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Case Report

Acardia twin-twin reversal arterial perfusion sequence

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ABSTRACT

Acardia twin is a rare, unique complication of monochorionic twin pregnancy. A 2.6% incidence among monochorionic twins has been cited. In such complications a twin with abnormal or non-functional heart is perfused by its co-twin via placental arterial anastomosis. The acardia twin has a poorly developed heart and upper torso. We present a case in which the fetal demise of one fetus of twin pregnancy has been diagnosed in the first trimester, but the patient wishes to continue the pregnancy. This resulted in the unexpected occurrence of an acardia twin after delivery along with one normal twin, placenta was single. A review is presented on therapeutic opportunities and necessity of thorough sonographic evaluation in monochorionic multifetal pregnancies.

Keywords: Acardia acephalus twin, TRAPS, Monochorionic twin

INTRODUCTION

It's double the giggles and double the grins and double the trouble if you are blessed with twins.¹ Twin reversed arterial perfusion is a rare condition of monochorionic twin pregnancies. It makes an appearance when the heart of one twin does the work of supplying blood for both twins. The twin supplying the blood is known as the "pump twin" and develops normally in the uterus. However, the increased pumping of the heart puts this twin at risk for cardiac failure. The other twin-known as the "acardiac twin"-lacks a heart or has one that is not fully formed. It usually has a poorly developed body and may also be missing a head, limbs and torso. It results due to abnormal artery to artery or venous to venous communication in the placenta. Reversal of blood flow in the umbilical artery of the recipient twin is characteristic, and deoxygenated blood is brought from the pump twin to the acardia twin. TRAP sequence has been reported to occur in about 1 percent of mono-chorionic twin pregnancies and 1 in 35000 pregnancies.² In present day obstetrics, the incidence appears to be much higher, when factors such as the use of first trimester obstetrics ultrasound examination, which detects twin demises early in gestation, and assisted reproductive techniques, which have increased the

incidence of twin including mono-chorionic twin, are held to be.³ Diagnosis of this complication was made with ultrasonography which is usually helpful from 11 weeks of gestation. TRAP may also occur in monochorionic triplet and higher-order multiple gestations.⁴ At present there is a controversy regarding elective versus therapeutic treatment of reversed arterial perfusion sequence.⁵ Here we are presenting a case of TRAP sequence, in which one twin was born acephalus and other was born normal.

CASE REPORT

A 27-year-old multigravida presented at second trimester of gestation in the outpatient department (OPD) of district hospital Chhatarpur for regular antenatal care (ANC) visit where she was advised ultrasonography (SG) due to discrepancy between gestational age according to date and height of fundus. Detailed USG revealed twin pregnancy with a surviving twin which was structurally normal with a fetal biometry of 23 weeks of gestation, and the other dead twin head was not seen with absent cardiac activity (Figure 1 A and B). Single placenta with a single pocket of amniotic fluid with no separating membrane was noted (Figure 1A).

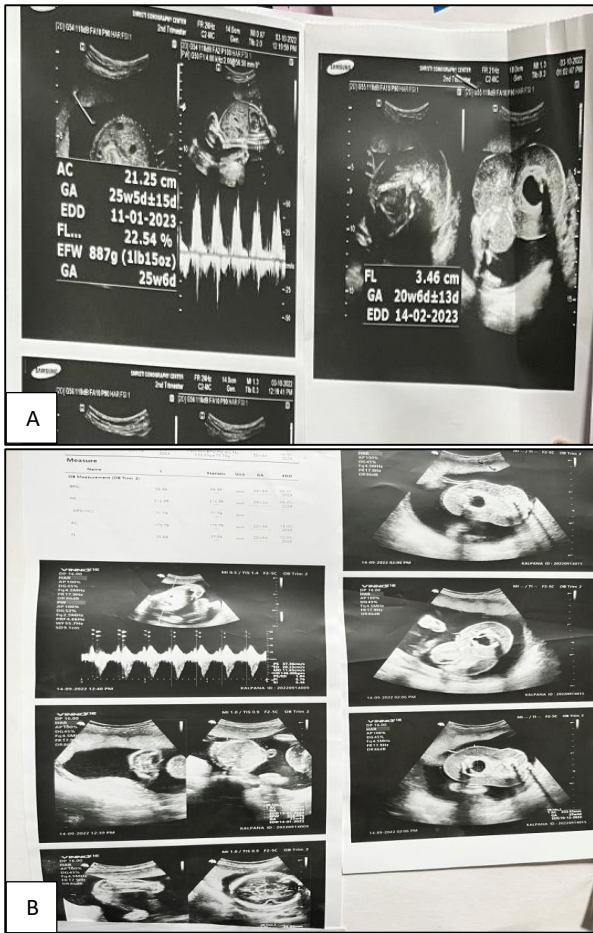


Figure 1 (A and B): Detailed USG revealing twin pregnancy with only one surviving twin and another twin with absent cardiac activity and no head.

