JACOBS PUBLISHERS

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Jacobs Journal of Gynecology and Obstetrics

Case Report

Expectant Management of Monochorionic Gestations Complicated by Fetal Anomaly of One Twin: Case Report and Review of Literature

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Received: 05/16/2015

Accepted: 07/16/2015

Published: 08/01/2015

Abstract

Background: The prenatal course of anomalous monochorionic, diamniotic (MC/DA)gestation requires intricate management. Literature on this topic is limited to management of normal monochorionic gestation and does not translate to management of anomalous monochorionic pregnancies. The only choice for selective reduction is cord ligation with the risk of losing the entire pregnancy. It is essential that we come up with some guidelines for MC/DA pregnancies as far as expectant management and proper timing of the delivery to prevent loss or damage of the normal twin. We report a case of a single anomalous MC/DA and its management dilemmas.

Case: A patient with MC/DA twin pregnancy remarkable for one fetus with multiple anomalies and the other twin completely normal. The patient was offered selective reduction. She opted for expectant management due to religious reasons and fear of losing the entire pregnancy. Cesarean section at 32 0/7 weeks was performed due to an acute change in the anomalous twin and the fear of loosing the normal twin due to placental connection. The anomalous twin expired immediately after delivery.

Conclusion: While there is some consensus regarding the expectant management of uncomplicated MC/DA twin gestations, the management of complicated MC/DA twins and timing of delivery requires additional studies, case reports and guidelines as far how to follow them and when to deliver.

Introduction

Assisted reproductive technologies (ART), including in vitro fertilization and ovarian stimulation, have contributed to the incidence of twinning. This technology specifically has been associated with a slight increase in the rate of monozygotic twins, which implies an increased rate of monochorionic twins (MC). Strategies for management of normal MC twin pregnancies have been discussed in the literature, yet MC twins are at increased risk for various anomalies (such as in the case reported here), and thus require intricate management. Information and guidelines regarding expectant management of anomalous MC pregnancies are very limited. Currently, the literature regarding management of anomalous MC twins is

largely limited to selective fetal reduction via cord ligation, increased frequency of prenatal monitoring, amnioreduction, or fetal surgery. Further studies are needed in order to advance the strategies for expectant management of anomalous MC gestations and to establish more extensive guidelines, specifically regarding the timing of delivery to prevent the loss of the normal twin.

Case

A 31 year old patient of Ashkenazi Jewish descent presented to our high risk obstetrical service with a MC/DA twin gestation. Her obstetrical history included 6 full term uncomplicated vaginal deliveries. Aside from morbid obesity, her past medical, gynecological, surgical, and social histories were noncontributory.

The patient's first trimester course was unremarkable. At 23 weeks, many anomalies were detected in Twin B. Those included total body edema, right pleural effusion, absence of a stomach bubble, bilateral hydrocephalus, hemivertebra and kyphoscoliosis in the thorax and thoraco-lumbar areas, and micrognathia. A fetal echocardiogram also demonstrated dextroposition with levocardia but normal segmental anatomy and valve function. In addition, the fetal head was hyperextended, and the arms were crossed over the chest with little or no upper body movement.

The anatomy scan and fetal echocardiogram for Twin A was unremarkable, and both twins had normal umbilical artery dopplers. The patient was counseled that due to Twin B's multiple anomalies, in utero demise was possible which could subsequently compromise Twin A due to sharing of the placenta.

She was offered amniocentesis and/or termination; both were declined. Weekly Biophysical profiles (BPP)/Dopplers starting at 28 weeks and growth sonogram every 2 weeks were performed.

Twin A developed polyhydramnios, while twin B developed oligohydramnios and progressive hydrocephalus. At 30 weeks gestation, Twin B was noted to also have an imperforate anus and rocker bottom feet.

At 31 0/7 weeks, the patient was admitted to Labor and Delivery with a sudden increase in blood pressures (160/100), new onset of bilateral lower extremity edema, and elevated uric acid. BPP for both twins was 8/8.

Steroids for fetal lung maturity and magnesium sulfate for seizure prophylaxis were initiated. On the 5th hospital day, Twin A (normal twin) demonstrated adequate growth, normal umbilical artery dopplers, and a BPP of 6/8 (2 off for breathing).

Growth for Twin B (the anomalous twin) was in the 15th percentile, with an abdominal circumference in the 6th percentile. Severe oligohydramnios was also noted and the S/D ratio was 5.1. The BPP for Twin B was 6/8 (2 off for amnionic fluid index). Due to the declining status of Twin B and fear of IUFD that could lead to demise of twin A, it was decided to proceed to an urgent Cesrean section.

At delivery, Twin A weighed 1410g with Apgar scores of 1/5/5, venous gases of 6.90, and a base deficit of 15.3. Twin A was intubated and taken to the Neonatal Intensive Care Unit (NICU).

Twin B weighed 1485g with Apgar score of 0/0. A brief attempt at intubation was unsuccessful due to an obstructed airway. Autopsy was offered to the family but the family refused due to religious reason.

Twin A remained in the NICU and was eventually discharged with no major neonatal events noted. Since then, Twin A has been doing well over past three years.

