Case Report

TWIN REVERSED ARTERIAL PERFUSION SEQUENCE / SEQUENCE ACEPHALUS ACARDIAC FETUS

Shahida Perveen[1], Huma Quddusi[2], Shakeel Ahmed Faiz[3]

ABSTRACT

Twin reverse arterial perfusion (TRAP) sequence occurs only in a setting of a monochorionic gestation and complicates approximately 1% of monochorionic twin gestation. In the TRAP sequence the acardiac / acephalic twin receives all of its blood supply from the normal "pump" twin.

KEY WORDS: Twin reversed arterial perfusion, Sequence acephalus acardiac fetus.

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INTRODUCTION

The term twin reversed arterial perfusion syndrome (TRAP) was first defined by Gruenwald in 1942, is probably responsible for acardiac foetus as one of the forms of twin to twin transfusion syndrome (TTTS).[1] It is a rare complication of monochorionic twinning. The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the normal pump twin.

CASE REPORT

A 25 years old primigravida was referred to Nishtar Medical College Hospital, Multan at 34 weeks of gestation with ultrasound report performed at a periphery hospital which revealed a twin pregnancy. One of the fetuses was reported dead. She was admitted in obstetrics and gynaecology unit-II Nishtar Medical College Hospital. Her marriage was non-consanguineous and she had no history of infection or drug intake during early pregnancy.

Patient was a house wife and belonged to poor socioeconomic class. She conceived spontaneously and was an un booked case. Her family history was not significant. Her clinical examination revealed uterus size of 36 weeks of gestation with multiple fetal parts. Routine investigations were done which showed Hb% 13.5gms/dl, Blood group B^+, Rubella antibodies negative, syphilis HBs Ag and HBc Ag –ve, Random Blood Sugar 80mg/dl, urea 52mg/dl, creatinine 0.8mg/dl, na 139, k 4.3, bilirubin 0.5, SGOT 24, SGPT 23, Alkaline Phosphate 393. Ultrasonographic evaluation was repeated which revealed that Twin A was found with vertex presentation, normal fetal anatomy and with normal liquor volume. No evidence of ascites, pleural effusion, skin oedema or polyhydraminos were noted. Grossly abnormal anatomy was noted in Twin B. There was no definition of fetal skull above the thorax and spine ended abruptly in cervical region as shown in Fig-1. Ultrasound findings were consistent with TRAP.

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Patient was already diagnosed to be a case of pregnancy induced hypertension and was taking Aldomet 500mg orally TDS. Patient remained admit in the ward for two days. She was given Betnesol injection 12mg intramuscular 24 hourly two doses. On the third day her blood pressure shoot up to 160/120 mm Hg and she was shifted to labour ward for her control of blood pressure and possible termination of pregnancy. In the labour ward Nitrosine infusion (10ud/min) and Tab Adalet retard 20mg BD was started. Patient was counseled about the whole situation and after stabilizing her blood pressure Caesarean Section was performed under general anesthesia.

The first twin was female with weight 2 kg without any external malformation as shown in Fig-2. The second twin was also female malformed stillborn weighing 2.5kg. The lower trunk was normal. Both feet showed equinovarus deformity and in right foot the middle toe was misplaced. Proximal part of the foetus was a multilocular cyst filled with transparent fluid, covered with gelatinous swollen skin. There was no upper limb or head and neck. A defect was found around the root of the umbilical cord which indicated umbilical hernia. It had a well developed female external genitalia, the urethral meatus was absent while the anal opening was present. Patient was transferred to ward in a satisfactory condition and was discharged from hospital on 5th postpartum day with a healthy female baby. Patient was given appointment for six weeks in postnatal clinic.

DISCUSSION

Twinning is the most common type of multiple gestation. Twin reverse arterial perfusion sequence occurs in setting of monochorionic gestation. The incidence of monozygotic twining is 3-5/1000 birth with a recurrence risk of 1%.[2] All monochorionic twins are at high risk for complications including twin to twin transfusion syndrome (TTTS), fetal growth restriction, fetal death and premature delivery. The first delineation of twin to twin transfusion syndrome was by a German obstetrician, Friedrich Schatz.[3] He described the placental "Third circulation" in 1875 and fully explored this concept in 1886.

