 Autopsy of the fetus showing serous fluid and fat, and one kidney (arrow).

Learning points

- ▶ Twin reversed arterial perfusion sequence is a rare entity and early diagnosis is possible by current investigative modalities.
- ▶ Acardiac twins are classified according to the degree of cephalic and truncal maldevelopment into four types. These are acephalus, aniceps, acornus and amorphous.
- ▶ Early antenatal diagnosis and close follow-up in these patients is required to prevent complication in the donor twin.
- ▶ Careful management strategies with minimal invasive therapy can decrease the morbidity and mortality of the donor twin.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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