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Undiagnosed Acardiac Anceps Twins: A Case Report

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Abstract

Acardiac twins refer to a developmental anomaly unique to monochorionic pregnancies. It is a very rare complication that manifest as twin reversed arterial perfusion (TRAP) sequence as a result of vascular anastomoses at the placenta bed. We present a case of an undiagnosed acardiac anceps, delivered at 31 weeks gestation. A 25-year-old unbooked multigravida presented in labour and had an emergency Caesarean section with a finding of a monochorionic diamniotic twin gestation. Twin I was an acardiac anceps weighing 3000g with ambiguous genitalia, the umbilical cord was short (25cm long) with two arteries and one vein. Twin II was a live female baby, weighing 1500g, APGAR scores were 6 and 8 at the first and fifth minutes respectively. She had features of hydrops fetalis and congestive cardiac failure with a significant patent ductus arteriosus (PDA). Twin II (pump twin) however died on day 21 from refractory heart failure. Antenatal diagnosis and appropriate intervention is crucial to good outcome.

Keywords

Acardiac anceps, Twin Reversed Arterial Perfusion (TRAP), Monochorionic, Hydrops foetalis.

I. Introduction

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Acardiac twinning is a dysmorphic anomaly unique to monochorionic pregnancies [1]. It is a very rare complication of multiple gestation that is associated with high perinatal mortality if not identified early and appropriately treated [1, 2]. The presence of an acardiac twin occurs in 1/ 35,000 twin pregnancies and in 1% of all monochorionic twin pregnancies [1-3] . Acardiac twin occurs only in monozygotic twins [1]. It is due to twin reversed arterial perfusion (TRAP) sequence which occurs in early embryogenesis as a result of vascular anastomoses at the placenta bed[3] . It is a spectrum of abnormality ranging from development of head, thorax and well formed limbs to an amorphous mass that has minimal semblance to human fetus [1-4] . Acardiac fetuses were first described by Benetti in 1533 [3,4]. Basically there are four morphological types *namely; acardiac acephalus* (most common type) here the fetus has no cephalic development, while *acardius anceps* has cranial structures with or without neural development. The *acardius acormus* has limited cephalic structures with no truncal development, while the *acardic amorphous* has the most severe malformation consisting of an indistinct mass with absent of cephalic and truncal differentiation. Based on the degree of cardiac development , acardiac fetus can be classified into two: hemiacardius when heart is incompletely formed and holo-acardius when heart is absent [6]. The pump twin suffers congestive cardiac failure and hydropic changes due to extra demand of pumping blood to the acardiac twin which is completely an obligate parasite. Mortality for pump twin is 50-70% while that of the acardiac twin is 100% [4]. However, early diagnosis, specialist follow-up and timely treatment will improve the survival rate of the pump twin [1-3,5]. This report emphasizes the importance of early and adequate antenatal ultrasonography in multiple gestation pregnancies. Antenatal scans will establish chorionicity, and identify TRAP and other Twin-Twin transfusion syndromes with a view to improving the fetal outcome.

II. Material and Methods

Study setting

An unbooked 25 year old gravida four para two plus one (G $_4P_2^+$ ^{1}) with two living children was referred from a private hospital on account of drainage of liquor with twin gestation and intra uterine fetal death (IUFD) of the leading twin at 31+4 weeks gestation. She had been draining liquor for 10 hours, the liquor was dark brown, with no associated fever or vaginal bleeding.

The pregnancy was spontaneously conceived and had been uneventful until the above complaints. No history of use of herbal medication.

She had two uneventful spontaneous vaginal deliveries and one spontaneous complete miscarriage at 13 weeks in the past. She had no previous surgery and was neither diabetic nor hypertensive.

