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Case Report

Acardiac twin: a commonly missed diagnosis

Neelamma Patil[1], Megha D Hittinhalli[1,*]

[1] *Dept. of Obstetrics and Gynecology, Shri B M Patil Medical college Hospital and Research Centre, BLDE University, Vijayapur, Karnataka, India*

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ABSTRACT

Twin reversed arterial perfusion syndrome is a rare condition seen in monochorionic twin gestation. The incidence is 1 in 35000 births and 1 in 100 monozygotic twin pregnancies. It is a complication of twin pregnancy that occurs due to preferential blood flow within the vascular communication between the two fetuses. We hereby report a case of 24-year-old primigravida referred in view of twin pregnancy with demise of one twin. On serial ultrasound it was misinterpreted as missed abortion of one twin and twin papyruses in further scans. But the consistent growth of the demised fetus made us think of acardiac twin as a differential diagnosis. When she presented in labour to our hospital, she delivered an acardiac anceps fetus along with a normal fetus.

1. Introduction

Acardiac twin constitutes a group of most bizarre malformations in human conception. Being a rare complication of monochorionic twin gestation, TRAP sequence accounts for approximately 1% of such cases i.e. 1 in 35000 pregnancies.[1,2] One twin with absent or nonfunctioning heart (acardiac twin) is perfused by its co twin (pump twin) via placental anastomosis resulting in poorly developed heart, upper body and a risk of heart failure in the pump twin.[2] It is commonly misdiagnosed as fetal demise because of absence of cardiac activity.

2. Case Report

A 24year old primigravida was referred to our hospital at 38 weeks of gestation in view of twin gestation with fetal demise of one of the twins. She was regularly booked and immunized. It was a monochorionic diamniotic twin gestation diagnosed at 10 weeks ultrasound with fetal demise of 2^{nd} twin. Repeat ultrasound further at 19 weeks of gestation gave the diagnosis as fetus papyruses of the 2^{nd} twin. Next ultrasound done at third trimester at 33 weeks gave the diagnosis of live fetus of 1^{st} twin and 2^{nd} twin at 18-19 weeks with fetal demise. Though all the scans had mentioned fetal demise of 2^{nd} twin, there was still slow, consistent growth of the " demised fetus ", which clinched the diagnosis of ACARDIAC TWIN. No doppler study was done. There was no hydrops or polyhydramnios in the surviving first twin.

The condition was diagnosed, and prognosis explained to the family. She underwent a vaginal delivery of the normal twin, alive male baby of weight 3 kg and an acardiac twin of weight 700gm. I t was Acarida Anceps type. The acardiac twin was a round mass of soft tissue with few hairs at one end and a small limb-like str ucture at the other end covered with thick, edematous skin and autopsy confirmed "the absence of heart ". Placen ta was monochorionic diamniotic with small thin cord attached to the acardiac twin.

3. Discussion

TRAP sequence is seen in monochorionic twin gestation. The Acardiac twin is dependent on the pump twin for its survival. The presence of large arterial anastomosis within the placenta helps in its development.[1]

* *Corresponding author.*
E-mail address: megha.anirudh@gmail.com (M. D. Hittinhalli).

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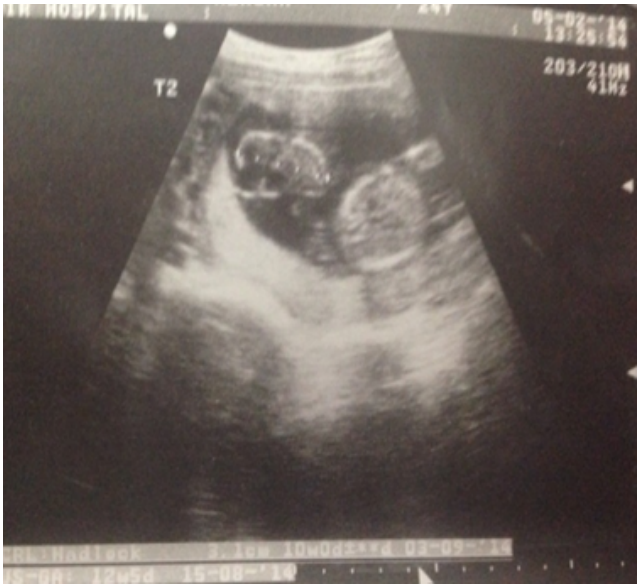

Fig. 1: Early scan with fetal demise (10^{TH} Week)

