10.5005/jp-journals-10016-1069

CASE REPORT

Acardiac Acephalus with Single Umbilical Artery in Acardiac Twin

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ABSTRACT

Twin reversed arterial perfusion (TRAP) sequence (acardiac twin) is one of the rare complications of multifetal gestation which is unique to monochorionic placentation. It affects about 1% of multifetal gestation and prevalence is one in 35,000 pregnancies. We are presenting a case of acardiac acephalus with ventricular hypertrophy and polyhydramnios of pump twin. Autopsy of acardiac twin was done and various anomalies associated with it were described. Acardiac twin showed anomalies consistent with VACTERL anomalies and single umbilical artery. This case is reported because of the rarity of presentation and to stress the importance of early diagnosis for the proper management of the case. This is taken as an opportunity to describe various modalities of treatment of acardiac twin.

Keywords: Acardiac acephalus, Monochorionic placentation, TRAP sequence, Multifetal gestation and anomalies.

How to cite this article: Muralidhar L, Govindraj SK, Venkatesh S, Thimmaiah R. Acardiac Acephalus with Single Umbilical Artery in Acardiac Twin. Int J Infertility Fetal Med 2013; 4(3):93-95.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Twin reversed arterial perfusion (TRAP) sequence (acardiac twin) is one of the rare complications of multifetal gestation which is unique to monochorionic placentation. It affects about 1% of multifetal gestation and prevalence is one in 35,000 pregnancies.[1] Though it was first described in 1533, the number of reported cases is very few. In TRAP sequence, there is either arterial to arterial or veno-venous anatomises between the pump twin and the recipient twin.[2] we are presenting a case of acardiac acephalus with ventricular hypertrophy and polyhydramnios of pump twin. Autopsy of acardiac twin was done, and various anomalies associated with it were described. Acardiac twin showed anomalies consistent with VACTERL anomalies and single umbilical artery. This case is reported because of the rarity of presentation and to stress the importance of early diagnosis for the proper management of the case.

Date of Received: 20-07-13
Date of Acceptance: 22-08-13
Date of Publication: September 2013

CASE REPORT

A 28-year-old primigravida with 24 weeks of period of gestation was referred from periphery in latent labor with ultrasonography showing monochorionic monoamniotic twin gestation, with first twin of around 24 weeks without any anomalies and massive hydramnios and 2nd twin acardiac acephalus and massive hydramnios.

On history, there were no significant findings. On examination, abdomen was over distended corresponding to 36 weeks of gestation, tense, fetal parts couldn't be made out distinctly and fetal heart rate was not localized. On pervaginal examination, cervix was 75% effaced, 4 cm dilated. Bag of membranes bulging. Repeat ultrasonography and Doppler was done which showed confirmed the findings of old scan with ventricular hypertrophy of pump twin.

Patient delivered an alive female baby of weight 450 gm and acardiac acephalus of around 200 gm delivered with monochorionic placenta arterioarterial anastomosis (Fig. 1). Normal twin shifted to neonatal ICU, and acardiac twin and placenta sent for autopsy in academic interest after taking the consent from mother and her relatives. Normal twin on gross examination was normal and no congenital anomalies. Acardiac twin showed absence of the structures in the upper half of the body, i.e. cranium, thoracic structures that is lungs and heart and upper limb. Prior to sending the specimen for autopsy, X-ray was taken which showed normal bone development in the lower half of the body and distal bone defects.

Acardiac twin was subjected to complete autopsy dissection. The following are the anomalies reported with

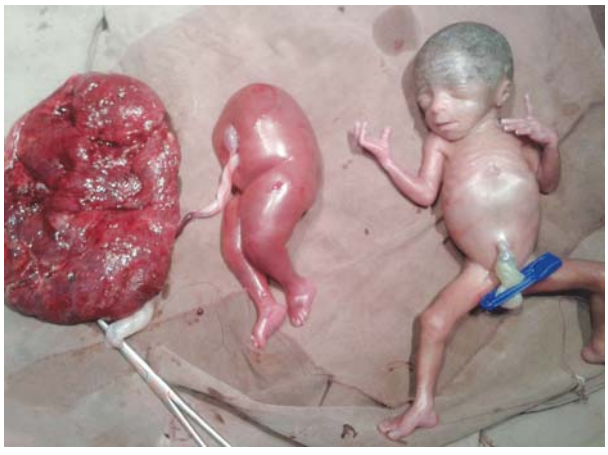

Fig. 1: The placenta, acardiac twin and normal fetus

it, i.e. single umbilical artery, limb anomalies in the region of toes, imperforate anus, improper development of external genitalia, equinovarus of the lower extremities, only small intestinal structures were developed, large intestine was absent, spleen and stomach were absent. Diaphragm was absent. Liver was developed. Internal genital organs were not developed. Small intestine which was present has blind ends on both the sides and was present within the umbilical cord (Fig. 2).