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Her clinical examination revealed a young woman in intermittent painful distress, not pale, axillary temperature of 36.5 Celsius with bilateral oedema up to the mid leg. Her pulse rate was 88 beats/minutes full volume and regular and blood pressure was 120/80 mmHg. Her abdomen was uniformly enlarged with no area of undue tenderness. Symphysio- fundal height was 40 centimeters (large for date) having two moderate uterine contractions in 10 minutes lasting 35 seconds each. Multiple fetal parts felt with leading twin in transverse lie. Single fetal heart was heard and regular -150 beats /minutes. Vaginal examination revealed vulva pad in-situ moderately stained with chocolate fluid, normal vulva, vaginal smeared with chocolate color fluid, no cord prolapse. The cervix was fully effaced, 5 centimeters dilated and membrane not felt. She was given a dose of dexamethasone (12mg). She had an urgent bed side Obstetric Ultrasound which showed viable twin II in transverse lie with normal cardiac and fetal activity at average gestational age of 31 weeks adequate liquor volume and estimated fetal weight (EFW) of 1500g, single pocket of amniotic fluid 1.5x 1.0 cm. Twin I was in transverse lie showing a complex irregular fetus with poorly formed head and indistinct trunk, no upper limb, no obvious cardiac activity, no pocket of liquor. Both shared a common placenta located posterior-fundal region and not low lying. She was counseled on her condition and need for an emergency caesarean section which she consented to. The neonatologist was informed of TRAP sequence. Her routine blood tests including virology were normal.

Intraoperative findings were twin I an acardiac-anceps , having poorly formed head subsumed into a poorly formed trunk , no upper limb, with relatively poorly formed lower limbs having 3 toes each and an ambiguous genitalia with birth weight of 3000g. It had a very short and thin umbilical cord measuring about 25 centimeters with two arteries and a vein. Twin II, a live female neonate, markedly pale, in respiratory distress, with hepatosplenomegally, weighing 1500g, with APGAR Score of 6 and 8 at the first and fifth minute, respectively. A single placenta weighing 900g. The tubes and ovaries bilaterally were normal, estimated blood loss was 800 millilitres.

The surviving pump twin had features of hydrops fetalis, with haemoglobin of 24mg/dl, which required three packed red cell transfusions. She also had a significant PDA. She had 21 days of neonatal intensive care but died of refractory cardiac failure.

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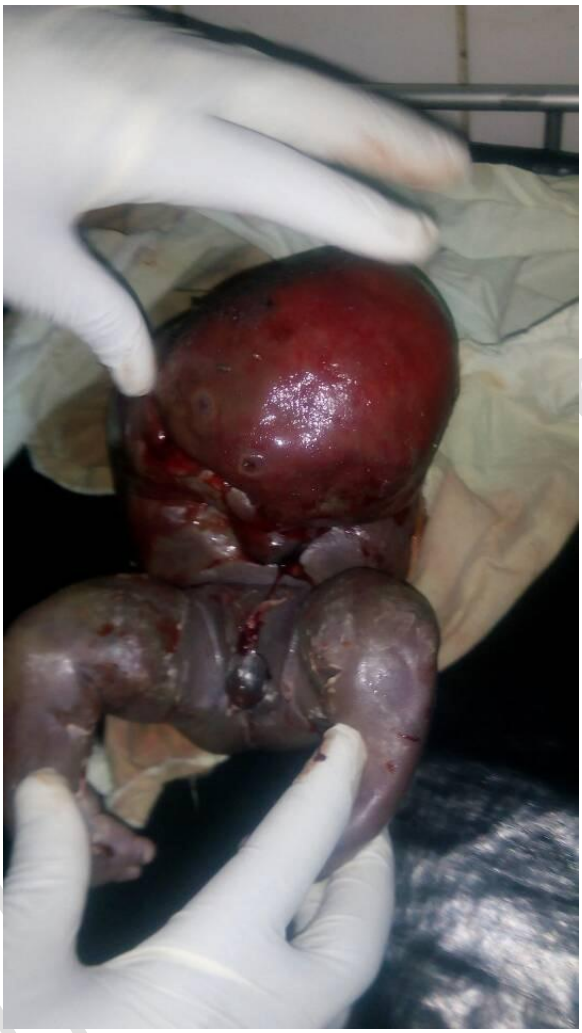

FIGURE 1 ACARDIAC ANCEPS (TWIN I)

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FIGURE II LIVE FEMALE NEONATE (Twin II in NICU)

