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A case of discordant monochorionic diamniotic twin with umbilical cord entanglement after spontaneous rupture of the dividing membrane

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Summary

Spontaneous antepartum rupture of the dividing membrane in monochorionic diamniotic twins with discordancy is extremely rare. The rupture is difficult to diagnose prenatally and has a poor outcome. The authors report a case of cord entanglement after spontaneous rupture of the dividing membrane within discordant monochorionic diamniotic twins. The subject was a 30-year-old woman pregnant with discordant monochorionic diamniotic twin at 27+4 gestational weeks. The relatively thin dividing membrane was sound until it passed parallel to the two umbilical cords where it then became ill-defined. The patient was managed cautiously due to the possibility of spontaneous rupture of the dividing membrane and potential cord entanglement. Upon delivery at 29+3 weeks due to fetal compromise, the patient presented with a monochorionic diamniotic placenta, a remnant of the disrupted dividing membrane, and entangled umbilical cords. The authors report this subject with literature review.

Key words: Spontaneous rupture; Dividing membrane; Discordant twin; Umbilical cord entanglement; Monochorionic diamniotic placenta.

Introduction

Discordant twins occur in monochorionic diamniotic (MCDA) placenta because of vascular anastomosis. The blood flow through the vascular connections becomes unbalanced, resulting in discordant twin such as twin-to-twin transfusion syndrome (TTTS) [1]. Monochorionic monoamniotic (MCMA) twin gestations have the highest perinatal mortality rate because of umbilical cord entanglement [2, 3]. Umbilical cord entanglement can occur in MCDA twins due to rupture of the dividing membrane (DM). Once the DM is disrupted, the pregnancy is similar to MCMA twins, with an increased risk of umbilical cord entanglement and an increased perinatal mortality rate [4]. Spontaneous rupture of the membrane in MCDA twins with discordancy is extremely rare. The authors report a case of rupture of the membrane in discordant MCDA twins with literature review.

Case Report

A 30-year-old nulliparous woman was transferred to the present hospital at 27+4 weeks gestation, because of discordant twin. The patient was diagnosed with MCDA twin pregnancy during the first trimester. On ultrasonography, a MCDA twin gestation with discordant 29+3 weeks and 26+0 weeks gestation size fetuses with amniotic fluid indexes (AFIs) of 25 and 7 cm was observed. The first fetus had cardiomegaly and an enlarged bladder. The second fetus displayed an absence of diastolic flow of the umbilical artery (Figure 1) and the bladder was not visible. In addition, a difference in the diameters of the umbilical cords was identified (Figure 2). Except for the obscure difference of amniotic volume, the discrepancy of fetal size, bladder appearance, and abnormal Doppler finding of the umbilical artery are appropriate for the diagnostic criteria of TTTS by Quintero *et al.* [1].

The relatively thin dividing membrane was sound until it passed parallel to the two umbilical cords where it then became ill-defined. This dividing membrane appeared loose and movable near the umbilical cords. It was suspected that the dividing membrane was either adherent to the umbilical cords or spontaneously disrupted (sonographic image unavailable). Therefore, the patient was admitted to the present hospital for careful observation because of suspected TTTS with rupture of DM.

The patient presented with severe dyspnea and premature uterine contractions. The authors performed amnioreduction at 27+5 weeks and removed 1.1 L of amniotic fluid. After this procedure the patient's symptoms improved. The NST pattern was reactive and the diastolic flow of the umbilical artery was observed through Doppler sonography in the smaller fetus. At 28+5 weeks, on ultrasonography, the fetuses were of sizes 30+5 and 27+5 weeks gestation and had each AFI of 26 and 7 cm. The smaller twin displayed an absence of diastolic flow of the umbilical artery and the bladder was not visible. Therefore, the authors removed 1.8 L of amniotic fluid by secondary amnioreduction. After amnioreduction, both the diastolic flow of the umbilical artery and a small bladder were observed in smaller twin at 29+1 weeks. At 29+3 weeks gestation, a cesarean section was performed due to decreased reactivity with deceleration in NST and absent diastolic flow of the umbilical artery in smaller twin. Discordant twin female fetuses with umbilical cord entanglement were delivered (Figure 3). First twin weighed 1,550 grams and had Apgar scores of 4 at one minute and 6 at five minutes. An-

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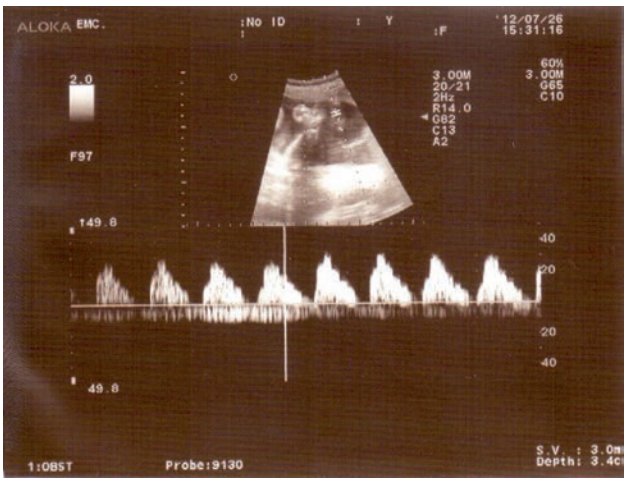

Figure 1. — Absent diastolic flow of umbilical artery in smaller twin.

