Acta Obstet Ginecol Port 2012;6(3):145-148

Case Report/Caso Clínico

Acute per-caesarean twin-to-twin transfusion syndrome or twin anemia-polycythemia sequence?

Sindrome de transfusão feto-fetal agudo per-cesariana ou sequência anemia-policitémia?

Rui Artur Coimbra*, Ana Patricia Domingues, Clara Morais***, Etelvina Fonseca***, Paulo Moura******

Obstetrics Department, Coimbra University Hospitals

ABSTRACT

Monochorionic twin pregnancies present a significantly high rate of fetal mortality and morbidity, in part due to the twin-to-twin transfusion syndrome (TTTS) and its consequences. TTTS typically occurs in the second trimester and generally develops chronically. The risk of TTTS is unpredictable and despite its classical chronic development, it can also occur in a sub-acute or even acute manner. In the literature, few cases describe an acute or sub-acute peri-partum TTS. The authors present one case of acute per-caesarean twin-to-twin transfusion syndrome that could also be classified as a twin anemia-polycythemia sequence (TAPS), a type of chronic twin-totwin transfusion associated with chronic anemia in the donor and polycythemia in the recipient, without twin oligo-polyhydramnios sequence (TOPS).

Keywords: Keywords: monochorionic twin pregnancy, twin-to-twin transfusion syndrome, acute per-partum syndrome, twin anemia-polycythemia sequence.

INTRODUCTION

The incidence of spontaneous multiple pregnancies is about 1%, being twin pregnancies in most of the cases; one third of these will have a monochorionic placentation1.
In general, the mortality of twins is 5-10 times greater than single pregnancies. Monochorionic twins are considered a high risk pregnancy due the higher incidence of fetal and perinatal morbidity and mortality. The rate of fetal loss in dichorionic twins is about 2% and in monochorionic it is about 10%[2]. That high mortality is related to TTTS, higher prematurity and congenital malformations[3].

TTTS is the result of vascular connections between placentas that allows the connections of both fetoplacental cir-

* Interno da Especialidade de Ginecologia/Obstetrícia, Serviço de Obstetrícia/Maternidade Dr. Daniel de Matos, H.U.C., Coimbra
** Assistente Hospitalar de Obstetrícia da Maternidade Dr. Daniel de Matos, H.U.C., Coimbra
*** Assistente Hospitalar Graduada de Obstetrícia da Maternidade Dr. Daniel de Matos, H.U.C., Coimbra
**** Professor Associado da Faculdade de Medicina de Coimbra e Chefe de Serviço de Obstetrícia/Maternidade Dr. Daniel de Matos, H.U.C., Coimbra

culations (this connections are found in 98% of monochorionic-diamniotic placentas). Therefore, unidirectional or unbalanced blood flow is necessary to explain the changes noted in TTTS[4]. These anastomoses may be arterial to venous (AV), arterial to arterial (AA), and venous to venous (VV). In fact, the majority of monochorionic-diamniotic placentas have AV connections and its presence does not necessarily results in TTTS[5]. The presence of AA anastomoses was associated with a lower likehood of TTTS[6]. Pathological studies have demonstrated that AV anastomoses are deep, but the vessels that feed them are invariably superficial[7]. In about 15-30% of the monochorionic twins, the flow imbalance in AV communications of one fetus, the donor, to the other, the receptor, will result in the TTTS[8]. The clinical case presented, at time of occurrence, was classified as a TTTS intra-partum, a very rare and unpredictable situation, even with a regular surveillance to delivery. With the recently described as twin anemia-polycythemia sequence, this earlier diagnose is contested and the value of accurate Doppler evaluations, namely the middle cerebral artery peak systolic velocity (MCA-PSV), assume another importance.

CASE REPORT

AMRG, 30 years, second time pregnant, with a previous spontaneous abortion at 6 weeks of gestation, iatrogenic hypothyroidism (thyroidectomy in 2008, for papilar carcinoma), had a spontaneous monochorionic diamniotic twin pregnancy diagnose at 12 weeks. The prenatal surveillance included a regular ultrasound (every 2 to 3 weeks) with normal fetal anomaly scan and echocardiography in both of fetus.

