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CASE REPORT

A Rare Case of Acardiac Twin

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ABSTRACT

This is a very rare condition happening on average once in every 35000 pregnancies. One twin is usually normal. The other is body like tissue often with malformed legs and lower body but no upper body or heart. Abnormal blood vessels on the placental surface allow the normal twin (pump twin) to pump blood through the tissues of abnormal twin. Because the pump twin heart has to pump for two, there is high risk of going into heart failure and death of normal twin. In our cases nature miraculously interrupted the blood circulation to acardiac twin which resulted in normal growth of normal twin. I present here this interesting case diagnosed and followed up and delivered at our institution.

Keywords: Twin pregnancy, Twin reversed arterial perfusion.

INTRODUCTION

The acardiac anomaly is a rare and bizarre phenomenon of monochorionic twin with incidence of one in 35000.[1] It results from large artery-to-artery and vein-to-vein anastomoses in their common placenta. In such cases, one twin becomes the 'pump twin' with its heart supplying blood both to itself and to the acardiac co-twin. The direction of blood flow in the co-twin is reversed with blood entering this twin through its umbilical artery and exiting through its umbilical vein.[2] This circulatory pattern leads to severe anomalies in the acardiac twin, most often with no formed heart (occasionally with an inadequate micro two-chambered heart), poor or absent development of the upper extremities and head.[3]

I am presenting this interesting case, which was incidentally detected during routine antenatal sonographic examination at 20 weeks pregnancy.

CASE REPORT

A 21-year-old female, married for 1½ years, visited the hospital on 31August 2009 with H/O 5 MA and pain in abdomen since two days. Last Menstrual period (LMP) of 17 March 2009.

Her clinical examination revealed 30 weeks size, tense distended uterus. The ultrasound on 31 August 2009 revealed the following:

- Monochorionic twin pregnancy with one live fetus in changing presentation of 22 and 23 weeks showing no apparent detectable major anomalies. Severe poly-hydramnios with AFI of 29.6 cm.
- Second acardiac and severely malformed fetus showing only portion of pelvis and malformed lower extremities with soft tissue swelling around. Absent upper body, heart and head.

The patient was referred to sonologist for a second opinion and Doppler study who confirmed diagnosis of acardiac anomaly as shown in Figures 1 and 2. Normal blood flow to normal fetus as shown in Figure 3. Reversed blood flow in umbilical vein of acardiac fetus[4] as shown in Figure 4.

