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Research Letter

Twin–twin transfusion syndrome in a monozygotic pregnancy with obvious lambda sign in first-trimester ultrasound

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Dear Editor,

A 30-year-old woman, gravida 2, para 1, was referred to our institution at 23 weeks' gestation after receiving a diagnosis of threatened premature labor and twin–twin transfusion syndrome (TTTS). Prenatal ultrasonography performed at 6 and 13 weeks of gestation at the referral hospital revealed thick dividing membranes called the "lambda sign." This finding suggests a dichorionic (DC) twin pregnancy (Figure 1). Initial ultrasonography in our institution revealed polyhydramnios (maximum vertical depth, 14.4 cm) in one fetus and oligohydramnios (maximum vertical depth, 0 cm) in the other fetus. The fetus with oligohydramnios showed a collapsed bladder. Doppler measurements of the umbilical artery, middle cerebral artery, ductus venosus, and umbilical vein in each fetus did not show abnormal flow patterns. TTTS Stage II was suspected according to these results. Fetoscopic laser photocoagulation (FLP) of placental vascular anastomoses was considered a therapeutic option for the condition at that time but was not performed because of the unknown effectiveness of FLP for TTTS in DC twin pregnancies.

After written informed consent was obtained from the patient and her family, serial amnioreduction was performed. In addition, nifedipine was administered as a tocolytic agent to control uterine contraction. Amnioreduction was performed 13 times prior to the delivery. The ductus venosus of the fetus with polyhydramnios (suspected recipient twin) deteriorated, showing reversed end-diastolic flow at 31 weeks' gestation. As the fetal cardiac function was considered to have worsened, termination was determined. Emergency cesarean delivery was performed at 31 weeks' gestation because the TTTS had progressed to Stage III;. The first twin (polyhydramnios, recipient fetus) weighed 1332 g, whereas the second twin (oligohydramnios, donor twin) weighed 998 g. The Apgar scores at 1 and 5 minutes were respectively 5 and 7 for the first twin, and 1 and 5 for the second twin. The birth weights differed by 25%. Both infants required intubation, artificial ventilation, and surfactant treatment because of respiratory distress syndrome. The smaller infant developed renal failure and necrotizing enterocolitis, requiring peritoneal dialysis from the 4th day of life and surgical treatment at 24 days after birth. The neonatal course of the larger infant was uneventful.

On macroscopic pathological examination, the placenta was composed of two distinct placental masses conjoined together through a small placental lobule (Figures 2, 3A, and 3B). The insertion sites of the umbilical cords of the twins were located almost at the center of each placental lobe. Staining studies of the anastomoses revealed a single arterial–venous connection from the donor's (oligohydramnios) umbilical artery to the recipient's (polyhydramnios) umbilical vein (Figure 3B). It was preferred to categorize the placenta as monochorionic (MC) placenta on macroscopic findings. The histological examination results of the dividing membrane showed diamniotic and DC fetal membranes with a trophoblastic layer (Figure 4). Studies of DNA zygosity that were performed using DNA extracted from the neonatal blood samples from each twin indicated a 99.99% probability of monozygosity by using the short-tandem repeat method.

The present case of monozygotic twin pregnancy was finally diagnosed as a DC twin pregnancy complicated with TTTS, which was confirmed through histological examination of the dividing membrane and DNA analysis of neonatal blood samples. Although FLP is the first therapeutic option for typical TTTS complicating MC twins [1], it was not performed in this case because its effectiveness for TTTS in DC twin pregnancies is unclear. Hence, serial amnioreduction was performed instead for the recipient fetus with polyhydramnios to prevent preterm labor.