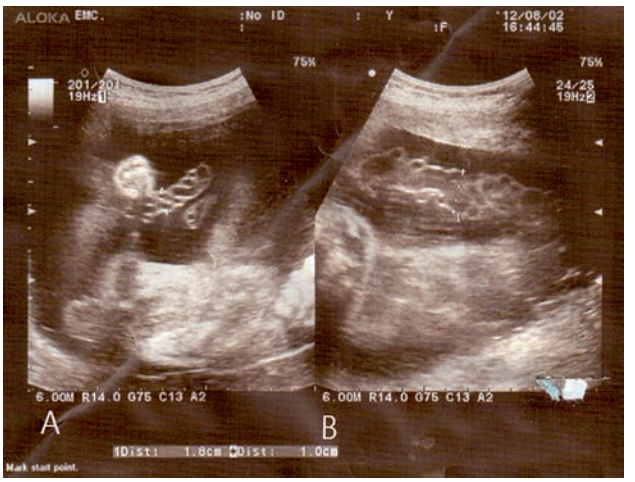

Figure 2. — Difference in diameters of the umbilical cords between smaller twin (A) and larger twin (B).

other twin weighed 1,130 grams and had Apgar scores of 4 at one minute and 7 at five minutes. However, the babies needed tracheal intubation and were admitted to the neonatal intensive care unit. The hemoglobin level of first baby was 17.6 mg/dL, and that of second baby was 15.7 mg/dL. An examination of the placenta disclosed an MCDA organ with a remnant of the disrupted dividing membrane. Histopathology confirmed the MCDA placentation. The patient's postoperative course was uneventful and she was discharged on postoperative day 5.

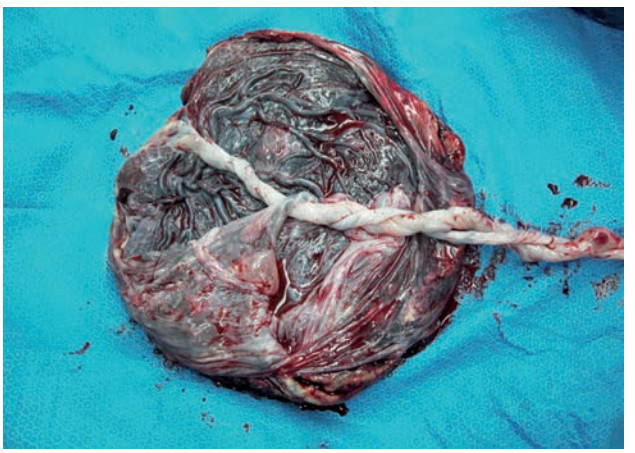

Figure 3. The placenta with entangled umbilical cords after delivery.

Discussion

In monochorionic diamniotic placenta, discordant twins develop from vascular anastomosis. The unbalanced blood flow through the vascular anastomosis results in discordant twins such as TTTS [1]. The present case was appropriate for diagnosis of TTTS, except for discrepancy of amniotic volumes. Based on the criteria by Quientero *et al.*, this case was stage III because of the absent bladder and abnormal Doppler finding of the umbilical artery [1]. However, the difference in amniotic volume was not greater, and there were abnormal findings of the umbilical vessels and DM, which were suggested to rupture the DM and cord entanglement. The present authors confirmed the discordant twins with ruptured DM and cord entanglement after delivery. TTTS was not diagnosed in the babies of the present case because of the difference in the hemoglobin level. The authors thought that this result might be attributed to the rupture of the DM. Actually, iatrogenic rupture of the DM, called septostomy, could be used as a modality of TTTS treatment. Through the procedure, the progression of the disease used to be delayed, and the babies' condition improved, especially, that of the donor twin because of increased amniotic volume [6]. In this case, the spontaneous rupture of the DM was suggested to delay the progression to TTTS by the same effect as septostomy.

During pregnancy, spontaneous rupture of the dividing membrane of MCDA twins is extremely rare and difficult to diagnose prenatally. The major etiologic factors are suspected to include a traumatic or physical rupture of the membrane [5]. The other probable cause is septostomy, which is a potential therapeutic modality for TTTS [6]. In addition, iatrogenic septostomy following amniocentesis and other invasive procedures in MCDA twins have also been reported [7].

The present group also utilized amniocentesis as there was minimal risk of a secondary rupture from amnioreduction. The risk was determined very low due to the following reasons: 1) suggested evidence of rupture such as an obscure difference between the amniotic volume and the parallel running pattern of two umbilical cords were observed prior to the amnioreduction; 2) there was no observable difference in the ultrasound findings prior to and post procedure, ex-

cept the difference of amniotic volume in the larger fetus; 3) the puncture site was specifically chosen as it was distant from the dividing membrane and near the larger twin, that showed marked polyhydramnios.

In the present case, the definite cause of the disrupted membrane is still uncertain. The authors speculate that the spontaneous rupture of the dividing membrane occurred due to a combination of thin membrane and active fetal movement [8]. The exact timing of the spontaneous rupture of the dividing membrane and the resulting umbilical cord entanglement is unknown. However, the diagnosis is very important for the prediction of umbilical cord entanglement and adverse fetal prognosis. Therefore the diagnosis of MCDA twins should be made cautiously, with special attention paid to the thickness of the membrane, the running pattern, and the insertion site of umbilical cords. Once the dividing membrane is ruptured, the MCDA twin gestations have high perinatal mortality rates of up to 70%, and a greater than 50% chance of umbilical cord entanglement [9]. Umbilical cord entanglement may be initially loose but it still has the potential to tighten and compromise fetal circulation later in pregnancy. In the present case, suspicious signs of fetal compromise were detected and a timely cesarean delivery was performed. Therefore, the authors recommend a thorough assessment of the DM at ultrasound examination in severe discordant MCDA twin which show obscured discordant amniotic volume. In addition, the umbilical cords should be assessed for evidence of entanglement.

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