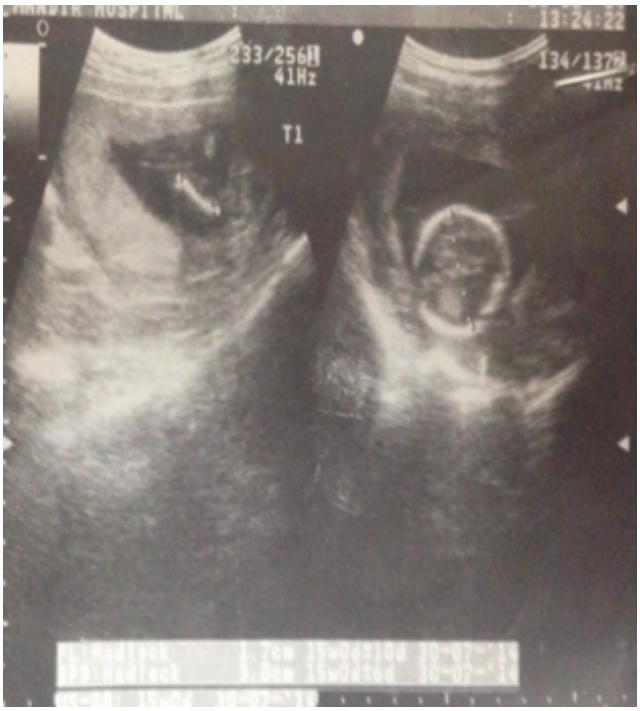

Fig. 2: 19 Week scan- fetus papyraceous

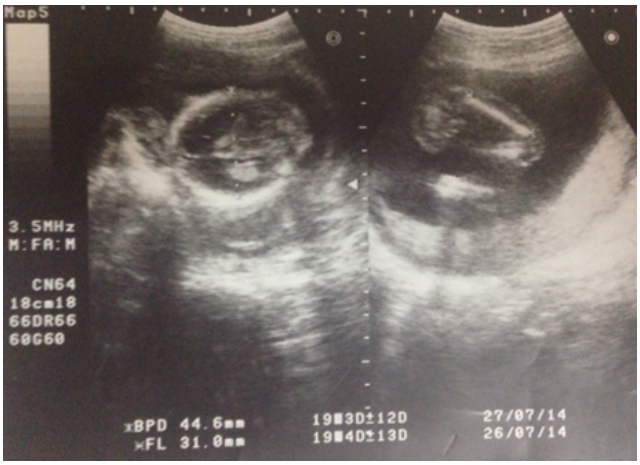

Fig. 3: 3^{RD} trimester scan, 2^{ND} twin IUD

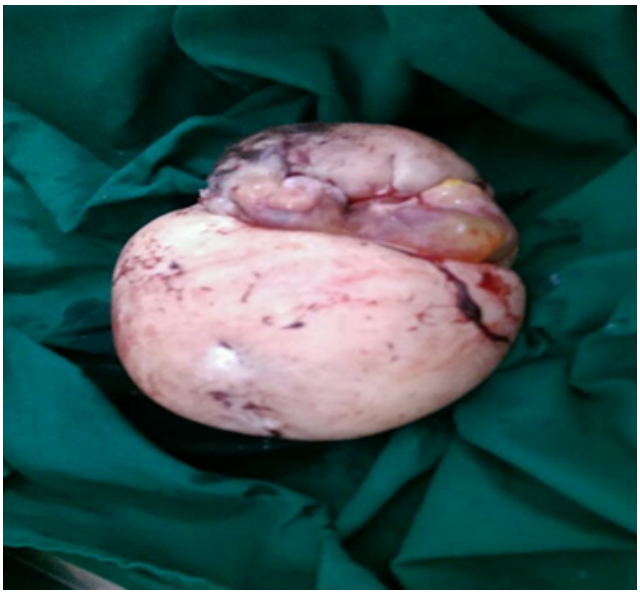

Fig. 4: Anterio aspect of acardiac anceps

Prenatal diagnosis by ultrasound is suspected when there is discordancy with retrograde flow in acardiac twin seen by color flow doppler. The differential diagnosis would be anencephaly, cystic hygroma, conjoined twin, twin demise, intraamniotic placental tumors.[3] Classic sonographic findings of the acardiac twin include absent cardiac activity, bizarre skeletal findings, and a large soft tissue mass with no limbs. Diagnosis of cardiac activity at 5-6 weeks gestation and their subsequent reversal of blood flow has been demonstrated with in the placental vascular anastomosis.[4,5] It is the pathognomic feature of this entity.

In the acardiac fetus blood circulation is provided by artery-artery and vein-vein anastomosis on the placental surface. It uses this deoxygenated blood from the cardiac twin and the normal tissues become atrophic.[6] It

