



Twin anemia polycythemia sequence in a dichorionic diamniotic pregnancy: a case report

Soo-Young Lee¹, Jin Young Bae², Seong Yeon Hong²

¹Department of Obstetrics and Gynecology, Yeungnam University Hospital, Daegu, Korea

²Department of Obstetrics and Gynecology, Daegu Catholic University School of Medicine, Daegu, Korea

Complications related to the vascular anastomosis of the placental vessels in monochorionic twins are fatal. The clinical syndromes of fetofetal transfusion include twin anemia polycythemia sequence (TAPS), twin-twin transfusion syndrome, and twin reversed arterial perfusion sequence. We present an extremely rare case of TAPS in a dichorionic diamniotic pregnancy. A 36-year-old woman, gravida 0, para 0, was referred to our hospital with suspected preterm premature membrane rupture. Although her pelvic examination did not reveal specific findings, the non-stress test result showed minimal variability in the first fetus and late deceleration in the second one. An emergency cesarean section was performed. The placenta was fused, and one portion of the placenta was pale, while the other portion was dark red. The hemoglobin level of the first fetus was 7.8 g/dL and that of the second one was 22.2 g/dL.

Keywords: Dichorionic diamniotic twins; Fetofetal transfusion; Twin anemia polycythemia sequence; Twin to twin transfusion syndrome

Introduction

In monochorionic twins, complications may occur because of vascular anastomosis, as the placenta is shared. An imbalanced blood supply due to vascular anastomosis can lead to fetofetal transfusion, increasing fetal morbidity. Clinical syndromes related to vascular anastomosis include twin anemia polycythemia sequence (TAPS), twin-twin transfusion syndrome (TTTS), and twin reversed arterial perfusion sequence. TTTS is a chronic form of fetofetal transfusion that affects approximately 9% of all monochorionic twins [1]. It occurs due to unidirectional flow through the arteriovenous anastomosis, creating an imbalance of blood volume between the donor and recipient twins. The donor twin is growth restricted and develops anemia, and the recipient twin becomes polycythemic and develops heart failure, resulting in fetal hydrops.

TAPS is diagnosed antenatally based on the middle cerebral artery-peak systolic velocity (MCA-PSV) when there is no amniotic fluid discordance. The term TAPS was first defined by Lopriore et al. [2] in 2007. It may occur spontaneously in up to 5% of all monochorionic twins as a result of incomplete laser treatment in TTTS cases [3]. We present a rare case of TAPS in a dichorionic pregnancy, of which only two cases have been reported so far.

Case

Ethical statements: This study was approved by the Institutional Review Board (IRB) of Yeungnam University Hospital (IRB No: 2021-04-042) and written informed consent from the patient was waived by IRB.

Received: March 17, 2021 • Revised: May 25, 2021 • Accepted: June 4, 2021

Corresponding author: Seong Yeon Hong, MD, PhD

Department of Obstetrics and Gynecology, Daegu Catholic University School of Medicine, 33 Duryugongwon-ro 17-gil, Nam-gu, Daegu 42472, Korea
Tel: +82-53-650-4078 • Fax: +82-53-650-4078 • E-mail: magu815@cu.ac.kr

Copyright © 2021 Yeungnam University College of Medicine, Yeungnam University Institute of Medical Science

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

A 36-year-old woman, gravida 0, para 0, was referred by a local clinic to the obstetric department following a preterm premature rupture of membranes at 34 1/7 weeks of gestation related to a dichorionic diamniotic pregnancy. She had a history of hypothyroidism, gestational diabetes, unexplained infertility, and laparoscopic left salpingectomy due to an ectopic pregnancy. She had conceived via *in vitro* fertilization. Although her pregnancy was uneventful, she was hospitalized for a week because of preterm labor at 33 weeks of gestation. She was transferred to the hospital under the suspicion of preterm premature rupture of membranes. Pelvic examination revealed no specific findings. The nitrazine and Amnisure ROM (rupture of membrane) test (QIAZEN, Hilden, Germany) results were negative. The non-stress test (NST) revealed minimal variability in the first fetus, but the heart rate of the second one showed late deceleration. An emergency cesarean section was performed immediately. During delivery, the donor twin was a 1,940-g male infant with Apgar scores of 2 and 6 at 1 and 5 minutes, respectively, and the recipient twin was a 2,360-g female infant with Apgar scores of 4 and 7 at 1 and 5 minutes, respectively. Birth weight discordance was 17.8%. The hemoglobin (Hb) count of the male infant was 7.8 g/dL, with a hematocrit level of 25.2%, whereas the female infant had a Hb count of 22.2 g/dL, with a hematocrit level of 70% on day 1, fulfilling the postnatal criteria of TAPS stage 3 (inter-twin Hb difference being 14.4 g/dL). After delivery, the donor twin was transfused with 20 mL of packed red blood cells, and the recipient twin received 28 mL of exchange transfusion with normal saline after phlebotomy. The twins were discharged from the hospital after conservative treatment without brain damage or hemorrhagic shock during postnatal care. Both twins were under follow-up without any complications, but congenital nephrotic syndrome developed in the recipient twin 3 months after birth.

The placenta was macroscopically fused. One portion of the placenta was pale, while the other was dark reddish due to congestion (Fig. 1). Due to an emergency cesarean section, the vascular connection of the placenta was not confirmed.

