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.

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 anastomoses 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.
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.

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.

REFERENCES


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