Case Report

A Case Report of TRAP Sequence with Preeclampsia and Review of Literature

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Abstract:
We are reporting a case of the Twin Reversed-Arterial Perfusion (TRAP) sequence, specifically highlighting the autopsy findings of an acardiac twin and the histopathological findings in its placenta.The TRAPsequence or acardiac twining, is an uncommon congenital anomaly involving a grossly abnormal acardiac twin and an otherwise normal pump twin. Thiscomplex congenital anomaly is unique to monochorionic-monozygotic twin gestations.It requires an artery-to-artery(A-A) anastomosis leading to reversal of arterial perfusion with complex pathophysiology. A 19 year-oldgirl delivered twins by C-section at 31 weeks because of preeclampsia and non-reassuring fetal heart tracings.One was a preterm female; the other was an acardiac twin. This is perhaps the first case report of autopsy findings of an acardiac twin pregnancy associated with preeclampsia.

Keywords: *Twin Reversed-Arterial Perfusion, acardiac twin, monochorionic-monozygotic twin gestations, preeclampsia*

INTRODUCTION

Monozygotic twinning is an uncommon variety of twin gestation with an incidence of approximately 3-4 (0.3%) per 1000 births.[1] The birth of an acardiac twin, a condition otherwise known as Twin Reversed-Arterial Perfusion (TRAP) sequence, is a rare and serious complication of monochorionic twins since they are prone to develop a disproportionate blood supply. The incidence is estimated 1 in 35000-48000 births accounting an average risk of 1% among monochorionic twins.[2] Similarly, preeclampsia (and eclampsia) is also twice as likely to develop in twin (especially monochorionic twin) gestations contributing significantly to perinatal morbidity and mortality by limiting the blood flow through placenta.[3] In the setting of monoamniotic/monochorionic gestations the placenta has two umbilical cords inserted in close proximity to each other. This close insertion of two umbilical arteries leads to the formation of an A-A (or V-V) connection resulting into twin-to-twin transfusion (TTT); the phenomena termed as twin reversed arterial perfusion (TRAP). As a result, there is shunting of hypoxic/hypercapneic blood from a donor or pump twin to a recipient or acardiac twin. This mostly deoxygenated blood supply to recipient twin gives rise to an atrophic mass of tissue, with absent or rudimentary heart and upper body structures.[4] The pump twin is usually structurally normal but may develop heart failure due to excess burden of pumping blood to the acardiac mass as well as to its own growing tissues. If left untreated, up to 50% of these otherwise normal twins may die in utero (stillbirth) or die shortly after birth.[4] Early recognition of the TRAP sequence and appropriate management of the co morbid conditions (such as preeclampsia in our case) may have significant impact on survival of donor or pump twin.On the contrary, due to the presence of dramatic deformities, there are no chances of survival for acardiac twin.

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CASE REPORT

A 19-year-old Hispanic primigravida presented at 31 weeks of gestation to the emergency department in Texas, (USA)with an absence of fetal movements and abdominal pains for one day. Examination revealed a term sized gravid uterus with multiple fetal parts and increased amount of amniotic fluid. She had high blood pressure and fetal heart sounds could not be localized. She denied any previous visit to the gynecologist for antenatal care or any medication for her high blood pressure. Routine investigations were unremarkable except for urine dipstick protein of 2+. A stat ultrasonography showed a twin intrauterine gestation with single placenta and polyhydramnios. Twin A was bradycardiac but structurally normal; twin B was grossly abnormal and acardiac. Immediate C-section was performed because of preeclampsia and non-reassuring fetal heart tracings. One premature female infant was delivered (twin A) who was immediately shifted to neonatal intensive care unit due to bradycardia and cyanosis. We received twin B with an autopsy request and placenta for histopathological examination.