Comment

Monozygotic (MZ) twin pregnancies, which result from a division of a single fertilized egg, account for about one third of all twin pregnancies. MZ pregnancies divide further into monochorionic (MC) if the two fetuses share a placenta and dichorionic (DC) if each fetus has a separate placenta. All types of twin pregnancies are at increased risk for almost every pregnancy complication, including but not limited to preterm delivery, intrauterine demise, and various anomalies. Nevertheless, compared to DC twins, MC monozygotic twins are known to be at a higher risk for further complications due to the vascular anastomoses between the two fetal circulation systems. Those complications include twin-twin transfusion syndrome (TTTS), twin anemia polycythemia sequence (TAPS), twin reversed arterial perfusion (TRAP) sequence, monoamnionic twinning [1], intrauterine Fetal Demise (IUFD), early preterm delivery, and intrauterine growth restriction (IUGR) [2]. MC twins, depending on the time of cleavage, further divide into monochorionic diamniotic (MCDA) and monochorionic monoamniotic (MCMA). MCMA are at the greatest risk for all complications, but they account for only approximately 1% of monozygotic twins [2].

Despite having identical DNA, monozygotic twins are not necessarily identical in structure formation and despite having a normal karyotype (as in the case presented) can often develop independent abnormalities. Moreover, structural anomalies are three times higher in MZ twins compared to dizygotic twins, and five times higher compared to singletons.

Having severe anomalies in one of the fetuses can put the unaffected twin at high risk for preterm delivery and even death due to the development of polyhydramnios by the affected twin [4] and thromboembolic phenomenon. Such severe anomalies of-

ten involve central nervous system (CNS) abnormalities, twin reversed arterial perfusion (TRAP), cardiovascular system (CVS) abnormalities, abdominal wall/gastrointestinal abnormalities, skeletal abnormalities, and hydrops [3]. In such cases, selective fetal reduction is often recommended and performed in order to optimize the outcome of the unaffected twin.

Different techniques for selective fetal reduction in MC twins have been discussed in literature [3]. However, unlike in singleton and DC pregnancies, performing selective fetal reduction in MC twins imposes additional risks, including procedure-related risk to the unaffected twin, as well as small maternal risks from retention of a volume of nonviable fetal tissue [3]. Due to the additional risks of selective fetal reduction in MC and/or refusal of the mother to allow the procedure for various reasons (as in the case described), conservative expectant management is often practiced. These cases then require a very challenging and closely monitored expectant management.

Strategies of uncomplicated (normal developing) MC twin pregnancies have been discussed in the literature. Barigye et al. [5] published data from 151 uncomplicated MCDA pregnancies to investigate IUFD. Heightened fetal surveillance was practiced in order to detect TTTS and IUGR as early as possible, to allow timely treatment in an attempt to prevent adverse perinatal outcomes.

The surveillance included: 1) first trimester nuchal translucency assessment and chorionicity determination 2) a mid-trimester anatomical survey 3) fetal echo followed by growth scans, amniotic fluid assessment, and Doppler studies every 2 weeks throughout the pregnancy. Despite the intensively monitored pregnancies, 10 unexpected IUFD (4.6%) occurred, suggesting that apparent healthy MCDA pregnancies are still at risk for IUFD. Moreover, the IUFD occurred predominantly after 32 weeks of gestation.

Overall, it appears that premature delivery risks such as respiratory distress syndrome, intraventricular hemorrhage, and necrotizing enterocolitis are significantly lower than the risk of IUFD. This led Barigye et al. [5] to conclude that in addition to intensive monitoring, elective delivery at 32 weeks should be practiced in MCDA as a preventative approach.

There are not clear guidelines regarding expectant management of complicated MC twin pregnancies with one anomolus pair, most likely because it is a fairly rare condition,

Specifically, there is a lack of information regarding the recommended frequency of monitoring visits, management of the third trimester, and the most optimal delivery time to prevent demise or damge of the normal twin.

Furthermore, currently it is uncertain whether the best treatment of complicated MC pregnancies is expectant management or selective fetal reduction [6].

The majority of the literature that does discuss MC pregnancy with an anomalous twin, focuses on techniques for selective fetal reduction. Those techniques are based on occluding blood flow within the umbilical cord in various ways, including radiofrequency ablation, bipolar cord occlusion, and endoscopic laser coagulation [3,6].

The case described in this report was further complicated by the onset of severe pre-eclampsia, an increased risk with multiple gestations. The management of pre-ecplamsia prior to 34 weeks of gestation is controversial and case specific. In general, in cases of women with severe pre-eclampsia, a viable fetus, and before 34 weeks, a policy of expectant management is recommended. However, in cases of uncontrolled maternal hypertension, increasing maternal organ dysfunction, or fetal distress, delivery is recommended [7].

In the case presented, expectant management was attempted; however, fetal compromise (i.e. oligohydramnios and abdominal umbilical artery Dopplers) necessitated immediate delivery in order to protect the normal twin. Despite all we had done, there was a very low PH at delivery for the normal twin, raising the question of proper timing of delivery to have a better umbilical cord PH and associated improved outcome.

In conclusion, expectant management of MC twins with discordant anomalies (including the frequency of follow up visits, the management of third trimester, and the optimal delivery time) is not well established and requires further study. More research is needed to aid physicians with management of these cases, hence all cases on this topic should be published so we could eventually have a guideline on management of these pregnancies.

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