Journal of Gynecology Obstetrics and Human Reproduction 48 (2019) 359–361

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Case Report

Unusual twin anemia-polycythemia sequence in a dichorionic diamniotic pregnancy

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ARTICLE INFO

Article history:
Received 14 November 2018
Received in revised form 12 February 2019
Accepted 19 February 2019
Available online 19 February 2019

Introduction

Placental vascular anastomoses, present in almost all monochorionic twin pregnancies, give rise to specific complications [1].

Twin Anemia Polycythemia Sequence (TAPS), described by Lopriore et al [2] in 2007 is a transfusional syndrome defined in the antenatal period by the presence of cerebral Doppler anomalies, with a middle cerebral artery (MCA) peak systolic velocity (PSV) greater than 1.5 MoM in the anemic twin, and PSV less than 1 MoM in the polycythemic twin [3,4]. More recently, Fishel-Bartal et al suggested that the antenatal diagnosis of TAPS should instead be based on a differential intertwin PSV value of more than 0.5 MoM [5]. In the post-natal period, TAPS is defined by an intertwin difference in the hemoglobin level of more than 8 g/dL, associated with a reticulocytosis ratio of more than 1.7 [5].

The placental angioarchitecture in TAPS is unique and specific, and is characterized by the presence of numerous very small diameter (< 1 mm) arteriovenous anastomoses [6]. Placental vascular anastomoses are present in almost all monochorionic twin pregnancies, but only extremely rarely in dichorionic pregnancies [7].

We report below an unusual case of TAPS complicating a dichorionic diamniotic pregnancy, secondary to a bulky velamentous arteriovenous anastomosis.

Case presentation

This was a 35-year-old patient, gravida 3 para 2, with a spontaneous dichorionic diamniotic pregnancy. The dichorionic nature of the pregnancy had been unequivocally established in the first trimester of pregnancy after demonstration of a lambda sign [8].

Clinical and monthly ultrasound monitoring were unremarkable until 31 weeks' gestation, with a final ultrasound one week prior to this date showing smooth growth patterns, adequate amniotic fluid and perfectly normal fetal dopplers.

Ultrasound scan then raised the suspicion of anemia in twin A (TA) in view of an increased middle cerebral artery (MCA) peak systolic velocity (PSV) of 72 cm/s (*i.e.* 1.71 MoM) and signs of cardiac decompensation (generalized fetal edema, significant subcutaneous edema and pleural effusion). Ultrasound scan of twin B (TB) was unremarkable aside from an abnormal cerebral Doppler with a MCA PSV of 17 cm/s (*i.e.* 0.40 MoM). The estimated intertwin weight difference was 15.9%. Furthermore, there was a marked difference in echogenicity between the two placentas: hypoechogenicity of the TA placenta and hyperechogenicity of the TB placenta (Fig. 1).

Ultrasound findings therefore supported the hypothesis of sudden-onset TAPS in a patient with a dichorionic diamniotic twin pregnancy. It may be noted that her parvovirus B19 serology suggested an old infection and there was no evidence of a feto-maternal allo-immunization issue. A Kleihauer test was performed at this time but there was no fetal red cells found in the maternal blood.

In view of the suspicion of anemia in TA, *in utero* transfusion was considered. Owing however to impairments of the fetal heart rate in TA, emergency cesarean section was performed for fetal salvage at 31 weeks.

