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Case Report

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Rare case of twin to twin transfusion syndrome with posterior urethral valves

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Abstract

Twin to twin transfusion syndrome (TTTS) is a rare complication of monochorionic twin pregnancies. Posterior urethral valves (PUV) is the main cause of urethral obstruction found almost exclusively in male embryos. A 23 - year - old gravida 1 para 0 presented in our department with diamniotic monochorionic twins at 21 weeks of gestation for ultrasound B - scanning and TTTS accompanied with PUV was diagnosed. The first trimester screening test for nuchal translucency was normal. This case presentation can provide more information regarding the pathophysiology of TTTS.

Keywords: Twin to twin transfusion syndrome; posterior urethral valves; bladder obstruction; oligohydramnios; polyhydramnios

Twin to twin transfusion syndrome (TTTS) is a very rare complication occurring in monochorionic twins. The risk of TTTS development is estimated to be about 1 in 2,000 pregnancies and 10 - 15% of monochorionic twin pregnancies[1]. TTTS can be diagnosed by ultrasound scan. Two criteria are necessary for the diagnosis of TTTS: i) the presence of monochorionic twins, and ii) the existence of oligohydramnios in one sac and of polyhydramnios in the other sac[2]. It is believed that this complication occurs as a result of a non - equal exchange of nutrients, fluid and oxygen through the anastomoses of the placenta[3].

Five stages of TTTS have been proposed by Quintero et al revealed by ultrasound findings, including absence of the fetal donor bladder, abnormal umbilical artery Doppler for both embryos, fetal hydrops, and fetal death[4]. However, many other theories have been proposed, as the absence of differences in hemoglobin, erythropoietin and iron metabolism, the up - regulation of renin - angiotensin system in donor, the fetal renin - angiotensin, the decreased antidiuretic hormone in the recipient or the differences of leptin and insulin among the fetuses[5 - 7].

The pathophysiology of TTTS remains to be clarified despite the growing understanding and the evolution of ultrasound.

We present here a case of diamniotic monochorionic TTTS presented with polyhydramnios in the amniotic sac of one fetus and oligohydramnios in the amniotic sac of the other, and abnormal Doppler for both fetuses. Moreover, posterior urethral valves (PUV) were diagnosed in the donor fetus.