Twin reverse arterial perfusion syndrome (TRAP) was first defined by Gruenwald in 1942, is probably responsible for acardiac foetus as one of the forms of TTT.[1] It is a rare complication of monochorionic twinning. It is caused by arterio-arterial and veno-venous placental anastomoses leading to circulatory problems in one twin. The acardiac twin is usually grossly abnormal with severe reduction anomalies of the upper part of the body as seen in our case. The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the pump twin.

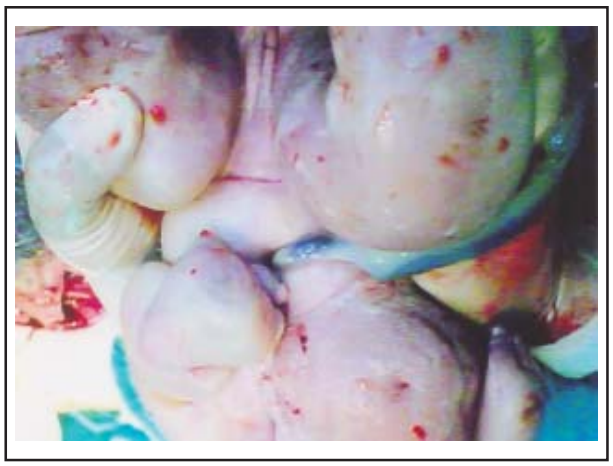

Figure-1: Acaridae Twin

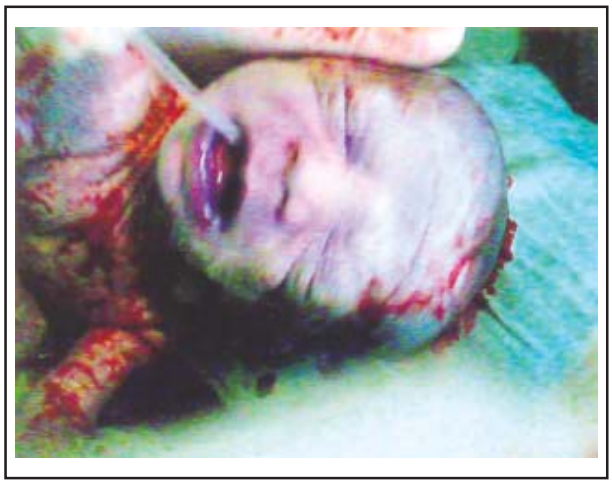

Figure-2: Normal Twin

Deoxygenated, low-pressure blood from the pump twin, which would normally return to the placenta, instead flows directly to the acardiac twin, resulting in a wide array of structural abnormalities. Mortality of the pump twin is 50-75% usually due to the result of heart failure and of the acardiac is 100%.[4]

In some cases the co-twin will be affected with infection, hematological and neurological complications but in the present case the donor twin was normal. Some authors have observed absence of external genitalia in acardia twins.[5] However in the present case the external genitalia was well developed and both were female babies.

TTTS cannot be prevented; therefore an early diagnosis of this disorder in an identical twin pregnancy can possibly save one or both babies. This can be detected in the early stages of pregnancy by ultrasound scanning and Doppler velocimetry.[6] Currently to stop the blood flow to the acardiac twin, a high energy Radiofrequency Ablation (RFA) is utilized to destroy the blood vessels and surrounding tissue at the site where they enter the acardiac twin.[7] The other available therapy for TTTS is fetoscopic placental laser surgery directed at the vascular connections between the twins.[8]

It is important to exclude a chromosomal abnormality before offering a fetoscopic procedure in TRAP sequence, because the incidence of chromosomal abnormality in the pump twin may be as high as 9%.

Quintero et al., reported the first successful umbilical cord ligation for TRAP sequence. The pregnancy continued until 36 weeks of gestation, when a healthy boy was delivered. Thus "curing" the twin to twin transfusion syndrome before birth by elimination of the anastomoses is possible. However, in the present case the baby was saved spontaneously.

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