CASE REPORT

A case of a surviving co-twin diagnosed with porencephaly and renal hypoplasia after a single intrauterine fetal death at 21 weeks of gestation in a monochorionic monoamniotic twin pregnancy

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Abstract

Monochorionic monoamniotic (MM) twin pregnancy carries a high risk of intrauterine fetal death (IUFD). Single IUFD in an MM twin pregnancy prior to 22 weeks of gestation has been reported to be strongly correlated with double twin demise. To our knowledge, there are no case reports on the natural course of a surviving co-twin in an MM twin pregnancy resulting in live birth after a single IUFD prior to 22 weeks of gestation. Here, we report a case of a surviving co-twin, after a single IUFD at 21 weeks of gestation in an MM twin pregnancy, with an antenatal diagnosis of renal hypoplasia and severe neurological damage leading to porencephaly, and live birth at 36 weeks of gestation.

INTRODUCTION

Monochorionic monoamniotic (MM) twin pregnancy is the least common type of twin pregnancy and carries a high risk of intrauterine fetal death (IUFD). Single IUFD in a monochorionic twin pregnancy is a devastating condition for the surviving co-twin due to acute fetal hypovolemia and anemia through blood transfusion from the surviving fetus into the demise fetus [1]. Previous studies have suggested that almost all single IUFDs that occur prior to 22 weeks of gestation in an MM twin pregnancy have been reported to result in double twin demise [2-4]. Here, we report on an extremely rare case of a surviving co-twin after single IUFD at 21 weeks of gestation due to cord entanglement in an MM twin pregnancy, with an antenatal diagnosis of renal hypoplasia and severe neurological damage, resulting in porencephaly and live birth.

CASE REPORT

A 33-year-old female, gravida 2, para 2, was referred to our hospital for the management of an MM twin pregnancy at 9 weeks of gestation. At 20 weeks of gestation, the entanglement of the umbilical cords was observed on ultrasound. At 21 weeks of gestation, IUFD of one fetus was confirmed and the surviving fetus displayed mild hydrops fetalis with the findings of subcutaneous edema and pleural effusion. Increased middle cerebral artery peak systolic velocity was observed (42 cm/s: >1.5 MoM), suggesting that acute anemic status caused by
blood transfusion from the surviving fetus to the demise fetus [5]. Serial counseling was provided for the mother and her family regarding the expected poor prognosis of the surviving co-twin and the possibility of severe neurological impairment and double IUFD; however, the mother chose to continue with the pregnancy.

At 23 weeks of gestation, the hydroptic changes of the surviving fetus disappeared. However, atrophy of cerebral parenchyma with an enlarged posterior horn of the lateral ventricle was observed. At 26 weeks of gestation, magnetic resonance imaging (MRI) of the fetus revealed severe ventriculomegaly, atrophy of the cerebral parenchyma and hypoplasia of the corpus callosum (Fig. 1). Additionally, at 26 weeks of gestation, oligohydramnios with amniotic fluid index (AFI; normal range: 5–25 cm) of 1.8 cm acutely emerged. At 30 weeks of gestation, bilateral renal hypoplasia became apparent [6] (Fig. 2). Interestingly, the volume of the amniotic fluid started to increase gradually from 28 weeks of gestation up to AFI of 15 cm at 34 weeks of gestation (Fig. 3). With regard to the fetal growth, the growth in the size of the head was almost arrested from 22 weeks of gestation; however, it started to increase again from 28 weeks of gestation (Fig. 4).

At 36 weeks and 0 days of gestation, the mother developed rupture of the membranes and vaginally delivered a male infant. The live male infant had a weight of 2775 g with an Apgar score of 7 and 8 at 1 and 5 min, respectively. Seven days after the birth, a head MRI revealed the characteristic findings of microcephaly and porencephaly (Fig. 5). Additionally, bilateral renal hypoplasia was also apparent [long axis length of right/left kidney: 18 mm/31 mm (normal range: 39–51 mm)] [7]. Two days after the birth, the serum creatinine level was 1.44 mg/dl; however, it spontaneously decreased to 0.80 mg/dl at 11 days after birth. The general condition of the infant was good, and the infant was discharged 12 days after the birth without any medication. The infant is currently under follow-up and has only slight rigidity in the limbs, predominantly on the left side, at 3 months of age (Fig. 5).

**DISCUSSION**

Previous reports have suggested that there is a high rate of neurological co-morbidities in the surviving co-twin after a single IUFD in a monochorionic twin pregnancy [8]. In our case,
the surviving fetus displayed not only neurological damage resulting in porencephaly, but also bilateral renal hypoplasia, which were speculated to be caused by acute detrimental circulatory changes on the brain and kidneys after single IUFD. To our knowledge, there have been no case reports that describe the antenatal course of a surviving co-twin after a single IUFD prior to 22 weeks of gestation in an MM twin pregnancy.

Genova et al. [9] reported that severe kidney failure was recognized in only 1 of 44 cases (2.3%) of single IUFD in monochorionic twin pregnancies. In our case, the surviving fetus exhibited signs of kidney failure from the severe oligohydramnios and renal hypoplasia seen on ultrasound. It is noteworthy that the volume of the amniotic fluid started to increase continuously from 28 weeks of gestation up to a normal range. One possible explanation for this change is that the fetal renal function might be gradually ameliorated because of the recovery from aberrant circulatory status of acute blood loss and hypovolemia as a result of single IUFD. Additionally, it is speculated that a certain number of nephrons were still preserved even if bilateral kidneys were morphologically impaired, which is supported by the fact that a blood creatinine level spontaneously decreased after birth. Our case provides valuable evidence that severe renal damage during the fetal period could recover, resulting in a live birth. However, it is difficult to predict the long-term prognosis given the short 3-month follow-up. Further close attention to renal function is necessary to assess the risk of chronic kidney disease later in life.

In conclusion, this is a case report on a surviving fetus with an extremely rare clinical course after a single IUFD in an MM twin pregnancy. The important aspect of this case is that a monoamniotic twin not only survived after early demise of its co-twin but also demonstrated recovery of renal function in the neonatal period despite the prenatal diagnosis of bilateral renal hypoplasia. We feel that this case report provides valuable information to enable both obstetricians and patients to make informed choice and furthers the understanding of the fetal pathophysiological changes after a single IUFD in an MM twin pregnancy.

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