Accurate diagnosis of chorionicity was of major clinical importance because of the associated high risk of mortality and morbidity in MC twin pregnancies [2]. In MC twins, the prevalence of adverse perinatal outcomes, including a significant risk of low birth weight, preterm birth [3], and neurological morbidity, is higher than in DC

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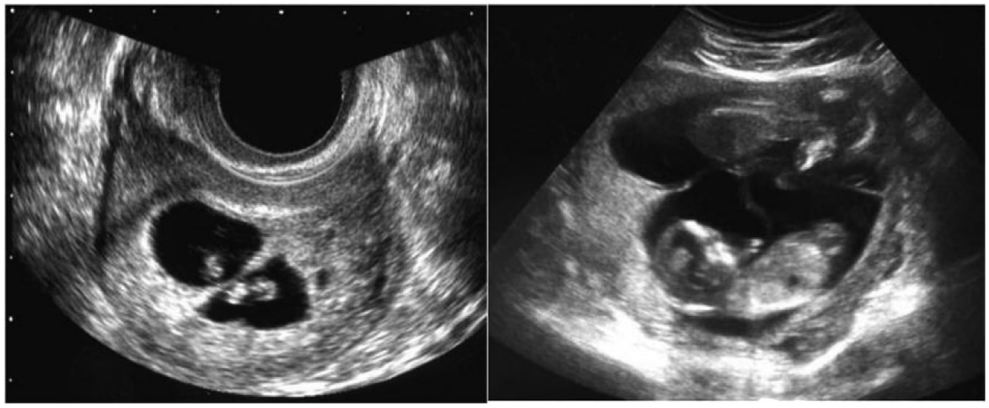

Figure 1. Lambda sign or twin peak sign in 6 weeks of gestation (left) and 13 weeks of gestation (right).

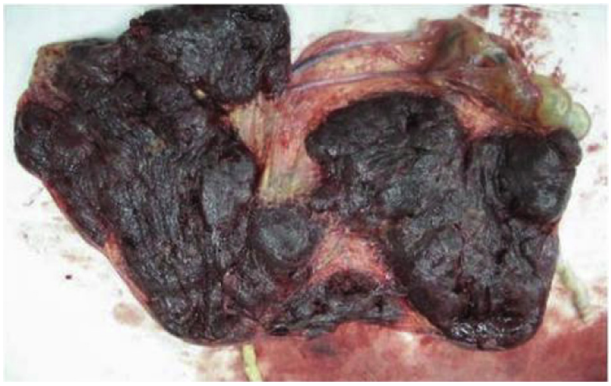

Figure 2. Two distinct placental masses.

twins [4]. Placental vascular anastomoses lead to the development of TTTS [5], selective intrauterine growth restriction [6], and intrauterine single fetal demise in MC twins. Dichorionicity and monochorionicity were distinguished because the chorionic membrane was considerably thicker than the amniotic membrane, and because the membrane is composed of two layers of chorion and two layers of amnion in DC twins and only two layers of amnion in MC twins [7]. Accurate diagnosis of fetal chorionicity is important in order to predict the prognoses of DC and MC twins in the first and second trimesters. With a cutoff dividing membrane thickness of 2 mm, the accuracy rates in predicting MC and DC twinning was 82% and 95%, respectively [8]. However, in the second and third trimesters, the prediction rate of DC twinning decreases because of the decline in membrane visualization during these periods [7]. Unfortunately, in this study, membrane thickness was not assessed because the intertwined membrane was not measured at 6 and 13 weeks' gestation.

This study has several limitations. In terms of determining the chorionicity of the twin pregnancy, only the dividing membrane thickness was used. Finberg [9] described the "twin peak sign" as an intertwin membrane producing a triangular projection of the placental tissue, extending between the layers. All of the 15 DC twin pregnancies in their study manifested this sign. During 10–14 weeks of gestation, sonographic determination of chorionicity was highly reliable because DC twins were easily distinguished according to the presence of the lambda sign [10]. Wood et al [11] described that ultrasonographic assessment of chorionicity using the "twin peak sign" or "lambda sign" had high sensitivity (94%) and specificity (88%), but its accuracy may not be sufficient to guide clinical management in all cases. If the lambda sign is observed on ultrasonographic examination in twin pregnancies, the fetuses should be diagnosed as DC twins [12]. However, Lopriore et al [13] concluded that two separate placental masses in twins were not by themselves a sign of dichorionicity and may occur in almost 3% of MC placentas. Owing to the lambda sign on the ultrasonograms obtained at 6 and 13 weeks' gestation (Figure 1), we inferred that our case was most likely a DC twin pregnancy. If the presenting typical TTTS in twin pregnancy is diagnosed as DC twinning in the first and second trimesters, the possibility of MC twinning should be considered.

