Caso Clínico/Case Report

Megabexiga num contexto de síndrome de transfusão feto-fetal: um possível factor de confusão

Monochorionic twin pregnancy: a puzzling case of a megacystis in a twin-to-twin transfusion syndrome setting

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ABSTRACT

Monochorionic twin placentation occurs once in every 3000-4000 pregnancies and twin-to-twin transfusion syndrome (TTTS) is its main complication, expected in 4-17% of the cases. Twins are also at higher risk for birth defects. Fetal megacystis occurs more frequently in twins and may obscure the diagnosis of TTTS. We report the case of a monochorionic twin pregnancy with urethral obstruction in one twin, which eventually developed TTTS.

Keywords: megacystis; monochorionic twins; twin-to-twin transfusion syndrome

INTRODUCTION

Monochorionic twin placentation occurs in every 3000-4000 pregnancies and twin-to-twin transfusion syndrome (TTTS) is the main complication expected in 4-17% of the cases. The twins are at higher risk for birth defects[1]. Fetal megacystis at 11-13 weeks of gestation (longitudinal bladder diameter of 7 mm or more) is found in about 1 in 1500 pregnancies[2]. It occurs more frequently in twins[1,3] and may obscure the diagnosis of TTTS. The risk ratio of bladder or urethral obstruction in twins over singletons is 3.34 (95% confidence interval (CI), 1.20-9.27)[1]. In a case series with 335 consecutive pregnant women, with 15 twin pregnancies, megacystis was diagnosed significantly more in twins than in singletons (20% of twins vs 2% of singletons, p = 0,007)[3].

Only few reports describe megacystis in twins[4,5]. In a case series including 19 fetuses with megacystis in the first half of pregnancy, one of them was a fetus from a monochorionic twin pregnancy[4]. The diagnosis in that fetus was made at 16 weeks. The renal parenchyma was bright and no other associated findings were observed. Vesicoamniotic shunt was inserted at 18 weeks after vesicocentesis at 14 and 16 weeks. The fetus died in utero at 22 weeks. The pregnancy and postnatal outcome were uneventful for the surviving twin.

We describe a monochorionic twin pregnancy with urethral obstruction in one twin, which eventually developed a TTTS.

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CASE REPORT

A pregnant woman, 31 years old, IGOP, was referred at 12 weeks to our centre for first trimester ultrasound scan. A monochorionic diamniotic twin pregnancy was detected. Fetus 1 had a crown-rump length (CRL) of 59,5 mm, a nuchal translucency (NT) of 2,3 mm, an abnormal flow in the ductus venosus (DV) and a megacystis with a longitudinal diameter of 13,9 mm. Fetus 2 had a CRL of 57,8 mm, a NT of 1,4 mm and normal flow in the DV (Figure 1). A week later, the bladder measured 24,6 mm and at 14 weeks, the diagnosis of urethral obstruction was established due to the presence of the "keyhole sign". Signs of TTTS appeared at 17 weeks. The donor presented megacystis, renal hyperechogenecity and oligohydramnios. The recipient showed cardiomegaly, pleural effusion, polyhydramnios, inverted A wave in the DV and the bladder measured 14 mm. The patient was referred at 17 weeks to the Harris Birthright Centre for laser ablation of placental anastomosis. The receptor died a day after the procedure. The couple opted for termination of pregnancy of the ex-donor with the urethral obstruction (bladder of 42 mm). Karyotype for both fetuses was normal. The necropsic examination confirmed the prenatal findings. The donor (Figure 2) (fetus 1) presented megacystis, bilateral hydronephrosis and megaureter, and multifocal renal dysplasia. The recipient (fetus 2) showed cardiomegaly (double volume compared to fetus 1).