Discussion

In 2007, Lopriore et al. [2] reported a case of severe fetal or neonatal hematological complications due to chronic inter-twin transfusion without a twin oligo-polyhydramnios sequence (TOPS) sign, which was defined as TAPS. It was diagnosed when the MCA-PSV increased to > 1.5 multiples of the median (MoM) in one fetus and decreased below 1.0 MoM in the other twin following an antenatal Doppler examination. Only 40% to 63% of TAPS cases are diagnosed antenatally [4,5]. Therefore, postnatal diagnostic criteria were proposed. These criteria are fulfilled when the difference

in Hb count between the twins is > 8.0 g/dL and the reticulocyte count ratio is > 1.7 , or when the placenta has only a small (diameter of < 1 mm) vascular anastomosis. Antenatal classification of TAPS categorizes this condition into five stages: stage 1, MCA-PSV > 1.5 MoM in the donor and MCA-PSV < 1.0 MoM in the recipient, without any other signs of fetal compromise; stage 2, MCA-PSV > 1.7 MoM in the donor and MCA-PSV < 0.8 MoM in the recipient, without any other signs of fetal compromise; stage 3, as stage 1 or 2, with cardiac compromise in the donor twin that is defined as a critically abnormal flow (absent or reversed end-diastolic flow in the umbilical artery, pulsatile flow in the umbilical vein, and increased pulsatility index or reversed flow in ductus venosus); stage 4, hydrops of donor twin; and stage 5, intrauterine demise of one or both fetuses preceded by TAPS [5]. The postnatal classification categorizes TAPS into five stages based on the inter-twin Hb difference as follows: stage 1, > 8.0 g/dL; stage 2, > 11.0 g/dL; stage 3, > 14.0 g/dL; stage 4, > 17.0 g/dL; and stage 5, > 20.0 g/dL [5].



Fig. 1. Gross finding of the placenta of twin anemia polycythemia sequence in a dichorionic diamniotic pregnancy. The maternal surface shows the pale placental share of the donor twin (arrow) and the plethoric share of the recipient twin (arrowhead).

Theoretically, TAPS and TTTS do not occur in dichorionic twins; however, two cases of TAPS have been reported to date [6,7]. Besides, our study has some limitations. As emergency cesarean section was immediately decided after NST monitoring, MCA-PSV could not be measured; however, the difference in the single deepest pocket between the two fetuses was insignificant. The patient underwent antenatal care at a local clinic where complications related to pregnancy or signs of TOPS were not observed. In addition, because the genders of the two fetuses were different, TTTS and TAPS were excluded. However, such a mistake occurred because it was overlooked that TTTS and TAPS might also occur in dichorionic twins. Therefore, a biopsy was not performed to confirm placental vascular anastomosis. In this case, a differential diagnosis between TTTS and TAPS was necessary. TTTS was excluded because clinical signs of acute perinatal blood loss were observed in the donor infant whereas TOPS did not occur during antenatal care.

As described above, TAPS can only be diagnosed antenatally with MCA-PSV. According to Movva and Rijhsinghani [8], heterogeneity in placental echogenicity is helpful for the early diagnosis and management of TAPS along with timely delivery.

In conclusion, although it is very rare, if there is a TOPS sign during antenatal care in dichorionic twins or if the fetal MCA-PSV is increased in dichorionic twins, it is important to be aware that TTTS or TAPS may also occur in dichorionic twins.

Notes

Conflicts of interest

No potential conflict of interest relevant to this article was reported.

Funding

None.

Author contributions

Conceptualization, Formal analysis, Supervision: JYB, SYH; Project administration: SYH; Writing-original draft: SYL; Writing-review & editing: SYL.

ORCID

Soo-Young Lee, <https://orcid.org/0000-0001-8672-0751>

Jin Young Bae, <https://orcid.org/0000-0002-9287-8210>

Seong Yeon Hong, <https://orcid.org/0000-0001-6496-0498>

References

- Lewi L, Jani J, Blickstein I, Huber A, Gucciardo L, Van Mieghem T, et al. The outcome of monochorionic diamniotic twin gestations in the era of invasive fetal therapy: a prospective cohort study. *Am J Obstet Gynecol* 2008;199:514.e1–8.
- Lopriore E, Middeldorp JM, Oepkes D, Kanhai HH, Walther FJ, Vandenbussche FP. Twin anemia-polycythemia sequence in two monochorionic twin pairs without oligo-polyhydramnios sequence. *Placenta* 2007;28:47–51.
- de Villiers SF, Slaghekke F, Middeldorp JM, Walther FJ, Oepkes D, Lopriore E. Placental characteristics in monochorionic twins with spontaneous versus post-laser twin anemia-polycythemia sequence. *Placenta* 2013;34:456–9.
- Slaghekke F, Kist WJ, Oepkes D, Pasman SA, Middeldorp JM, Klumper FJ, et al. Twin anemia-polycythemia sequence: diagnostic criteria, classification, perinatal management and outcome. *Fetal Diagn Ther* 2010;27:181–90.
- Tollenaar LS, Slaghekke F, Middeldorp JM, Klumper FJ, Haak MC, Oepkes D, et al. Twin anemia polycythemia sequence: current views on pathogenesis, diagnostic criteria, perinatal management, and outcome. *Twin Res Hum Genet* 2016;19:222–33.
- Zilliox M, Koch A, Favre R, Sananes N. Unusual twin anemia-polycythemia sequence in a dichorionic diamniotic pregnancy. *J Gynecol Obstet Hum Reprod* 2019;48:359–61.
- Tollenaar LS, Prins SA, Beuger S, Slaghekke F, Oepkes D, Lopriore E. Twin anemia polycythemia sequence in a dichorionic twin pregnancy leading to severe cerebral injury in the recipient. *Fetal Diagn Ther* 2021;48:321–6.
- Movva VC, Rijhsinghani A. Discrepancy in placental echogenicity: a sign of twin anemia polycythemia sequence. *Prenat Diagn* 2014;34:809–11.