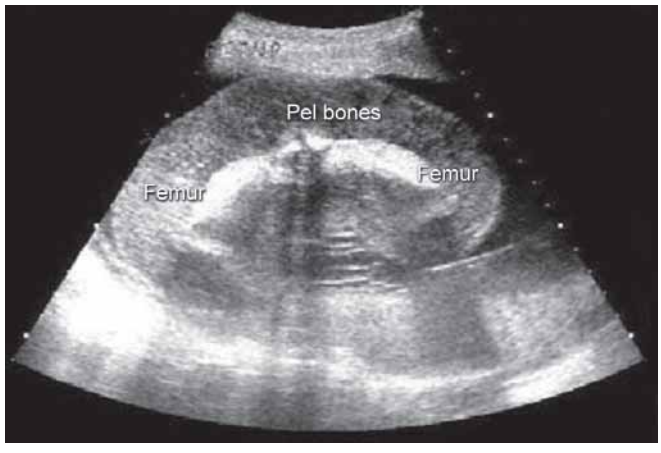

Fig. 1: Sonography showing pelvis of acardiac fetus

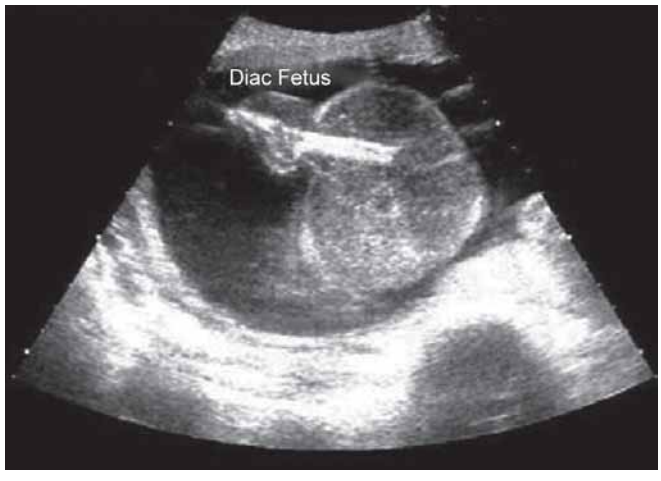

Fig. 2: Sonography showing malformed lower limbs of acardiac fetus

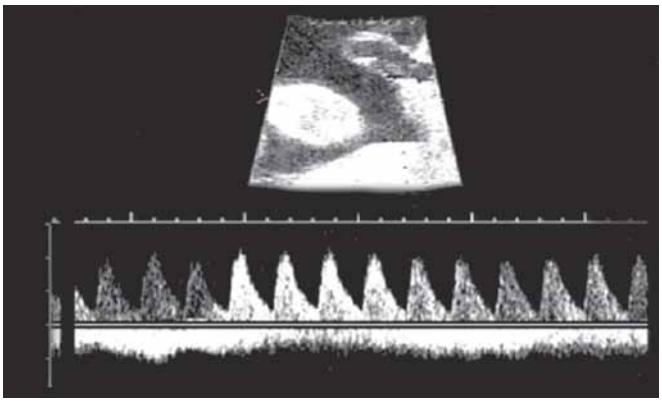

Fig. 3: Doppler showing normal blood flow to normal fetus

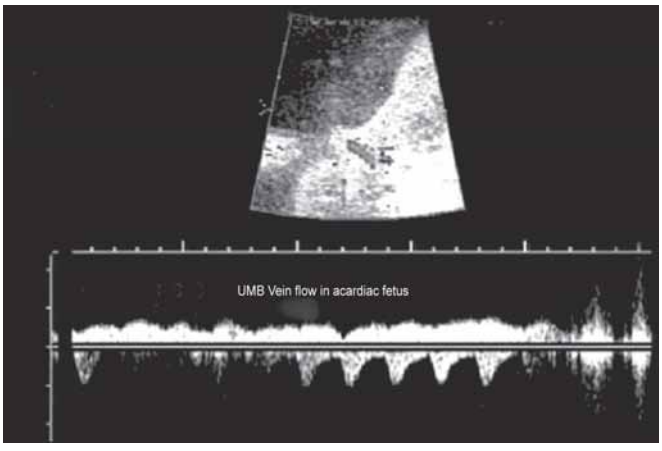

Fig. 4: Doppler showing reversed blood flow in umbilical vein of acardiac fetus

The patient and her relatives were counseled about this rare twin pregnancy and were advised to go to higher center for umbilical cord embolization or radiofrequency laser ablation.[5]

Due to their family's financial constraints they decided against it and opted to continue pregnancy as it is. In view of severe polyhydramnios and short cervix, cervical circlage was done and the patient was put on restricted activity, uterine relaxant and intravaginal micronized progesterone. She was followed-up at regular intervals.

Her examination on 3 November 2009 revealed corresponding fundal height of 32 weeks with breech presentation and soft nontender abdomen. Sonography showed twin pregnancy with one normal fetus of 32 weeks in breech presentation, no signs of hydrops and other acardiac. Surprisingly, her hydramnios had disappeared with AFI of 10.5 cm. Doppler was normal in normal fetus and showed no flow in umbilical vein of acardiac fetus. She was followed-up regularly till 36 to 37 weeks receiving two pints of blood as her Hb dropped to 7.7 gm.

In view of her persistent breech presentation, she was counseled for elective lower segment cesarian section (LSCS). Elective LSCS was done on 2 December 2009, primi with breech. First full-term male child weighing 2.3 kg was delivered with breech presentation, cried immediately and handed over to pediatrician. Then acardiac fetus weighing 300 gm was delivered. Placenta and membranes delivered completely. Placental weight 600 gm, showing two cord insertions as shown in Figures 5 and 6.