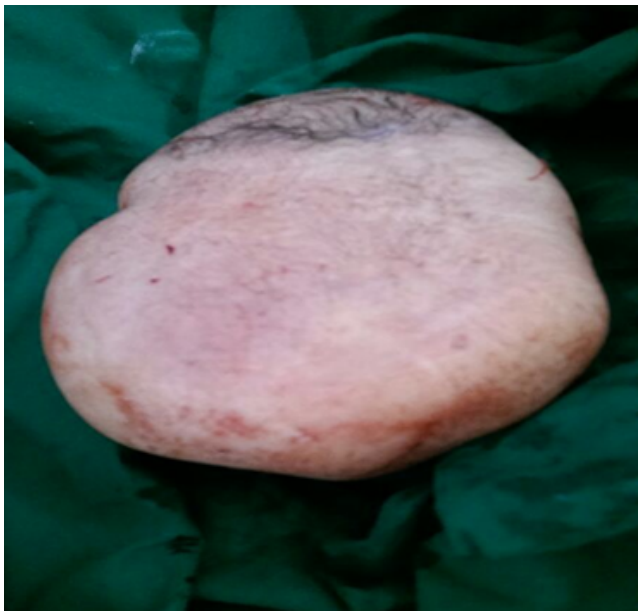

Fig. 5: Posterior aspect of acardiac anceps

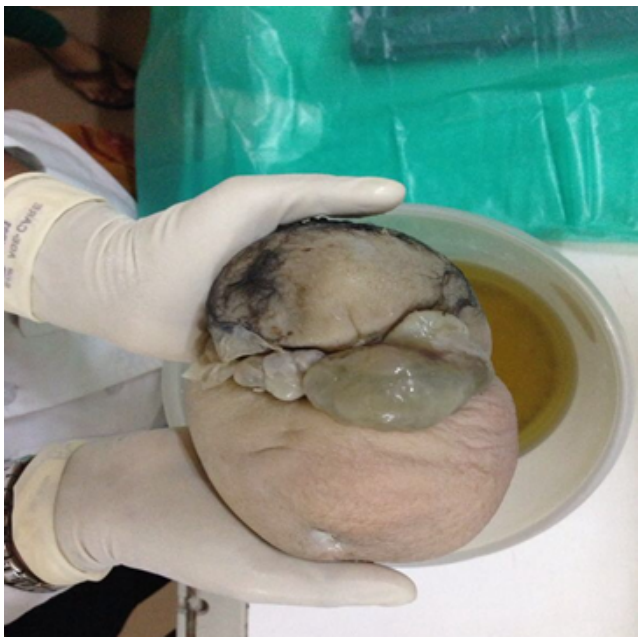

Fig. 6: Partialy developed upper part

preferentially perfuses the lower part of the body hence the entity known as " twin reversed arterial perfusion " which leads to maldevelopment or no development of the upper part.[1,6]

Acardiac fetus is divided into two types[6,7]

1. Pseudocardiac: presence of cardiac structures although rudimentary
2. Holocardia: absence of cardiac structure development.

It is again divided into 4 types:

- Acardiac acephalic
- Acardiac anceps
- Acardiac acormus
- Acardiac amorphous

In our case it was partially acardiac anceps type. Because of absence of cardiac activity, it was diagnosed as fetal demise at first trimester scan, but subsequent scans had shown growth of the demised twin. This clearly indicates that the cotwin must be acardiac twin. So previous USG reports must be considered which was suggesting a slow but consistent growth of the acardiac twin.

The prognosis in TRAP sequence is 100% for the acardiac twin. Mortality around 50% for the pump twin ; cause being cardiac failure. The risk of cardiac failure depends on the relative size of the acardiac twin in relation to its pump.[1] Cardiac failure develops because the pump twin must maintain two circulations. Eventually it ends in hydrops with polyhydramnios.[8] Chromosomal anomalies are reported to be around 10% and thus should be excluded before the management of TRAP sequence.[6] Sharan Athwal et al in the year 2010 in a case report on TRAP sequence demonstrated a case of TRAP sequence with association of VACTERL syndrome.[9] The surviving twin was diagnosed prenatally with VACTERL association i.e. vertebral defects (V), anal atresia (A), congenital heart lesions (C), tracheoesophageal defects (TE), renal and distal urinary tract anomalies (R) and limb lesions (L); any 3 of them. Another report was presented by W. Blaicher et al in 2000 demonstrated two cases of acardiac twin associated with trisomy 2. It was concluded that cytogenetic investigations should be carried out routinely to evaluate the impact of aneuploidy in the cardiac twin.[10]

Optimal management is controversial. Salvaging the pump twin should be the goal of the therapy. The best modality of treatment must be provided. In addition to that the optimal timing for treatment, the management options include expectant management, delivery, and intrauterine interventions should be determine. The technique of choice in early intervention is intrafetal laser coagulation and radiofrequency ablation done between 12-14 weeks of gestation.[1] Advance d technology such as Radiofrequency ablation procedure have a favorable outcome and support early intervention of TRAP sequence.[11] Further in second trimester they can undergo ligation of umbilical cord of acardiac twin, fetoscopic laser coagulation, bipolar diathermy of vessels in the umbilical cord. USG guided ablation of intrafetal vessels is a newer and popular method. Modalities in third trimester include expectant management with proper fetal monitoring, doppler flow pattern, betamethasone therapy if preterm and termination by cesarean section if term.

4. Conclusion

A high index of suspicion is required for accurate diagnosis and optimal management of acardiac twin. Acardiac twin should be thought as differential diagnosis whenever there is a twin pregnancy with fetal demise of one twin.[1] This case also stresses the importance of correlation of the previous ultrasound reports with the present ultrasound.

5. Source of funding

None.

6. Conflict of interest

None.

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Author biography

Neelamma Patil Associate Professor

Megha D Hittinhalli Post Graduate

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