Elusive Prediction of Monoamniotic Acute Twin-Twin Transfusion Syndrome: A Case Report and Review

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Introduction

Monochorionic monoamniotic (MCMA) pregnancies occur in 8 in 100,000 pregnancies and exhibit a substantial peripartum mortality risk of 40%.[1] Twin-twin transfusion syndrome (TTTS), a typical finding of monochorionic diamniotic (MCDA) pregnancies, with incidence of 15%, has only 4% incidence in MCMA pregnancies.[1] The main cause of unexplained MCMA intrauterine fetal demise (IUFD) is attributed to cord entanglement, however reports suggest this may be invalid.[1,2] Moreover, this supposition is insufficient to explain why a percentage of such twins succumb when all MCMA twin pairs have cord entanglement in all trimesters. An acute TTTS has been proposed as an alternative explanation for unclear IUFD of MCMA twin pairs.[1,4-6] **Figure 1** shows an example of the classic visual appearances of a surviving TTTS twin pair postpartum.

We present a case of mid-trimester MCMA twin demise with features of TTTS at delivery (discordant fetal size, color) within 1 week of healthy antenatal ultrasound (US) testing.

Case Presentation

A 36-year-old patient G2P1001 with known MCMA twin gestation at 26 0/7 weeks presented with decreased fetal movement. The patient had been followed with biweekly US examinations and outpatient serial fetal heart rate (FHR) monitoring. She was seen 4 days previously for fetal testing which revealed healthy FHR tracing and BPP 8/10 for both twins. Growth US 1 week prior revealed concordant fetal growth- Twin A 780g, Twin B 768g, weight discordance 1.5% and unremarkable structural survey. On admission, US showed MCMA twin pair demise. The patient was taken for repeat C-section that was performed without complications of demised discordant twins- the larger twin weighed 1005g and exhibited marked erythema, the smaller twin weighed 726g and pale. Birthweight discordance was 27.8%. **Table 1** shows the calculated fetal placental blood volumes at 1 week check-up and delivery day using the regression equation from Mandelbrot et al.[7]

Figure 1: TTTS twin pair with erythematous recipient (left) and pallid donor (right).[8]

	1 week prior to delivery	Delivery day
Twin A	110.25 ml	141.75 ml
Twin B	108.57 ml	102.69 ml
Twin-Twin Volume Difference	1.68 ml	39.06 ml

Table 1: Estimated fetal blood volumes at prenatal structural survey check-up and delivery.

Discussion

Cord entanglement occurs in virtually all MCMA pregnancies.[1-3] Detectable from the first trimester, the phenomenon is monitored for fetal blood restriction to each twin by US with umbilical cord and middle cerebral artery doppler and, more commonly, FHR tracing to detect cardiac distress.[2,3] As most IUFDs occur mid-trimester, tight cord entanglement should increase mortality equally across trimesters. However, the last trimester has the least risk of mortality, and the first trimester does not have a death rate as high as the second.[9] In our case, the patient presented with twins with no signs of cardiac distress with BPP 8/10 only 4 days prior to delivery. The tight cord entanglement theory fails to explain twin-pair death here against a 27.8% twin pair-weight discordance on delivery day versus 1.5% difference a week prior.

Mechanistically, extensive vascular anastomoses drive TTTS presentation and exist naturally in monochorionic placentas.[4] MCMA pregnancies are posited to be protected from TTTS due to increased number of superficial arterioarterial (AA) anastomoses in the placenta that allow balanced blood flow between twins.[4,6] The unidirectional flow of placental arteriovenous (AV) anastomoses is canceled out in effect. The observed TTTS in this case may have resulted from blood shunting subacutely through placental AA anastomoses by a path of least resistance to 1 twin, as has been previously discussed in other reports.[4,6]

With the absence of an oligohydramnios twin the determination of MCMA TTTS relies on less indicative findings of uneven bladder filling, doppler studies and presence of polyhydramnios with maximum amniotic vertical pocket of greater than 10 cm after 20 weeks gestation.[1,4] In MCDA TTTS twins, 1 fetus has polyhydramnios (maximal vertical pocket of above 10 cm amniotic fluid after 20 weeks) and the other oligohydramnios (characterized with a pocket length under 2 cm) with both twins occupying 1 placenta.[4] In MCDA pregnancies, with the presence of the classic oligo- and polyhydramnios twin pair, the diagnosis is clearer and likely leads to higher incidence rate. Lacking the oligohydramnios twin component in addition to otherwise healthy FHR tracing and antenatal twin-pair weight difference on prenatal screening, MCMA TTTS was challenging to predict in our patient.

The ability to screen for developing TTTS in MCMA twin pairs is therefore compromised, making surveillance and treatment standards for TTTS in MCMA pregnancies deficient.[4,6] The absence of reliable sonographic markers and sudden onset of acute TTTS frustrates the ability to design an effective antenatal surveillance strategy. The acuity of the process also limits the potential effectiveness of serial middle cerebral artery dopplers looking for disparate blood flow peak velocities.[6,10] Acute alterations in fetal behavior- analogous to those seen in acute massive fetomaternal hemorrhage- are less reliable in the setting of twin gestation and may occur too late for any clinical intervention to alter the outcome.[6,10] Therefore, to detect acute TTTS, a provider would theoretically need to continuously have a patient under US and FHR monitoring throughout the pregnancy course, which is financially costly and logistically challenging to arrange.[11]

Acute TTTS thus remains an elusive diagnosis to predict in MCMA pregnancies. More research into screening modalities attuned to the rapid onset of acute TTTS is needed to lower the mortality risk of the condition.

Works Cited

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