Fig. 5: Normal full term male child along with acardiac foetus

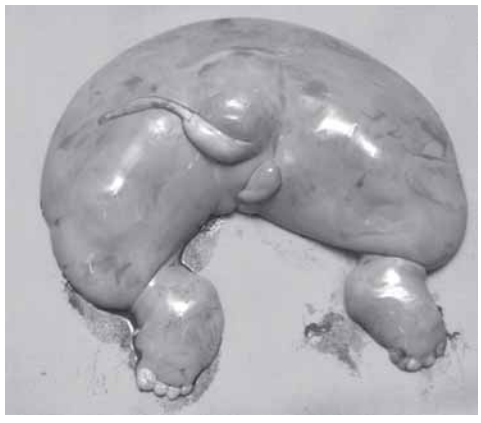

Fig. 6: Acardiac fetus with malformed lower body and limbs, absent upper body and heart

X-ray of acardiac fetus showed absence of bony cranium. Both upper limbs, rudimentary iliac blades and bony pelvis, and malformed lower limbs as seen in Figure 7.

The patient's intraoperative and postoperative period was uneventful. The mother and the baby were discharged on the 5th day.

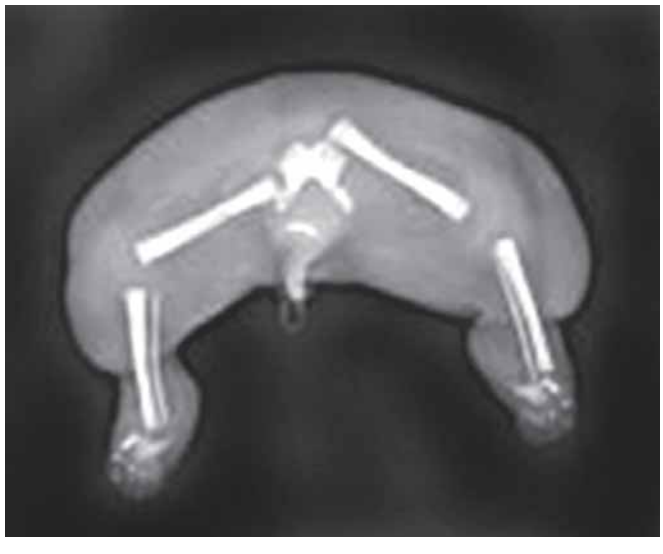

Fig. 7: X-ray of acardiac fetus showing rudimentary iliac blades and bony pelvis

Placental autopsy to study artery-to-artery and vein-to-vein anastomoses were not done due to absence of facilities in Jalgaon.

DISCUSSION

Acardiac twining is considered to occur as a sequel of the TRAP sequence in monochorionic twin pregnancy. The acardiac twin is perfused by the normal co-twin (referred to as the pump or donor twin) by means of reversal of circulation through large vein-to-vein anastomosis within the placenta. The acardiac/ recipient twin, therefore, has no direct vascular communication of its own with the placenta.

This reversed circulation in the anomalous twin alters the homodynamic forces needed for normal cardiac morphogenesis resulting in acardia.

PROGNOSIS

Overall perinatal mortality is 55% for the donor twin.[6] This is directly related to weight of the donor and recipient twins. Higher the weight of the recipient twin, higher is the likelihood of polyhydramnios, cardiac insufficiency in donor twin and premature delivery.

Moore's ratio or twin weight ratio is defined as weight of the acardiac twin expressed as percentage of pump twin weight. When this ratio exceeds 70%, preterm labor occur in 90%, polyhydramnios in 40% and congestive heart failure in 30%. With acardiac twin weight less than 70%, these rates are preterm delivery in 75%, polyhydramnios in 30% and CCF in 10%.[6]

In our case, since nature showed its own way of miraculous working, we were lucky to give the patient one normal full-term live child.

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