Autopsy revealed a malformed fetus with crown-rump length of 16 cm, crown-heel of 20 cm. It weighed 879 grams. Gross examination exhibited lack of development of facial structures, neck, shoulders and upper extremities (Figure 1). The scalp had scant thin hairs; the head, by palpation, showed open cranial sutures. The nuchal area exhibited marked edema consistent with cystic hygroma. In the area of the face there was a single orifice with partial formation of the

lips that exhibit dark purple discoloration; no eyes, nose, or ears were seen. The lower extremities were deformed and twisted around each other; the foot exhibited 3 toes. No anus was noted and no genitalia are seen. Attached to the abdominal wall was a 2 cm markedly dilated segment of umbilical cord; it exhibited 2 blood vessels and intestinal loops consistent with patent omphalomesenteric duct. A specimen's x-ray was obtained prior to sectioning revealing incomplete cranial bones, vertebral body defects and rudimentary ribs (Figure 2). There were malformed pelvic bones but normal appearing femurs and lower leg bones. An incision was made from the base of the facial orifice to the lower abdomen passing to the right of the umbilical cord. No thoracic organs were noted other than the esophagus. There were abdominal organs including the stomach, small intestine, and kidneys. No liver, spleen, adrenal glands, colon or internal genitalia were noted. There was a balloon like structure in the pelvis that appears to correspond to the bladder or a dilated urachus; it was connected directly to the umbilical cord appearing to enter at its base. In addition to this structure there were two blood vessels. Upon sectioning the scalp, there was release of abundant clear, yellowish fluid from a cavity in the nuchal area and corresponding grossly to a large cystic hygroma. Upon opening the cranium, the brain was autolysed and liquified.

Specimen 2 was received in formalin and consisted of a twin placenta that was irregular and measured 17 x 16 x 3 cm. It weighed 815 grams with two centrally attached two vessels umbilical cords separated by a distance of 1.5 cm without intervening T zone (monoamniotic) (Figure 3). One umbilical cord (designated arbitrarily as "A") measured 7.5 cm in length and 0.7 cm in width and exhibited tight left-handed twists. The other umbilical cord (designated arbitrarily as "B") measured 17 cm in length and varied in width from 0.7 cm near the base at the point of insertion to 1.8 cm at the distal end where it exhibited marked edema; In addition, there were tight right-handed twists and varicosities creating false knots but no true knots. The fetal surface showed multiple areas of nodular fibrinous deposits and markedly congested torturous vessels surrounding the insertion of umbilical cords. The dissection of the congested vessels revealed one of the vessels running from one umbilical artery to the other (artery to artery shunt). Maternal surface showed partially broken and ill-defined cotyledons. Sectioning revealed dark brown spongy parenchyma with multiple foci of fibrinous deposits and gritty calcification

Twin A was a pale-greyish preterm, female infant born at 31 weeks and 5 days, weighing 1550 g. Subsequently, she died on day 5 in neonatal intensive care unit as a result of congestive heart failure.

DISCUSSION

The incidence of multiple pregnancies is increasing due to popularity of ovulation inducing drugs and other assisted reproductive techniques. Twin, especially the monochorionic twin gestation is often a hazardous pregnancy due to high risk of development of twin-twin transfusion syndrome (TTTS). Acardiac twinning or twin reversed arterial perfusion (TRAP) sequence is an uncommon but the most extreme manifestation of monochorionic twin gestations resulting in poor survival of twins. In normal twin pregnancy, oxygen- and nutrient-enriched blood is distributed through the umbilical cords of each twin separately and independently(Figure 4). Oxygenated blood from the placenta enters the fetuses through the umbilical veins and travels immediately through the ductus venosus to the right atrium. In the TRAP sequence, there is a large A-A or V-V connection between superficial chorionic vessels cutting the direct placental blood supply to one of the twins. This twin is termed as acardiac twin since there is absent or malformed heart. Acardiac or recipient (also known as parasitic) twin receives all of its blood supply from the normal co-twin giving rise to the term "pump twin."

