CEOG Clinical and Experimental Obstetrics & Gynecology

Case Reports

Management of monochorionic monoamniotic twins: case reports and a review of literatures

J.Y. Lee, H.-H. Cha, W.J. Seong

Department of Obstetrics and Gynecology, Kyungpook National University Hospital, Kyungpook University School of Medicine, Daegu (Republic of Korea)

Summary

Monochorionic monoamniotic (MCMA) twins are rare but are associated with higher morbidity and mortality rates than monochorionic diamniotic (MCDA) twins. Although the prenatal prognosis of MCMA pregnancy has improved with advanced prenatal care, no consensus has been reached for the management for MCMA twins. Here, the authors report two cases of MCMA twin pregnancies diagnosed at 30 weeks of gestation and complicated with congenital heart disease, respectively, with a review of literature on MCMA twin pregnancies.

Key words: Monochorionic monoamniotic (MCMA) twins; Monochorionic diamniotic (MCDA) twins.

Introduction

Monochorionic monoamniotic (MCMA) twins are rare, accounting for approximately 1–2% of monozygotic twin pregnancies [1], but are associated with a 10–20% rate of intrauterine death beyond viable gestational age [2, 3]. Advanced prenatal care, including ultrasonography, non-stress test (NST), or cardiotocography, have improved the perinatal outcomes in MCMA twin pregnancies. In addition, early detection can lead to an increase in requirements for proper antenatal counseling about MCMA twins. However, considerable practical variations exist among maternal fetal medicine specialists [4]. The authors aimed to report two cases of MCMA twin pregnancies along with a review of the literature.

Case Report

Case 1

A 37-year-old woman (gravida 3, para 2) with twin pregnancies was referred to this institution at 30+1 week of gestation for the management of short cervical length and gestational diabetes (GDM) A1. She became pregnant naturally and known to have monochorionic diamniotic (MCDA) twins. However, inter-twin membranes were not visible, and the presence of significant cord entanglement was suspected in a color Doppler study (Figure 1). She was admitted for antenatal corticosteroid administration and fetal surveillance using ultrasonography and non-stress test (NST). After two days, the NST result showed recurrent variable decelerations in both fetuses. After completion of antenatal corticosteroid dose, the authors decided to perform immediate cesarean section, suspecting cord entanglement. Shortly before the cesarean section, the authors administered magnesium sulfate ($MgSO_4$) for fetal neuroprotection [5]. Umbilical cord entanglement and true knots between two cords were observed during the operation (Figure 2).

Baby A was female and weighed 1,430 grams, with one- and five-minute Apgar scores of 6 and 8, respectively. Baby B was also female and weighed 1,290 grams, with one- and five-minute Apgar scores of 4 and 7, respectively. Both neonates were admitted to the intensive care unit (NICU) and required ventilator care and surfactant therapy for respiratory distress syndrome (RDS). Both neonates were discharged after seven weeks, but Baby B was readmitted shortly for LASER operation because of retinopathy of prematurity (ROP). Both neonates are currently doing well.

Case 2

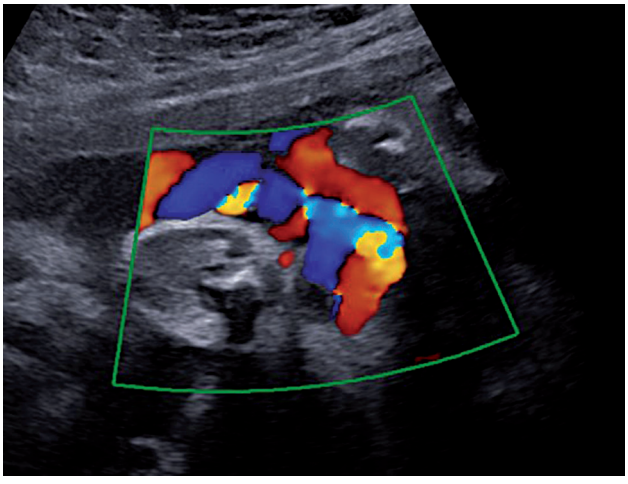

Figure 1. — Cord entanglement.

