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Twin anemia-polycythemia sequence: the importance of an accurate diagnosis

Abstract: Twin anemia-polycythemia sequence (TAPS), which was first described in 2007, is an atypical chronic form of twin-twin transfusion syndrome (TTTS). It presents as a large intertwin hemoglobin difference, with one twin developing anemia and the other polycythemia, without the traditional “polyhydramnios-oligohydramnios sequence” that is usually seen in TTTS. So far, ideal management of TAPS is not clear. We report here the case of a monochorionic biamniotic gestation, complicated by TAPS, detected at 30 weeks’ gestation that led to an emergent cesarean section. The recipient twin had an uneventful evolution. The donor twin was severely anemic, with a hemoglobin level of 2.5 g/dL that required immediate blood transfusion and intensive care management measures. Clinical evolution was favorable with good outcome in both twins. This case report intends to emphasize the association between hydrops fetalis and severe anemia, as well as the relevance of antenatal Doppler ultrasound investigations and the importance of an accurate management decision.

Keywords: Anemia; monochorionic gestation; polycythemia; twin-twin transfusion syndrome (TTTS).

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Introduction

Monochorionic multiple gestations may be complicated by twin-twin transfusion syndrome (TTTS). Virtually, all monochorionic twins develop vascular anastomosis, allowing the transfer of blood from one fetus to the other and vice versa; however, TTTS occurs in only 9–15% of them. This condition increases the risk of fetal/neonatal mortality and survivors are at a high risk of severe cardiac, neurological, and developmental disorders [6, 7].

Twin anemia-polycythemia sequence (TAPS) is an atypical chronic form of TTTS, first described in 2007, which presents as a large intertwin hemoglobin difference, with one twin developing anemia and the other polycythemia, without the traditional “polyhydramnios-oligohydramnios sequence” usually seen in TTTS [1, 6].

Twin anemia-polycythemia sequence results from slow intertwin blood transfusion through small caliber placental arteriovenous anastomosis, leading to large intertwin hemoglobin differences (superior to 8 g/dL) in the absence of amniotic fluid discordance [1, 4].

This slow transfusional process, with the passage of 5–15 mL of blood in 24 h, allows the development of hemodynamic compensation, which, in turn, avoids the development of polhydramnios [7].

This entity can occur spontaneously in up to 3–6% of previously uncomplicated third trimester monochorionic biamniotic twins, and may also develop after incomplete laser therapy of TTTS, in about 2–13% of cases [2, 6, 7].

According to the degree of severity, the outcome spectrum may vary from delivery of two healthy neonates to the death of both twins [6].

Twin anemia-polycythemia sequence can be diagnosed antenatally with Doppler studies and postnatally by hemoglobin and reticulocyte measurements [1, 4].

Case report

We report here the case of a monochorionic biamniotic gestation, with an unremarkable course until the 30th week, when, in a routine ultrasound (US), an asymmetry in the placenta echogenicity associated with hydrops of one of the fetus and paucity of fetal movements were
noticed. Umbilical cords were symmetric and there was no difference in the volume of amniotic fluid as well as in blade dimensions (Figure 1).

Under the suspicion of TTTS, an emergent cesarean section was performed at gestation 30+6. The recipient twin, with birth weight 1525 g, was plethoric, with a hemoglobin level of 19.3 g/dL (hematocrit 59.6%), and an Apgar score of 8/9/9 (Figure 2). She developed a slight acute respiratory distress, with hemodynamic stability. The remaining clinical evolution of this twin was uneventful, including the neurological examination and imaging.

The donor twin, with a birth weight of 1715 g and an Apgar score of 3/5/6, was severely anemic, with a hemoglobin level of 2.5 g/dL. She was hydropic, poorly perfused and severely hypotonic, with low hematocrit (9.5%), fetal heart rate of 173 bpm, hypertensive (46/28 mm Hg), and acidotic (pH 7.029; HCO$_3^-$ 13.9; base deficit of 14.3) (Figure 3). After resuscitation, she was ventilated and an urgent blood transfusion with 20 mL/kg of packed red cells was performed in 2 h, followed by intravenous furosemide. After the transfusion, arterial blood pressure became normalized and hemoglobin value raised to 12.6 g/dL (hematocrit 36.4%). Acidosis was corrected with sodium bicarbonate. Due to a respiratory distress syndrome, surfactant was administered. Chest X-ray revealed a right pleural effusion with marked edema of the thoracic wall and ascites. Therefore, in an attempt to improve the respiratory function, a paracentesis was performed, with drainage of a pale yellow fluid.

