Case report

Dichorionic pregnancy complicated by a twin-to-twin transfusion syndrome

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SUMMARY

Twin-to-twin transfusion syndrome (TTTS) is a serious complication typical of monochorionic pregnancies. Cases in dichorionic pregnancies have been described. The authors present a case report of a presumed TTTS in a dichorionic pregnancy followed at our institution in which the patient presented to the emergency department at 31 4/7 weeks' gestation complaining of lack of fetal movements. Both fetuses did not show any cardiac activity. On the first twin, an oligohydramnios was present and his bladder was not visualised and, on the second twin, a polyhydramnios and an augmented bladder were visualised. Pathological findings of the fetuses were compatible with a diagnosis of TTTS and placental study confirmed the presence of a dichorionic/diamniotic placenta, with superficial vessels crossing the dividing membrane.

BACKGROUND

Twin-to-twin transfusion syndrome (TTTS) is a serious complication that affects 1 in 40 multiple gestations.¹ In monochorionic twin pregnancies, both fetuses share one placenta that possesses vascular anastomoses on the fetal surface that connect the blood circulation of both twins.² Vascular anastomoses between the two placentae in dichorionic pregnancies, although rare, have been demonstrated.³ Unbalanced flow can, potentially, lead to TTTS and, therefore, to the death of one or both twins. Although TTTS is a complication of monochorionic twins, cases in dichorionic pregnancies have been described.

The authors present a case report of a presumed TTTS in a dichorionic pregnancy.

CASE PRESENTATION

The patient was a 37-year-old woman, gravida 2, para 0 with a history of a previous first-trimester spontaneous miscarriage, referred to our institution at 12 4/7 weeks' gestation with a dichorionic/diamniotic pregnancy. She had an uneventful medical history except for the presence of smoking habits. First-trimester blood analysis showed an immunised status for rubella and not for toxoplasmosis and no previous contact with cytomegalovirus. Blood pressure and glycaemic control for gestational diabetes were normal. Ultrasound examinations were performed monthly. Ultrasound markers for aneuploidy were absent and no morphological anomalies were found. Both fetuses were male. At 28 weeks of gestation, they were growing in the 25th percentile and the amount of amniotic fluid for each was normal.

At 31 4/7 weeks' gestation, the patient presented to our emergency department complaining of lack of fetal movements. She did not complain of pain, vaginal bleeding or loss of amniotic fluid. On ultrasound examination, both fetuses did not show any cardiac activity. The first fetus had an oligohydramnios and his bladder was not visualised. On the second fetus, a polyhydramnios and an augmented bladder were visualised. The diagnoses of fetal death of both twins and of a possible TTTS were made. After induction of labour, both fetuses were delivered vaginally.

INVESTIGATIONS

Autopsy findings for fetus 1 were of a male fetus weighing 1400 g, pale, without any congenital anomalies and with signs of acute anoxia. Autopsy findings for fetus 2 were of a male fetus weighing 1750 g, plethoric, with signs of acute anoxia and without congenital anomalies. On this fetus, a distended bladder without any urethral anomalies was present.

Placenta was dichorionic/diamniotic with two layers of amnion and intervening chorion on the dividing membrane (figure 1). There were two different sides: the first one, which corresponded to fetus 1, was smaller, 18×10 cm, and had thin vessels on the fetal surface. The cord had a velamentous and furcate insertion on the dividing membrane. The second, with 18×17.5 cm, had tortuous and congestive vessels on the fetal surface, the cord had velamentous and furcate insertion on the free membrane and had a vessel with long trajectory without Wharton substance. On this side, there were two large superficial vessels from the cord of

Figure 1 Dividing membrane composed of two layers of chorion and two layers of amnion. Fused chorion (*).