On complete study of placenta, no communication was found within the placenta but the communication was directly near the insertion of umbilical cord of pumping twin between two arteries. Placenta was velamentous and showed marginal insertion of both the cord structures. Placenta was monochorionic and, on complete dissection of the placenta, there was no vascular communication between the umbilical cord of the recipient twin and the placenta (Fig. 3).

DISCUSSION

There are two theories put forward for the development of acardiac twin: one is genetic defect in the fetus sufficient to cause failure of cardiac development in the fetus. Proposed hypothesis for the cardiac anomaly is post-zygotic nondysjunction. Other theory proposed is development of anastomosis in between the two vessels of the umbilical cord. Arterio arterial anastomosis early in the first trimester leading to vascular disruption of one twin and development of TRAP sequence. Our case strongly supports this theory as the placental dissection showed communication between umbilical arteries of acardiac twin with normal twin at insertion.[3,4]

Acardiac twin is associated with single umbilical artery in 66% and chromosomal abnormalities in 33% of the cases.[5] Mortality for pump is about 50 to 75%. Early diagnosis is very essential for the proper management of pump twin.

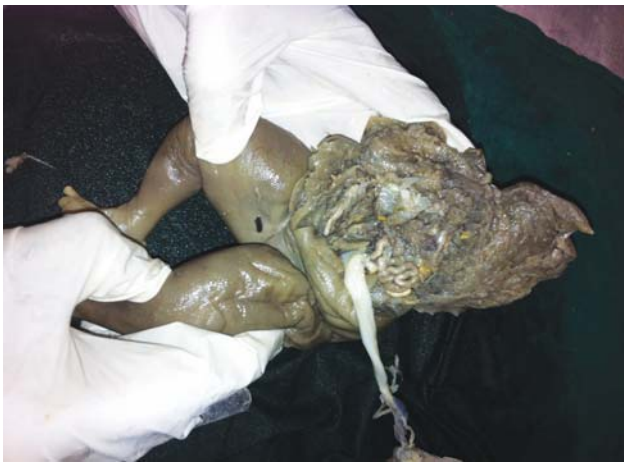

Fig. 2: The autopsy specimen of acardiac fetus

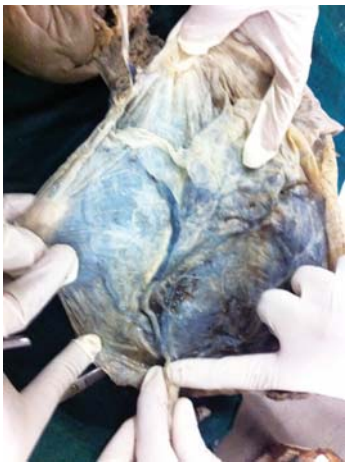

Fig. 3: The placenta with arterio-arterial anastomosis

Medical treatment has been tried with indomethacin and digoxin with limited success.[6] Conservative management is advised unless one of the following poor prognostic indicators is present. Unnecessary intervention will only increase the risk of mortality of the pump twin. Poor prognostic factors are development of: (1) polyhydramnios, (2) ultrasound indicators of cardiac insufficiency, like tricuspid regurgitation, pulsatile umbilical vein and abnormal pulsatile waveform in pulsatile twin, (3) large acardiac twins and rapid growth of acardiac twin.[7]

Various modalities have been described in the management of acardiac twin, earlier methods, like fetoscopic ligation of cord of acardiac twin and fetoscopic laser coagulation. The disadvantages of this methods are they are more invasive, frequently may require an additional port leading septotomy and iatrogenic pseudo amnionicity leading to increased chances of cord entanglement, the main disadvantage is technical difficulty in visualising the umbilical cord of acardiac twin. Perinatal mortality associated with this technique is 30% and preterm delivery is 70%. Newer treatment modalities include ultrasound-guided intrafetal approach. It includes intrafetal vascular ablation. The methods include monopolar coagulation, laser coagulation and radiofrequency ablation. Intrafetal approach simple, safe and effective. They are less invasive and have high rate of clinical success. Doppler clearly identifies the main intrafetal branch making it easier to access. Therefore, it is the preferred modality of treatment.[7]

Our case is presented because of the rarity of the case and to stress the importance of early diagnosis for the proper management of the pump twin.

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