On day 2, she was on invasive ventilation, hemodynamically stable without inotropic support, with a small volume pericardial effusion in the echocardiogram, which resolved spontaneously. Apart from the initial hypotonia, the remaining neurological examination was unremarkable, with no seizures or other manifestations. The cerebral US showed a transient and small hemorrhagic focus on the germinal matrix, which resolved spontaneously in subsequent examinations.

Histology of the placenta revealed a division into two unequal portions – the part corresponding to the recipient twin showing ectasia and congestion of blood vessels, with an umbilical cord diameter of 1.2 cm. The part corresponding to the donor twin was immature and edematous, with a pale and friable parenchyma, and an umbilical cord diameter of 1.5 cm (Figure 4). We have no data concerning vascular communications.

Both twins were discharged on day 38, the recipient twin weighing 2320 g and the donor twin 2040 g, with a normal neurological examination. Ophthalmologic and hearing screenings were unremarkable.

Both twins maintain follow-up and, at present (9 months of corrected age), both show a normal physical and psychomotor development.
Discussion

Twin anemia-polycythemia sequence may remain undetected during pregnancy, mainly in cases where Doppler measurements were not performed, such as the presented case. In fact, some studies have suggested that in monochorionic biamniotic gestations US examination with measurement of middle cerebral artery-peak systolic velocity in both fetuses should be performed every 2 weeks, even in the absence of intertwin discordance in amniotic fluid volume. Even knowing that the range of outcome in TAPS is wide and is probably due to the heterogeneity of this entity, close follow-up could indicate the best treatment option in each case, such as expectant management, labor induction, or fetoscopic laser coagulation of arterio-venous anonymosis [3, 7, 8].

In our case, antenatal Doppler US investigation was not performed, and the presence of hydrops fetalis in one of the fetuses, associated with an asymmetric placenta echogenicity in the US performed at gestation 30+6, raised the suspicion of anemia due to TTTS and led to an emergent cesarean section. The diagnosis of TAPS was suspected antenatally based on the presence of hydrops fetalis and confirmed postnatally based on the presence of an intertwin hemoglobin difference of 16.8 g/dL.

Polycythemia in the recipient twin is a known risk factor for fetal and placental thrombosis, which may lead to severe cerebral injury and fatal outcome [1, 4]. In the present case, the recipient’s hemoglobin level was not as high as it would have been expected, presumably due to a concomitant fetomaternal transfusion. In order to clarify this hypothesis, flow cytometry was performed on the mother’s blood revealing an estimated fetomaternal transfused volume of 20 mL, corresponding to 16.4% of the theoretical volemia of the newborn. However, we cannot assume that fetomaternal hemorrhage occurred solely between the recipient twin and the mother or also between the donor twin and the mother.

Several studies have been performed in reference centers in order to evaluate some aspects in monochorionic twin gestations complicated by TAPS. It has been shown that, in a selected population, neonatal mortality and morbidity rates are similar to control neonatal rates [5].

Neurological injuries may occur in TAPS, both fetuses being potentially affected: in the recipient twin neurological damage can be caused by polycythemia and venous stasis, and in the donor twin, anemia and hypotension may lead to neurological injury [4].

However, it seems that the cerebral US findings do not have a direct correlation with the future outcome and neurodevelopment. Therefore, it is extremely important to perform a close follow-up, not only to prevent the development of sequelae, but also to detect early manifestations of some deficits.

In this case, after a troubled beginning of life, the twins are growing well and seem to have a favorable psychomotor development.

With this case report we intend to emphasize the fact that severe anemia may present as hydrops fetalis in a donor baby, as well as on the importance of antenatal Doppler US investigations, which, if done accurately, may lead to an earlier diagnosis.

References


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