Recently, two cases of monozygotic twinning in an intermediate mono-DC and monodiamniotic twin pregnancy were reported [14]. The first case was diagnosed as a partial MC twinning according to

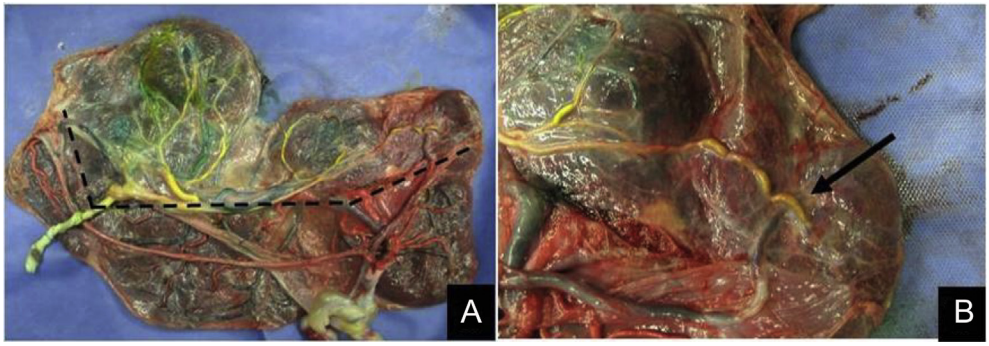

Figure 3. (A) Placental field occupied by donor fetus (oligohydramnios; above dashed line) and recipient fetus (polyhydramnios; under the dashed line) by function. (B) A single arterial–venous connection (arrow) from donor's umbilical artery to recipient's umbilical vein by color dye injection.

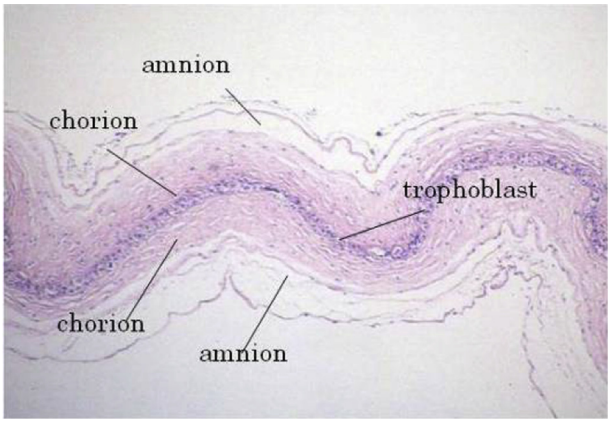

Figure 4. Microscopic findings of the dividing membrane confirmed two amnion layers, two chorionic layers, and trophoblastic layer. Hematoxylin and eosin, ×40.

the imaging finding at the first trimester but was finally diagnosed as a partial MC and DC septa according to a pathological examination. Meanwhile, the second twin was diagnosed with typical MC and monoamniotic twinning according to the imaging finding at the first trimester but was finally diagnosed as partial monoamniotic twinning with a partial intertwin septa. Furthermore, Quintero et al [15] reported a case of a monozygotic and DC-diamniotic pregnancy with placental vascular anastomoses complicated by TTTS Stage I that was successfully treated with FLP. This twin case was diagnosed with dichorionicity according to the presence of a "lambda sign" on preoperative ultrasonography. TTTS should be described as a typical condition in MC gestations, but the possibility of its occurrence in DC pregnancies should also be considered.

In our case, whether the histological specimens were indicative of DC or MC twinning remains unclear because only one part of the dividing membranes was used in the microscopic examination. A folding error of the dividing membrane that was fixed only for a part of the specimen for histological examination had an influence on a different diagnosis such as partial-DC twin pregnancy. According to the placental examination of single placenta with a narrow placental cotyledon between the two fetal territories, it would form a lambda sign diagnosed as DC twin by ultrasound scanning.

In conclusion, dichorionicity and monochorionicity were not completely distinguished based on the ultrasonographic examinations and course of the pregnancy. Our case was finally diagnosed as a monozygotic DC twin pregnancy according to the ultrasonographic findings at the first trimester and a postpartum histological examination. We recognized that complete detection of DC and MC twinning was impossible in the first and second trimesters, and the possibility of TTTS in DC and MC twin pregnancies.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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