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Figure 2 Dichorionic/diamniotic placenta: (1) placenta of fetus 1, (2) placenta of fetus 2; dividing membrane (yellow dashed line); vessels from umbilical cord of fetus 1 that anastomose with vessels of the placenta of fetus 2 (yellow arrows); vascular anastomoses between the placentae (black lines) and long vessel without Wharton substance (arrowhead).

fetus 1 which crossed the dividing membrane and anastomosed with vessels from the fetal surface of placenta 2 (figure 2). On the maternal surface, there was an area deprived of chorion near the dividing membrane (figure 3). On the histological study, there were some differences between the two placentae as the chorionic villi of second placenta were bigger and had oedematous stroma.

Zigosity determination was not possible due to technical limitations.

OUTCOME AND FOLLOW-UP

After this misfortune, the patient already had a second first-trimester miscarriage and a singleton pregnancy with the birth of a healthy baby.

DISCUSSION

TTTS is a known and serious complication of monochorionic twin pregnancies and arises from the presence of unbalanced vascular anastomoses between the circulation of each fetus leading to relative hypovolaemia in one twin and hypervolaemia on the other.⁴ The hypovolaemia enhances the release of

vasoactive mediators in the donor twin in an effort of restoring the intravascular volume. The upregulation of the renin-angiotensin system results in oliguria which then leads to oligohydramnios, anhydramnios and a 'stuck twin'.⁵ On the other hand, the hypervolaemia of the recipient twin results in a release of natriuretic peptides which leads to polyuria and then polyhydramnios.⁶ The continued presence of this unbalanced circulation eventually leads to hydrops fetalis and fetal death of one or both twins. In dichorionic twin pregnancies, the presence of vascular anastomoses has been recognised. Foschini *et al*, for example, describe a case of a twin-to-twin cytomegalovirus infection through a dichorionic/diamniotic placenta.³ Nevertheless, typically this is not the case and, therefore, TTTS will not occur.

In our case, pathological studies confirmed the presence of a dichorionic/diamniotic pregnancy with the presence of large superficial anastomoses between the two placentae and one of vessels of the cord of fetus 1 crossing the dividing membrane and connect to the other placenta. The process leading to the growth of these anastomoses is not well understood. Quintero *et al* speculated that a monozygotic conception separated during the latter morula/blastocyst stage may not be complete and a tissue bridge may remain between the two blastocysts.⁷ When sufficient trophoblast layers are present, a dichorionic/diamniotic membrane is formed. The same authors also suggested that a failure of the mechanism that leads to the inhibition of angiogenesis in the marginal zone of the placenta could lead to the development of anastomoses in dichorionic placentae. Their study advocated that a reduction in the trophoblast in the chorion leave could also be sufficient for continued local angiogenesis and development of crossing vessels through the dividing membrane. In our case, microscopic studies showed the presence of a central area deprived of placental cotyledons (figure 3).

During our research, we only found two recent published cases of TTTS in dichorionic pregnancies.^{7,8} In our case, clinical, placental and autopsy findings were compatible with a diagnosis of TTTS. Both the dividing membrane and the T-section were studied confirming the presence of two layers of chorion and two layers of amnion. This helps exclude a different diagnosis such as partial-dichorionic twin pregnancy and surely is one of the strongest points in favour of our case. The discordance

Figure 3 Maternal surface showing an area deprived of chorion near the dividing membrane (*).

Patient's perspective

I understand that nothing could have been done differently to alter the unfortunate ending. I want my case to raise awareness amongst doctors that exceptional situations like mine can happen.

Learning points

- ▶ Twin-to-twin transfusion syndrome (TTTS) is a serious complication typical of monochorionic pregnancies.
- ▶ Although extremely rare, vascular anastomoses can occur between placentae in a dichorionic pregnancy and potentially lead to an unexpected TTTS.
- ▶ Clinicians should be alert of this possibility if a discordance on the amount of amniotic fluid is present.
- ▶ As with previous reports by other authors, this should lead the medical community to admit that TTTS, although typical of monochorionic pregnancies, can occur in dichorionic pregnancies.

between fetal weights and between the two bladders are also in favour of the diagnosis of TTTS. However, we do recognise that it is only a presumptive diagnosis.

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