Revised manuscript accepted for publication November 9, 2017

Clin. Exp. Obstet. Gynecol. - ISSN: 0390-6663
XLV, n. 6, 2018
doi: 10.12891/ceog4350.2018

7847050 Canada Inc.
www.irog.net

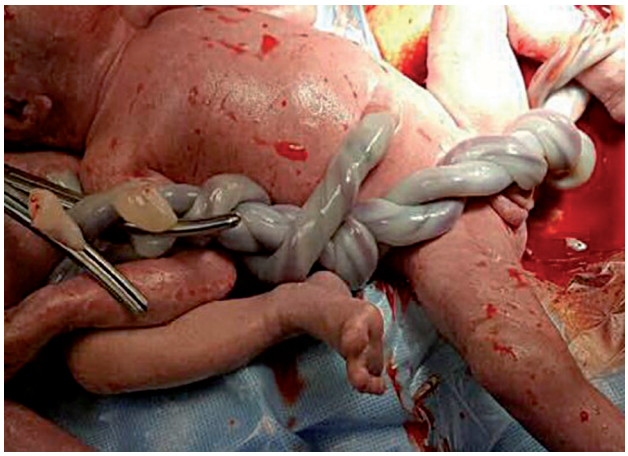

Figure 2. — Cord knotting.

A 32-year-old woman (gravida 1, para 0) was referred to this institution at 33+1 weeks of gestation for management of MCMA twin pregnancies complicated with heart anomaly in both fetuses. She was diagnosed as having MCMA twins at an early gestational age, and both fetuses were suspected of having ventricular septal defects on ultrasonography at 20 weeks of gestation. The presenting fetus had a large ventricular septal defect (VSD) 6 mm in diameter that required postnatal surgical correction. At her first visit to this institution, ultrasonography revealed one fused placenta and the inter-twin membranes were not visible between the two fetuses. In addition, both fetuses were presumed to have the same sex (female). Therefore, the authors diagnosed her pregnancy as MCMA twins. In addition, they performed fetal echocardiography and found a large ventricular septal defect, abnormal three-vessel view, and situs inversus totalis in the presenting fetus. Only two of the three vessels were found and showed parallel ventricular outflow tracts with a large VSD that was suspicious of double outlet of the right ventricle type of transposition of the great arteries (double outlet of the right ventricle [DORV] TGA type). The other fetus had no definitive abnormal finding. In addition, no definitive finding of cord entanglement was found. Considering the fetal congenital heart anomaly, the authors decided to continue her pregnancy up to at least 34 weeks of gestation. She was admitted for intensive fetal surveillance and administration of antenatal corticosteroid. Daily NST and Doppler study showed reassuring results. At 33+4 weeks of gestation, preterm labor developed with variable deceleration in both fetuses; therefore, immediate cesarean section was performed.

The first baby was female and weighed 2,180 grams, with one- and five-minute Apgar scores of 3 and 5, respectively. Postnatal echocardiography and chest CT revealed a double inlet of the left ventricle, DORV with a large subpulmonic VSD 7 mm in diameter, and situs inversus totalis. She was discharged 35 days after birth with a scheduled operation. The second baby was female and weighed 1,930 grams, with one- and five-minute Apgar scores of 7 and 9, respectively. Postnatal echocardiography revealed only patent ductus arteriosus without VSD.

Discussion

MCMA twins are well known to be associated with congenital abnormalities, preterm births, and perinatal death [1], but the evidence for antenatal counseling in cases of MCMA twin pregnancies is still insufficient [6]. Recently, research studies on MCMA twin pregnancies showed that about half of MCMA twins could reach 20 weeks of gestation and a decrease in mortality rate in MCMA twin pregnancies from 30–50% to 4–20% [7-10].

Chorionicity is the most important prognostic factor in multifetal gestations [6], so it should be confirmed in the first trimester using ultrasonography and recorded with a picture in the patient record [11]. However, diamniotic twins can become monoamniotic if the inter-twin membrane ruptures and can have prognostic progression similar to originally monoamniotic twins [12]. Therefore, the present authors suggest that the inter-twin membrane should be identified in each ultrasonographic examination in monochorionic twin pregnancies.

In the management of MCMA twin pregnancies, intensive fetal surveillance from 28 weeks onward and elective cesarean section at 32–34 weeks of gestation after antenatal corticosteroid administration have been recommended [4, 11]. Cord entanglement is often, even always [13], detected during closed fetal surveillance that could be associated with adverse perinatal outcomes such as acute fetal death [11, 13]. However, Rossi *et al.* found that ultrasonographic detection of cord entanglement could not improve perinatal outcome, and they suggested that prenatal management of MCMA twin pregnancies should focus on the detection of twin-to-twin transfusion syndrome, congenital anomaly, or assessment of preterm birth risks rather than the detection of cord entanglement [14].