The patient was hospitalized at 31 weeks, for surveillance and fetal pulmonary maturation protocol, as some Doppler abnormalities were seen: Umbilical Artery (UA) Doppler of first fetus (F1) revealed resistance (IR) and pulsatibility index (IP) >P90 with diastolic flow present and without evidence of centralization – middle cerebral artery (MCA) Doppler was normal with a peak systolic velocity within normal range. Anatomy, growth and amniotic fluid were normal. She had hospital discharge 7 days latter with normalization of UA Doppler, but was readmitted at 34 weeks again with UA Doppler abnormality of F1 and fetal growth of both fetus on the 5th percentile. The scan performed at 35 weeks and 1 day revealed: an estimated fetal weights (EFW) of F1 is 1900 grams(gr) and F2 is 1650gr, amniotic fluids (AF) were normal and AU and MCA Doppler's evaluations were also normal. Two days later, F1 UA doppler revealed IR>p90 with diastolic flow present and peak systolic velocity of MCA normal for both fetus (MCA-PSV of F1=45,5cm/s(p10) and F2=65,0cm/s(p80)). Due to the gestational age it was decided to perform an elective caesarean, under epidural analgesia, with easy extraction of both fetus (with a time break of one minute). Two males were born, the first with 1760gr, Apgar index (AI) of 9/10/10 and pH artery/vein=7.15/7.20 and the second with 1710gr, AI of 9/10/10 and pH artery/vein=7.17/7.22. Macroscopically the placenta was monochorionic diamniotic, which was histological confirmed, but the vascular connections have not been evaluated.

Because one of the twins has born pale and the other plethoric, the pediatric team has picked blood from both umbilical cords.

Twin 1 presented a whiteness cutaneous-mucosa and a respiratory distress syndrome always without oxygen need. It had also an asymptomatic anemia, with arterial pressure and cardiac frequency normal: day 0 (D0) with erythrocytes-2.40x10^6/microL, haemoglobin (Hg)-9.1g/dL; hematocrit level (Hct) -27%, platelets - 302000 U/L and in D3 with Hg-10.0g/dL; Hct-29.1%, 320000 platelets. Other morbidities included ictericia/hyperbilirubinaemia without criteria for phototherapy. This neonate got medical discharge on D8.

Twin 2 had a plethoric aspect with a polycythemia and thrombocytopenia: D0 with erythrocytes-6.49x10^6/microL, Hg-25.3g/dL; Hct-71.4%, 121000 U/L platelets and D3 with Hg-26.7 g/dL; Hct-81.3%, 63000 platelets being held sanguine exchange (blood by fisiologic serum) without intercurrences. On D4 Hg was 21.7g/dL, Hct-61.4%, 68000 U/L platelets and on D7 the Hg value was 23.2 g/dL; Hct-67.9%, 111000 platelets. Ictericia/hyperbilirubinaemia with criteria for phototherapy also occurred, from D1 to D5. Clinical discharge took place on D8.

In Pediatric evaluation, neurodevelopment outcome at 6 and 12 months was normal for both children

DISCUSSION

The TTTS is defined by the presence of polihydramnios (maximum vertical pocket of amniotic fluid of >8cm) in one fetus and oligohydramnios (maximum vertical pocket of fluid of<2 cm) in another. The staging system proposed by Quintero defined 5 stages: stage I, with a oligo/polihydramnios sequence, the bladder of the donor twin was still visible, whereas in stage II, the bladder was not visualized but the doppler studies were normal; by stage III, there are doppler abnormalities; in stage IV, hydrops

is present; and in stage V, there is a demise of one or both twins[9]. Untreated TTTS that develops earlier than 26 weeks has a perinatal mortality rate of 90%[10].

It has been suggested that some features in first trimester scan will be able to predict TTTS in monochorionic twin pregnancies. These features included crown-rump-lenght (CRL) and nuchal translucency (NT) discrepancy (the prevalence of increased NT thickness in at least one of the fetuses that subsequently develop TTTS was described to be about 30%, compared to 10% of those that do not develop TTTS[4,11]); and abnormal Doppler flow velocity waveform in the ductus venoso (DV)[11].

In this particular case reported and in all performed ultrasounds there were no suggestive signs of TTTS. Even the last one, performed in the same day of the delivery, also didn´t revealed any suspicion. The short time between the last ultrasound evaluation and the caesarean (one hour) excludes any chronic or sub-acute TTTS. The caesarean held without incidents and fetal extractions were simple and fast. Immediately, the newborns were delivered into the care of the Pediatric team, who suspected of TTTS.

A twin anemia-polycythemia sequence (TAPS) is a newly described form of chronic twin-to-twin transfusion associated with chronic anemia in the donor and polycythemia in the recipient, without twin oligo-polyhydramnios sequence (TOPS)[12].

In present case we admit the possibility of a spontaneous TAPS (although it is more often described after laser surgery)[10]. Both forms are characterized by the presence of a large inter-twin hemoglobin difference at birth without signs of TOPS as seen injuries the typical form of chronic TTTS. Whereas iatrogenic TAPS occurs in up to 13% of cases after laser therapy[10], the spontaneous form seems to complicate 5–6% of monochorionic twin pregnancies[13,14].