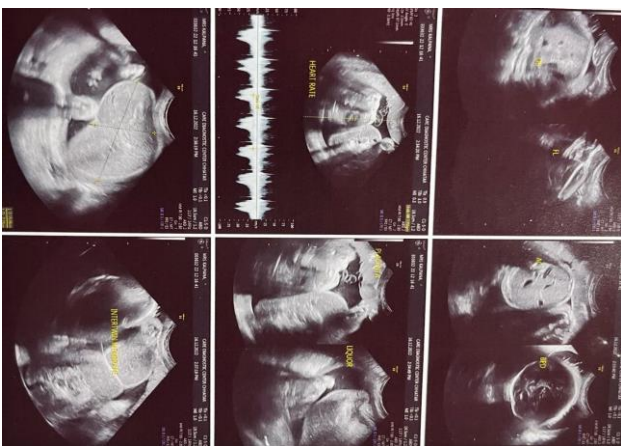


Figure 2: USG revealing one foetal demise.

On this basis, a diagnosis of twin pregnancy with one fetal demise was made. Patient wished to continue the pregnancy; her fibrin degradation product (FDP) and coagulation profile were checked which were within normal limits. Patient did not report back to OPD throughout the rest of her pregnancy. After three months she presented to emergency with lower abdominal pain and discharge pervaginum. One fetal heart sound (FHS)

was audible other not. Abdominal ultrasound scanning revealed one fetal demise (Figure 2). She was admitted and she had vaginal delivery. The first twin was female, weighing 2.7 kg alive and healthy with good cry. (Figure 3A) and second twin weighed 2.7 kg (Figure 3 B), upper portion contained soft globular mass, head and upper extremities were absent, external genitalia female type. Poorly developed, the lower spinal column and lower leg bones were apparently normal. Autopsy revealed absence of fetal heart. Fetus was classified as acardiac anencephalus structures. The placenta was single, monochorionic and weighted 500 (Figure 4). Postpartum period was uneventful, and she was discharged in three days.



Figure 3 (A and B): Twins. Surviving twin, female, 2.7 kg alive and healthy. Dead twin with absent upper torso.

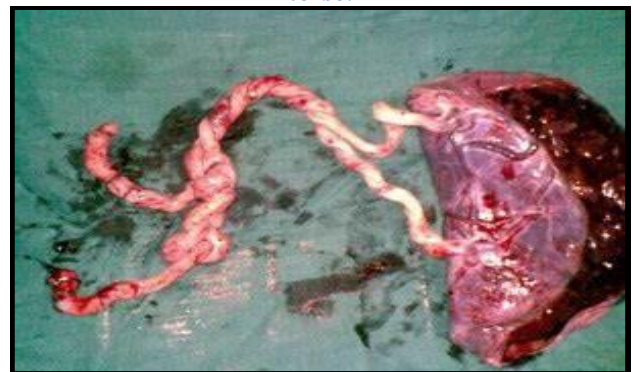


Figure 4: Monochorionic placenta.

DISCUSSION

First historical case of acardia were reported by Benedetti in 1533 and Benedictus in 1539, and later by Geoffroy in 1836. Prevalence of acardia is one in 35,000 deliveries. One of the variance criteria most widely accepted are morphological, which is, the presence or absence of head, body and cardiac tissue. Acardiac anomaly usually occurs in monozygotic twins, there are a few case reports of dizygotic twins with a fused placenta were also seen. It is more common in female twins, and because the disorder is monozygotic, twins share the same gender. The pathogenesis of this anomaly is abnormal placental vascular communication between the twins, leading to imbalance of interfetal circulation. Reversed blood flow in the umbilical artery of the acardiac twin causes atrophy of the heart and other organs.⁶

The mortality of the acardiac twin is 100%, and the perinatal mortality of the pump twin is reported to be around 50%.⁷ Since then, much advancement in ultrasound technology has been put into practice for detection of this anomaly. An acardiac twin should be suspected in all monochorionic, malformed fetuses with an absent cardiac pulsation with a non-functioning heart. An ultrasonography finding of twins revealing discordant or grotesque malformation along with reverse flow in the umbilical artery is usually diagnostic of an acardiac twin.⁸

CONCLUSION

This case shows that acardia can be diagnosed by means of prenatal ultrasound in front of a monochorionic twin pregnancy when one of the fetuses is deformed and has no cardiac activity. Continuation of pregnancy can be done after early diagnosis of acardia for viable foetuses with regular follow-up.

Recent studies and advancements in ultrasound technology have significantly contributed to enhancing the future of the field related to acardia twins and monochorionic twin pregnancies. The diagnosis of acardia twins can now be made in the early stages of pregnancy thorough sonographic evaluation. This allows healthcare professionals to identify cases where one twin lacks a functional heart or has a poorly developed body. The use of ultrasound helps in detecting twin demises early in gestation and provides essential information for making informed decisions regarding the continuation of the pregnancy.

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