The risk of congenital malformation is increased in multifetal gestations in comparison with singletons, and the risk increases in monochorionic twins. However, most studies about multifetal gestations did not include congenital anomalies. Therefore, the evidence for optimal delivery timing in multifetal gestations complicated with congenital malformations has remained unclear. The delivery indication of the present second case was not congenital heart disease but preterm labor with variable deceleration. Further studies about the optimal delivery timing in MCMA twins complicated with congenital abnormalities are needed.

References

[1] Post A., Heyborne K.: "Managing monoamniotic twin pregnancies". *Clin. Obstet. Gynecol.*, 2015, *31*, 643.
[2] Van Mieghem T., De Heus R., Lewi L., Klaritsch P., Kollmann M., Baud D., *et al.*: "Prenatal management of monoamniotic twin pregnancies". *Obstet. Gynecol.*, 2014, *124*, 498.
[3] Heyborne K.D., Porreco R.P., Garite T.J., Phair K., Abril D..: "Improved perinatal survival of monoamniotic twins with intensive inpatient monitoring". *Am. J. Obstet. Gynecol.*, 2005, *192*, 96.
[4] Desai N., Lewis D., Sunday S., Rochelson B.: "Current antenatal management of monoamniotic twins: a survey of maternal-fetal medicine specialists". *J. Matern. Fetal Neonatal Med.*, 2012, *25*, 1913.
[5] "Committee opinion No 652: Magnesium sulfate use in obstetrics". *Obstet. Gynecol.*, 2016, *127*, e52.

[6] Oepkes D., Sueters M.: "Antenatal fetal surveillance in multiple pregnancies". *Best Pract. Res. Clin. Obstet. Gynaecol.*, 2017, *38*, 59.
[7] Assuncao R.A., Liao A.W., Brizot Mde L., Krebs VL., Zugaib M.: "Perinatal outcome of twin pregnancies delivered in a teaching hospital". *Rev. Assoc. Med. Bras.*, 2010, *56*, 447.
[8] Ezra Y., Shveiky D., Ophir E., Nadjari M., Eisenberg VH., Samueloff A., *et al.*: "Intensive management and early delivery reduce antenatal mortality in monoamniotic twin pregnancies". *Acta Obstet. Gynecol. Scand.*, 2005, *84*, 432.
[9] Pasguini L., Wimalasundera R.C., Fichera A., Barigye O., Chappell L., Fisk N,M.: "High perinatal survival in monoamniotic twins managed by prophylactic sulindac, intensive ultrasound survelliance, and cesarean delivery at 32 weeks' gestation". *Ultrasound Obstet. Gynecol.*, 2006, *28*, 681.
[10] Prefumo F., Fichera A., Pagani G., Marella D., Valcamonico A., Frusca T.: "The natural history of monoamniotic twin pregnancies: a case series and systemic review of the literature". *Prenat. Diagn.*, 2015, *35*, 274.
[11] Dias T., Ladd S., Mahsud-Dorman S., Bhide A., Papageorghiou AT., Thilaganathan B.: "Systematic labeling of twin pregnancies on ultrasound". *Ultrasound Obstet. Gynecol.*, 2011, *38*, 130.
[12] Cunningham F.G., Leveno K.J., Bloom S.L., Spong. C., Dashe J.S., Ho B.L. (eds): "Williams Obstetrics" 24 ed. New York: McGraw-Hill Education, 2013, 901.
[13] Dias T., Mahsud-Dorman S., Bhide A., Papageorghiou A.T., Thilaganathan B..: "Cord entanglement and perinatal outcome in monoamniotic twin pregnancies". *Ultrasound Obstet. Gynecol.*, 2010, *35*, 201.
[14] Rossi A.C., Prefumo F.: "Impact of cord entanglement on perinatal outcome of monoamniotic twins: a systematic review of the literature". *Ultrasound Obstet. Gynecol*., 2013, *41*, 131.

Corresponding Author:
HYUN-HWA CHA, M.D.
Department of Obstetrics and Gynecology
Kyungpook National University Hospital
Kyungpook National University School of Medicine
130 Dongdeok-ro, Jung-gu, Daegu
700-721 (Republic of Korea)
e-mail: chh9861@knu.ac.kr