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III. Discussion

Acardiac twins, characterized by twin reverse arterial perfusion (TRAP) sequence is a rare complication unique to monochorionic twin gestation [1,2] . It is associated with a very high perinatal morbidity and mortality. This case clearly indicates the need for appropriate early diagnosis, treatment and need for regular ultrasound follow up with Doppler studies to promptly detect imminent fetal jeopardy and timely delivery. She was an unbooked patient who presented with preterm labour due to preterm premature rupture of membrane. The pumped twin was alive birth delivered with severe anaemia and obvious heart failure whom despite proactive and neonatal intensive care still died. The etiopathagenesis of acardiac twin results from an abnormal vascular communication in early embrogenesis leading to imbalance of interfetal circulation [1,5]. Usually there are large arterial-arterial placenta shunt and consist of a normally formed 'donor' or "pump'' twin with high arterial perfusion pressure and a recipient twin completely lacking a heart[2]. The recipient twin therefore receive deoxygenated blood via the umbilical arteries in a reverse pattern thus preferentially supply the lower body by the illac vessels, its mortality is 100% [1,7]. The donor twin on the other hand is at a higher risk due to the extra demand on the heart from the recipient twin; this put her at an increased risk of mortality of about 50-70% [6].

The diagnosis of acardiac twins is quite challenging. Early second trimester ultrasonography (USS) for twin gestation is very important to identify the chorionicity and amnionicity. If detected a monochorionic twins, effort should be made to confirm good fetal viability. An acardiac twin should be suspected in all monochorionic, malformed fetuses with cystic hygroma, generalized oedema and absent of cardiac pulsation [9]. In subsequent USS follow up, finding of growth discordant, monstrous nature of either twin along with reverse of flow in the umbilical artery should draw a high index of suspicion for TRAP sequence with acardiac twins [9].

Having made a diagnosisof acardiac twins with TRAP sequence, management can be very challenging, however several options have been used with variable successes. Among which are maternal digoxin and indomethacin therapy, this can help salvage the pump twin by preventing heart failure and polyhydramnios respectively [9]. After excluding gross fetal anomaly of the pump twin, regular USS follow up can be done till obvious sign of fetal compromise from Doppler studies. This conservative approach may be of valuable importance especially in resource poor counties with limited specialist and technology for active radical treatment [10]. It is very important to take note of the fetal growth, if marked discordant growth is observed then it is a poor prognostic sign. This is quite challenging to compare as it is difficult to estimate the weight of the acardiac twin. It been suggested that an approximate estimate can be done by comparing the ratio of the abdominal perimeter of the acardiac and pump twins, or by using the starndard prolate

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ellipsoid formula(ie. {LENGTH OF ACARDIAC}X { ABDOMINAL DIAMETER} divided by 2) [9].The weight of the acardiac twins is very important as a prognostic index. It has been documented that congestive heart failure developed in the pump twin in 100% of cases if the acardiac twin / pump twin twin ratio is greater than 70% at birth and when the weight of the acardiac twin was less than 25% compared to the pump twin, the prognosis is better [1,3,9,10]. This invariably shows a worse prognosis attributable to preterm delivery (90%), Polyhydramnios (40%), thus a perinatal mortality rate of 50-75%. This can account for the poor outcome in this index case despite proactive intensive neonatal care. Birth weight of the pump twins was 1.5 kg while that of the acardiac anceps was 3.0 kg.

Other treatment modalities are invasive or minimally invasive targeted at the vascular communications at the placenta bed usually by stopping the blood flow to the acardiac twin. Invasive once include hysterotomy to remove the anomalous fetus it is associated with high maternal and perinatal morbidity and mortality as such no longer done [1]. Most favoured are minimally invasive procedures includes embolization of the acardiac circulation with alcoholic substances, fetoscopic cord ligation and ultrasound guided coagulation of the umbilical cord or laser or radiofrequency ablation. Percutaneous ultrasound guided intra-fetal alcohol injection is less invasive compared to endoscopic procedure and may suffice in our environment. If early diagnosis is made with gross anomaly and poorer prognostic indexes, a termination of pregnancy can be done [3].
The management of acardiac twins with TRAP sequence is quite challenging, however the outcome is better with early diagnosis and adequate intervention [1-3].

Conclusion: An Acardiac twin is a very rare complication of monochorionic twin pregnancy that is associated with high perinatal morbidity and mortality. Increased index of suspicion with early USS for twin pregnancy especially monochorionic, may help in early diagnosis which will enhance close fetal surveillance and specialist care. Conservative treatment still remains a major tool for the poor resource countries.

AUTHOR DISCLOSURE STATEMENT

No competing financial interests exist.

All the authors participates fully in case diagnosis and treatment

IV. References

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