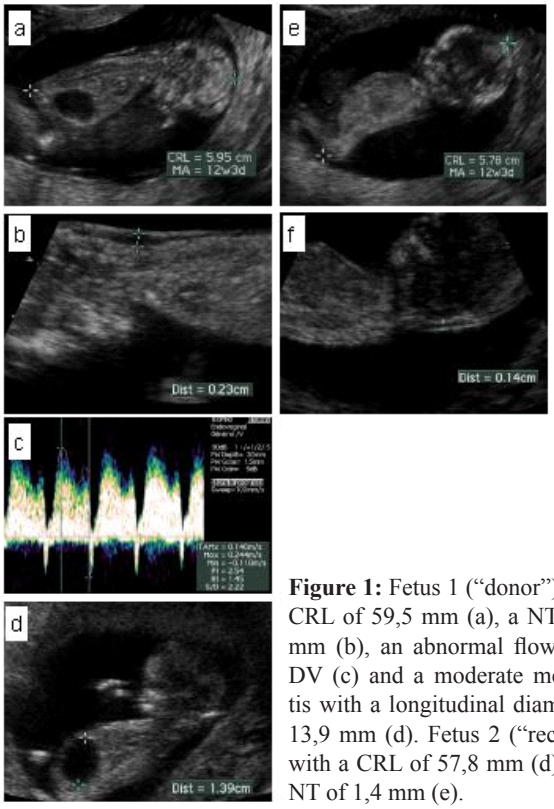

Figure 1: Fetus 1 ("donor") with a CRL of 59,5 mm (a), a NT of 2,3 mm (b), an abnormal flow in the DV (c) and a moderate megacystis with a longitudinal diameter of 13,9 mm (d). Fetus 2 ("receptor") with a CRL of 57,8 mm (d), and a NT of 1,4 mm (e).

Figure 2: Necropsic specimen of the donor at 17 weeks showing the megacystis.

DISCUSSION

This clinical case raised four important points:

1. Discrepancy for malformations between fetuses must be considered even in monozygotic twins[6], both in the type of malformation and its severity.
2. In monochorionic twins the diagnosis of megacystis in the first trimester should be careful and differentiated from distended bladder found in the recipient of a TTTS. In TTTS there is discordance in bladder size, with small or non-visible bladder in the donor and distended bladder in recipient.
3. Increased NT in monochorionic twins can be a predictor of TTTS [7]. In those who eventually developed TTTS, the fetus with the highest NT was the recipient. Increased NT can also be found in the donor if there is a coexisting malformation,

namely megacystis[2,8]. The underlying mechanism for the increased NT in fetal megacystis may be thoracic compression[9].

4. In all monochorionic twins the hemodynamic evaluation between 11-14 weeks should be performed by DV flowmetry to anticipate TTTS [10]. Increased NT and abnormal flow in the DV may be early manifestations of haemodynamic imbalance between donor and recipient[10].

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REFERENCES

1. Li SJ, Ford N, Meister K, Bodurtha J. Increase risk of birth defects among children from multiple births. Birth Defect Res A Clin Mol Teratol 2003; 67: 879-885
2. Sebire NJ, Von Kaisenberg C, Rubio C, Nicolaides KH. Fetal megacystis at 10-14 weeks of gestation. Ultrasound Obstet Gynecol 1996; 8: 387-390
3. Maymon R, Ben-Ami, Vaknin Z, Reish O, Herman A. Is there an increased rate of megacystis in twins? Ultrasound Obstet Gynecol 2005; 26: 197-199
4. Jouannic JM, Hyett JA, Pandya PP, Gulbis B, Rodeck CH, Jauniaux E. Perinatal outcome in fetus with megacystis in the first half of pregnancy. Prenat Diagn 2003; 23: 340-344
5. Hsu CD, Craig C, Pavlik J, Ninios A. Prenatal diagnosis of megacystis-microcolon-intestinal hypoperistalsis syndrome in one fetus of a twin pregnancy. Am J Perinatol 2003; 20: 215-218
6. Machin GA. Some causes of genotypic and phenotypic discordance in monozygotic twin pairs. Am J Med Genet 1996; 61: 306-307
7. Favre R, Kohler M, Gasser B, Muller, Nisand I. Early fetal megacystis between 11 and 15 weeks of gestation. Ultrasound Obstet Gynecol 1999 14: 402-406
8. Sebire NJ, D'Ercole C, Hughes K, Carvalho M, Nicolaides KH. Increased nuchal translucency thickness at 10-14 weeks of gestation as a predictor of severe twin-to-twin transfusion syndrome. Ultrasound Obstet Gynecol 1997; 10; 86-89
9. Liao AW, Sebire NJ, Geerts L, Cicero S, Nicolaides KH. Megacystis at 10-14 weeks of gestation: chromosomal defects and outcome according to bladder length. Ultrasound Obstet Gynecol 2003; 21: 338-341
10. Matias A, Ramalho C, Montenegro N. 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. 2005; 18: 79-86