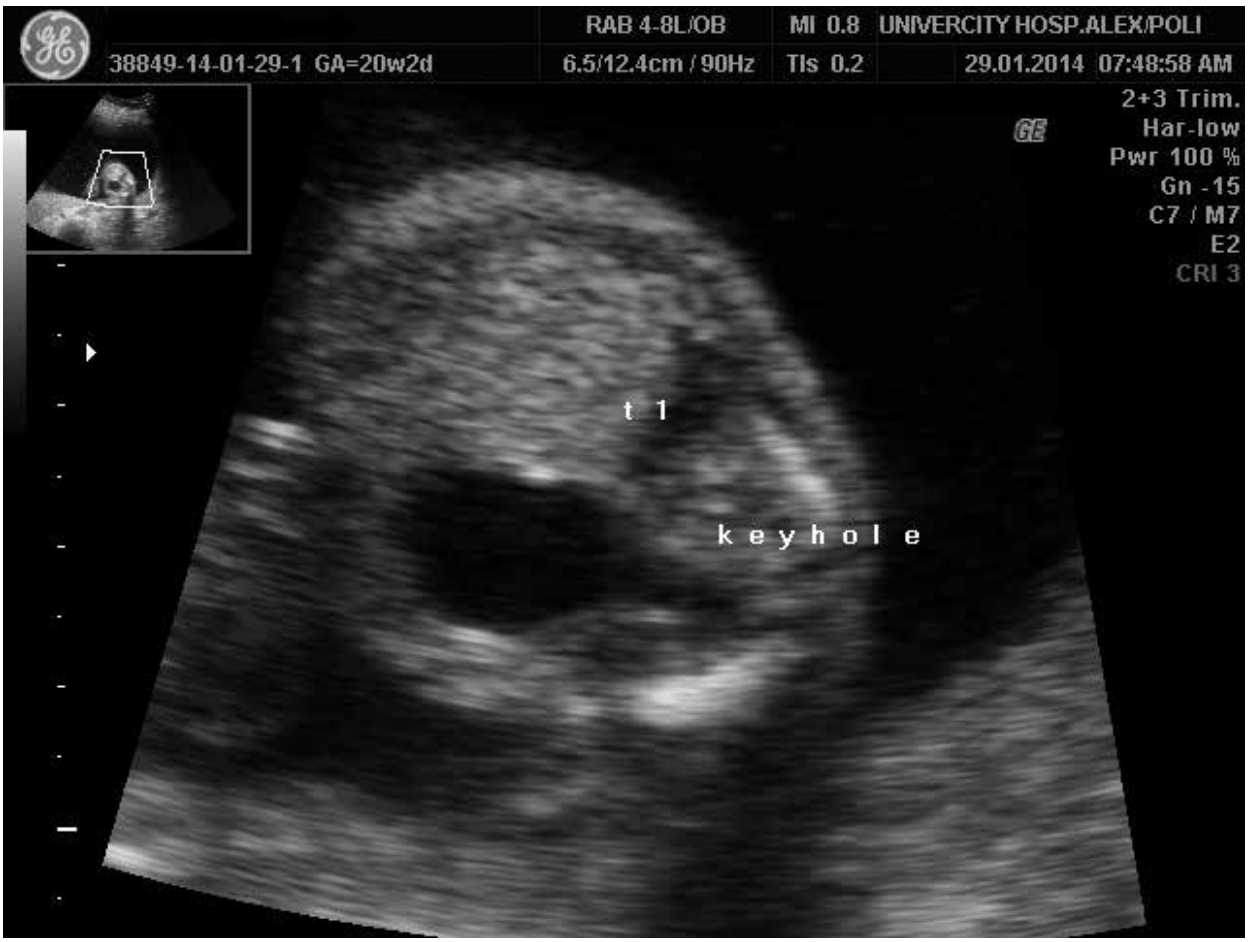

Figure 1: Posterior urethral valves (PUV) diagnosis presented in the recipient embryo: Fetal bladder with keyhole sign

Case report

A 23 - year - old woman gravida 1 para 0 referred to our department with diamniotic monochorionic twins at 21 weeks of gestation for ultrasound B-scanning. Her medical history was unremarkable. Ultrasound scan at 12 weeks of gestation revealed a twin gestation with a single placenta and no dividing membrane, indicating monochorionic diamniotic twins. Both twins were appropriate for gestational age, with absence of any disorders. The amniotic fluid volume appeared normal. Amniocentesis at 16 weeks of gestation revealed an heterozygous delta F 508 mutation, the most common mutation of cystic fibrosis. The ultrasound B - scanning revealed a polyhydramnios of twin A (donor) and oligohydramnios of twin B (recipient) and resulted in TTTS stage 3 diagnosis. Moreover, the examination revealed bladder outlet obstruction of twin A, with bilaterally dilated ureters and moderate hydronephrosis of both kidneys. Specifically, PUV were observed in the donor twin (Figure 1). Doppler findings were abnormal for both fetuses. Termination of the pregnancy was decided after consultation and informed written consent of the pregnant woman.

Discussion

We present a case of diamniotic monochorionic twins diagnosed with TTTS at 21 weeks of gestation. The syndrome was diagnosed by the presence of polyhydramnios of the one embryo and oligohydramnios of the other in the absence of other caus-

es. The case was a TTTS stage 3 assessed according to the criteria proposed by Quintero[4]. The importance in our case was that additionally the donor embryo was diagnosed with PUV. A presented keyhole sign in the donor's bladder suggested the presence of PUV after ultrasound examination. PUV is the most common cause of urethral obstruction in male infants occuring in 1: 5000 live births[8]. The urinary tract is the site with the most frequent congenital abnormalities in twin pregnancies leading to renal failure. The most common therapy for antenatal bladder outlet obstruction remains pregnancy termination, whereas early antenatal interventions are associated with high rates of fetal morbidity[9].

The survival rate in TTTS depends on the stage of diagnosis and it becomes poorer with the progression of stages. The detection of TTTS is reported in the literature to be between the 16 and the 28 weeks of gestation[3]. Even today, there is not a gold standard ultrasound monitoring for early diagnosis of the TTTS. In our case until the 16 week of gestation no signs of suspicious TTTS were detected. An ultrasound performed at 21 weeks of gestation revealed the signs of TTTS.

PUV have been associated with twin pregnancies[10]. However, to our knowledge our case is the first in the literature where PUV are detected in TTTS. Probably, the congenital abnormalities of the urinary tract are associated with the complications in amniotic fluid presented in the TTTS.

Our case may provide powerful insight into the pathophysiologic process of TTTS and the possible association with the PUV. Timely diagnosis of the TTTS is crucial to achieve less perinatal morbidity. We also suggest that in case of detection of PUV in a twin pregnancy, obstetricians should be suspicious for a possible TTTS and a close further ultrasound monitoring is needed.

Conflict of interest

All authors declare no conflict of interest.

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