Because the umbilical arteries arise from the iliac arteries, the lower body of the recipient is perfused first with any remaining oxygen and nutrients; it generally shows more recognizable development leading to typical morphology. Thus the blood reaching the upper body is severely deficient in oxygen and nutrients resulting into extensive atrophy of the upper body structures like heart, head and arms. Due to unequal perfusion, none of the two parasitic twins are alike. They are little more than a torso with or without legs, showing a spectrum of morphologies categorizing into four types. (5)

1. The acardius-acephalus fetus has complete absence of head. This is most common type of acardiac twin overall.
2. The acardius-anceps fetus has some cranial and neural development with recognizable torso and extremities. This is the most developed variety however lacks even a rudimentary heart. This is the type seen in our case too.
3. The acardiac-acormus fetus has head development with umbilical cord attached to it, but complete absence of truncal structures. It is the least common variety.
4. The acardius-amorphus fetus has no cephalic or truncal development. It has minimal development as a human and differs from teratoma only by its attachment to an umbilical cord. (5)

Although vascular anastomoses in cases of TRAP have been well described, a little disagreement over the

sequence of events exists in its pathogenesis:

One school of thought proposes that the primary defect is in embryogenesis (dysmorphogenesis). There is no cardiac development in the early period of organogenesis. This leads to establishment of an artery-to-artery anastomosis resulting in TRAP sequence. Other group favors the development of an abnormal artery-to-artery anastomosis between embryos during early embryogenesis. (6) This vascular communication in the placenta leads to reversed flow of blood to the hemodynamically disadvantaged or recipient twin, with resulting secondary atrophy of the heart and dependent organs.

Gross examination of monochorionic or monoamniotic twin placenta in our case confirmed the presence of an artery-to-artery (A-A) anastomosis (figure 3).Both umbilical cords showed a single umbilical artery. Single umbilical artery is a common occurrence and associated with 66% cases of acardiac twinning or TRAP sequence (2). In our case, preeclampsia was another contributing factor towards compromised blood supply and dismal outcome of both twins.Several studies have shown that utero-placental blood flow is decreased in PIH due to maternal vasospasm and accordingly it may lead to fetal hypoxia, distress and death. (3).

TRAP sequence is often complicated bycongestive heart failure of the pump twin due to extra burden of pumping blood to its own growing tissue as well as acardiac mass. However, the risk of having heart failure and dying of normal (pump) twin seems to depend predominantly on the size of the acardiac twin. A study conducted by Moore et al concluded that if twin-weight ratio is above 70%, the chances of having congestive heart failure is relatively less (30%). In our case, pump to acardiac twin weight ratio was less than 70%. Polyhydramniosis another complication that was present in our case too, may in turn cause hydrops and premature labor. It is a consequence of polyuria due to increased blood flow to the kidneys.In order to maintaining the viability of the donor/pump twin, close surveillance for the size of recipient and strict monitoring for other comorbid conditions via regular clinical and radiological checkups is necessary. However, the TRAP sequence does not appear to run in families and there are currently no reports of recurrence in the same family.

CONCLUSION

The case findings support the concept that placental vascular pathology is a major contributor to adverse outcome in monochorionic twin pregnancies. Based upon these pathological findings, we believe the sequence of events in this case was early twin-to-twin transfusion (A-A anastomosis), which resulted in circulatory reversal along with a further limitation of blood flow due to pre eclampsia. Postmortem examination of the placenta confirmed monochorionic, monoamniotic placentation with two adjacent two-vessel umbilical cords. The histological findings confirmed an artery-to-artery anastomosis and the histological features of pre eclampsia. Twin B was enormous hydrops with complete absence of the heart and multiple other congenital anomalies, which are summarized in autopsy findings. Due to paucity of published literature on the microscopic changes in the placenta regarding the TRAP sequence, the exact mechanisms that lead to twin-to-twin transfusion are not fully understood. Careful gross and microscopic examination of all monochorionic twin placentas is highly recommended to provide appropriate perinatal management for prevention of serious complications and to improve the survival of twins.