TAPS can be diagnosed both pre and postnatally. The prenatal diagnostic criteria is based on the inter-twin discordance in Doppler ultrasound measurement of middle cerebral artery peak systolic velocity (MCA-PSV). As described by Mari et al., MCA-PSV measurement >1.5MoM in one twin suggests severe anemia[15,16] and a simultaneous decrease of the MCA-PSV in the co-twin suggests polycythemia[13]. The postnatal diagnosis is based on three criteria: marked intertwin hemoglobin differences at birth, reticulocytosis in the donor and placental injection examination showing very small superficial AV anastomoses[13,15]. The case reported occurred in 2009 and, although MCA was evaluated in all ultrasounds performed (regarding de IP, IR and S/D index), the PSV wasn´t described in all the reports, so it´s not possible to classify this case as a TAPS, remaining the doubt based on the post-natal observation.

The understanding of the pathogenesis of TAPS is still incomplete but it appears to be mediated by a few small AV anastomoses[15]. Chronic inter-twin transfusion through these few minuscule AV anastomoses may occur so slowly that compensatory hemodynamic mechanisms maintain normal blood volume in both fetuses. The placental vascular pattern may also be dynamic as spontaneous thrombosis or infarction may occur as well as revascularization[17,18].

The prenatal monitoring of MCDA twins should imperatively include the measurement of the MCA-PSV of both fetuses during each follow-up visit, even in all uncomplicated pregnancies with absence of inter-twin discordance in amniotic fluid volumes. Although, the sensibility and the specificity of this prenatal marker (MCA-PSV) haven´t yet been demonstrated.

The special interest this clinical case lies on the need of MCA-PSV Doppler evaluation for a differential diagnostic between the rare form of acute TTTS (that may occur in any gestational age and in a sudden form) and the recently described TAPS.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

REFERENCES

1. Nicolaides KH. In: The 11-13 weeks scan. Fetal Medicine Foundaction, 2004: 95-96.
2. Sebire NJ, Snijders RJM, Hughes K, Sepulveda W, Nicolaides KH. The hidden mortality of monochorionic twin pregnancies. BJOG 1997;104:1203–7.
3. Callen, PW. In: Ultrasonography in Obstetrics and Gynecology (5th edition). 2009: 271
4. Matias A, Ramalho C: Search for hemodynamic compromise at 11-14 weeks in monochorionic twin pregnancy: is abnormal flow in the ductus venosus predictive of twin-twin transfusion syndrome? J Matern Fetal Neonatal Med 18:79, 2005.
5. Callen, PW. In: Ultrasonography in Obstetrics and Gynecology (5th edition). 2009: 281
6. Sebire NJ, Souka A, Skentou H, Geerts L, Nicolaides KH. Early prediction of severe twin-to-twin transfusion syndrome. Hum Reprod 2000;15:2008–10.
7. Benirsche K, Kim CK. Multiple pregnancy. N Eng J Med 1973; 288: 1276-84.
8. Nicolaides KH. In: The 11-13 weeks scan. Fetal Medicine Foundaction, 2004: 102.
9. Quintero RA, Morales WJ, Allen MH, et al: staging of twin-twin transfusion syndrome. J Perinatol 19:550, 1999
10. Robyr R, Lewi L, Salomon LJ, Yamamoto M, Bernard JP, Deprest J, Ville Y. Prevalence and management of late fetal complications following successful selective laser coagulation of chorionic plate

anastomoses in twin-to-twin transfusion syndrome. Am J Obstet Gynecol. 2006; 194: 796-803.

11. Matias A, Montenegro N, Areias JC. Anticipating twin-twin transfusion syndrome in monochorionic twin pregnancy. Is there a role for nuchal translucency and ductus venosus blood flow evaluation at 11–14 weeks? Twin Res 2000;3:65–70.
12. Weingertner AS, Kohler A, Kohler M, Bouffet N, Hunsinger MC, Mager C, Hornecker F, Neumann M, Schmerber E, Tanghe M, Viville B, Favre R. Clinical and placental characteristics in four new cases of twin anemia-polycythemia sequence. Ultrasound Obstet Gynecol. 2010; 35; 490-4
13. Lopriore E, Oepkes D. Fetal and neonatal haematological complications in monochorionic twins. Semin Fetal Neonatal Med. 2008; 13: 231-8.
14. Lewi L, Jani J, Blickstein I, Huber A, Gucciardo L, Van Mieghem T, Doné E, Boes AS, Hecher K, Gratacós E, Lewi P, Deprest J. 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.
15. 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.
16. Mari G. Hanif F. Fetal Doppler: umbilical artery, middle cerebral artery, and venous system. Semin Perinatol 2008; 32:253-257
17. Lopriore E, Deprest J, Slaghekke F, Oepkes D, Middeldorp JM, Vandenbussche FP, Lewi L. Placental characteristics in monochorionic twins with and without twin anemia-polycythemia sequence. Obstet Gynecol. 2008; 112: 753-8.
18. Lopriore E, Hecher K, Vandenbussche FP, van den Wijngaard JP, Klumper FJ, Oepkes D. Fetoscopic laser treatment of twin-to-twin transfusion syndrome followed by severe twin anemia-polycythemia sequence with spontaneous resolution. Am J Obstet Gynecol. 2008; 198: e4-7..