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Case Report

MCMA acardiac twin: a trap?

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ABSTRACT

Acardiac twinning or Twin Reverse Arterial Perfusion (TRAP) is a rare complication of monozygotic twin occurring due to retrograde perfusion of acardiac twin by structurally normal pump twin through abnormal arterio-arterial anastomosis in the placenta. Here we report a case of unexpected occurrence of acardiac twin diagnosed as monozygotic monoamniotic twin (MCMA) gestation with single foetal demise due to multiple anomalies, delivered vaginally suggesting a role for expectant management.

Keywords: Acardiac twin, MCMA twin, TRAP sequence

INTRODUCTION

Twin reversed-arterial-perfusion (TRAP) sequence is a rare and serious complication of monochorionic twin gestation. It occurs due to abnormal placental arterio-arterial anastomosis also often accompanied by a vein-to-vein shunt. "Reversed perfusion" is a term used to describe this scenario because blood enters the acardiac twin through reversed flow through its umbilical artery and exits through the Umbilical vein, which is opposite to the normal blood supply of the fetus. The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the pump twin. Mortality rate of acardiac twin is 100%. The reported incidence is 1 in 35000 pregnancies and 1 in 100 monozygotic twins.¹ Conservative management is a consideration though mortality among pump twin is 50%.²

CASE REPORT

A 25-year-old tribal patient, primigravida with irregular antenatal check-up diagnosed as monochorionic monoamniotic (MCMA) twin gestation with single foetal demise, presented to our Labour room with complains of

labour pains. Her clinical examination revealed stable vitals, uterus of 34-36 weeks' size, first twin in cephalic presentation, with good FHS and contractions +. Per vaginal examination finding was well effaced cervix, 2cm dilated with tense bulging membranes, vertex at -2, pelvis adequate.

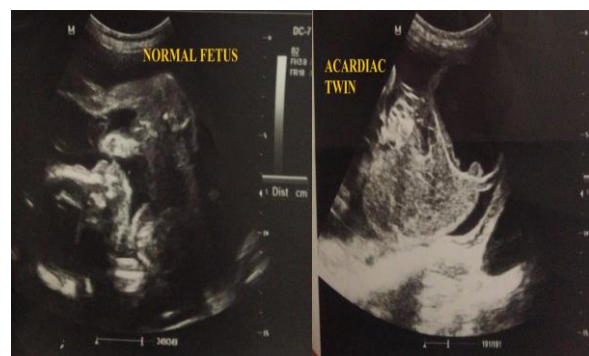


Figure 1: Ultrasound taken at 26 weeks showing MCMA twins.

She had a single antenatal visit at 26 weeks gestation in a local hospital where she was admitted and managed

expectantly in a local hospital. USG done at that time revealed MCMA twin pregnancy (Figure 1) Twin A- Transverse lie, normal foetus and biometry shows 24 weeks, Twin B-Anomalous foetus, dilated ventricles in brain, anterior abdominal wall defect present, parts of spine and long bones visualized. Placenta was single upper segment and adequate liquor.

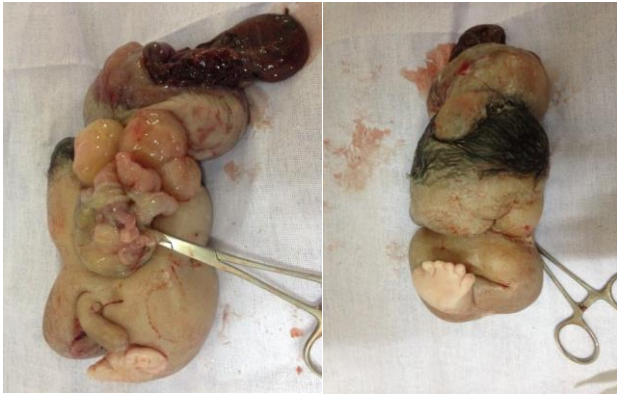


Figure 2: Acardiac aniceps.

Within three hours of admission, she delivered first a live male baby of weight 1.3 kg and second acardiac twin (Figure 2), sex not identifiable of weight 750g, amorphous mass with a cephalic element, poorly formed limbs, and anterior abdominal wall defect with protruding intestines. The pump twin had no visible malformation or features of congestive cardiac failure. Both twins shared same placenta. No obvious placental anomaly noted. Umbilical cord of acardiac twin was short. Patient refused autopsy of the acardiac foetus. Postnatal period was uneventful. The pump twin was admitted for low birth weight and discharged on postnatal day 10. (Figure 3).



Figure 3: Pump twin discharged on postnatal day 10.

DISCUSSION

Acardiac twinning is a unique complication of monozygotic twin pregnancies. It was first described by Bendetti as Chorioangiophagus parasiticum.³ There are two main hypotheses suggested for pathogenesis of

TRAP sequence.⁴ One is the deep placental anastomoses in early embryogenesis causes malformation of the acardiac twin. The early pressure flow in one twin exceeds that of other and leads to reversed circulation in the twin who exhibits perfusion. Another attributed cause is a primary defect in embryogenesis in one twin leads to failure of cardiac development. The normal twin then perfuses the acardiac twin via artery-artery anastomosis. The anastomoses are not responsible for the cardiac anomaly.

Acardiac twins are classified according to the degree of cephalic and truncal maldevelopment.⁵

- The first type is the most common one, Acardius acephalus, where no cephalic structures present.
- The second type is seen in our case Acardius aniceps where some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed. It is highly developed form.
- The third is Acardius acormus with cephalic structure but no truncal structures are present.i.e. Head without a body. The umbilical cord is attached to the head. It is rarest form of Acardia.
- The fourth type is Acardius amorphous with no distinguishable cephalic or truncal structure. It is least developed form.

Healthy pump twin succumbs to high output failure, hydrops fetalis and IUD. Prognosis depends on twin weight ratio.⁶ Management of acardiac twin pregnancy implies serial ultrasound to assess well-being of normal pump twin excluding any chromosomal anomalies and occlusion of blood flow to acardiac twin through modern techniques like endoscopic umbilical cord ligation, thrombogenic coil in umbilical artery of acardiac twin, endoscopic laser coagulation of arterio-arterial anastomosis, fetal image guided surgery using radiofrequency ablation.⁷ Ours was a rare case scenario of MCMA acardia aniceps twin diagnosed after delivery. The acardiac foetus was mistaken for a dead anomalous fetus in the earlier scan and Doppler study was not done. Vaginal birth was a possibility because first of the twin was the normal pump twin and its weight was more when compared to acardiac counterpart.

Conservative management is an option with a careful monitoring and ultrasound surveillance as no intervention was done in this patient. Early referral of high risk cases to tertiary centre and good antenatal care among the tribal population is a must. Goal of prenatal treatment is to stop blood flow to the acardiac twin without affecting the pump twin in order to improve its outcome in cases of increased twin weight ratio and cardiovascular impairment in pump twin.

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