Figure 1: Acardiac twin (Twin B) 121X91mm

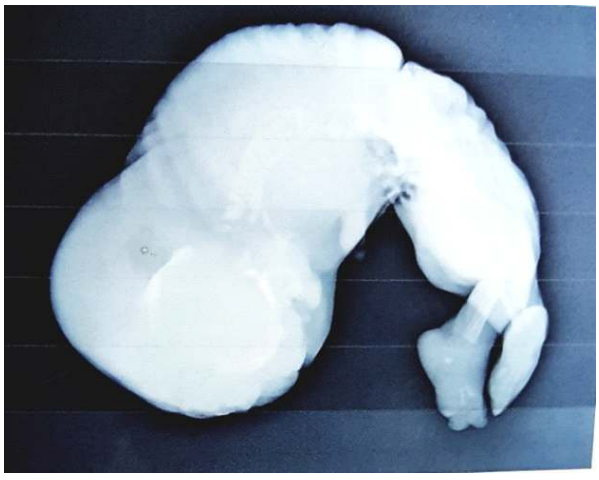

Figure 2: Specimen X ray

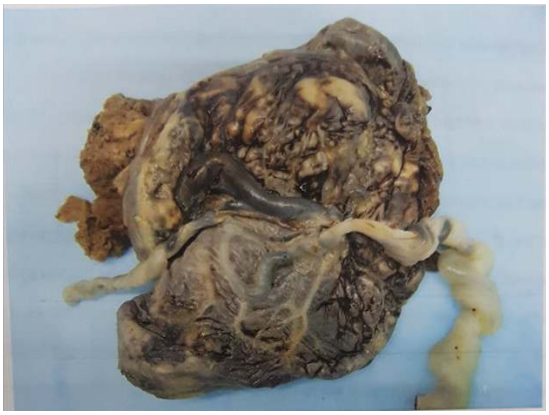

Figure 3: Placenta (Gross) with A-A anastomosis

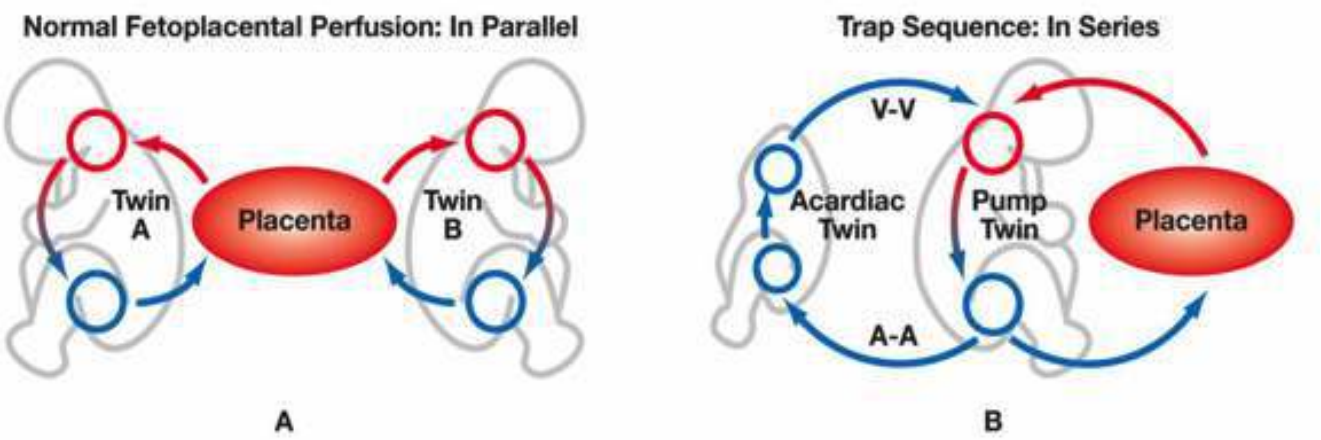

Figure 4: Diagrams illustrating fetoplacental circulation in twins. A: Normal twin fetoplacental circulation B: Twin reversed arterial perfusion (TRAP) sequence.

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