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http://dx.doi.org/10.1016/j.jogoh.2019.02.005

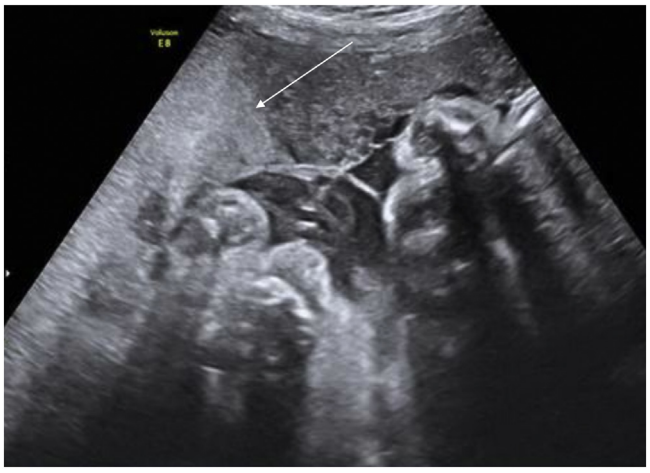

Fig. 1. Echographic image at 31 weeks: placental hypoechogenicity of twin TA thought to be anemic and hyperechogenicity of the TB placenta. The arrow indicates the interline between the 2 placentas.

Twin TA weighed 1725 g and TB 1554 g, which therefore represented a weight difference of 9.9%.

Twin TA showed extreme pallor of the skin and mucosae, generalized diffuse edema and severe signs of cardiorespiratory decompensation. The hemoglobin level was 2.4 g/dL with 103 100 reticulocytes per mm^3, *i.e.* a reticulocytosis rate of 17.5%. In spite of intensive care measures, the infant died at 24 h of life.

Twin TB presented erythrosis of the skin without any cardiorespiratory complications, suggesting a favorable neonatal development. The hemoglobin level was 20.9 g/dL, with 329 000 reticulocytes per mm^3, *i.e.* a reticulocytosis rate of 5.8%. This corresponds to an intertwin differential of 18.5 hemoglobin units and reticulocytosis ratio of 3.3.

Macroscopic examination of the placenta confirmed the dichorionic nature of the pregnancy by revealing 4 dividing membranes (Fig. 2). Moreover, there was a difference in the coloration of the maternal surface of the two placentas: the TA placenta was a pale pink whereas the TB placenta was bright red.

After injection of staining fluids into the vessels of the umbilical cord following Lopriore's method (arteries injected blue and veins

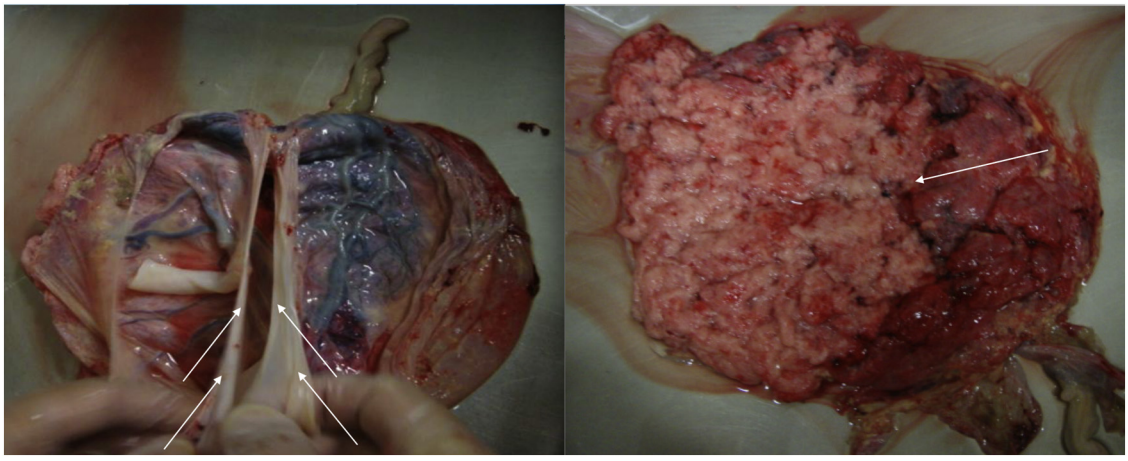

Fig. 2. Photographs of the placenta without preparation: on the left, fetal surfaces with the presence of 4 dividing membranes (indicated by 4 arrows); on the right, maternal surface showing a clear demarcation between the two chorionic plates (arrow) and difference in placental coloration.

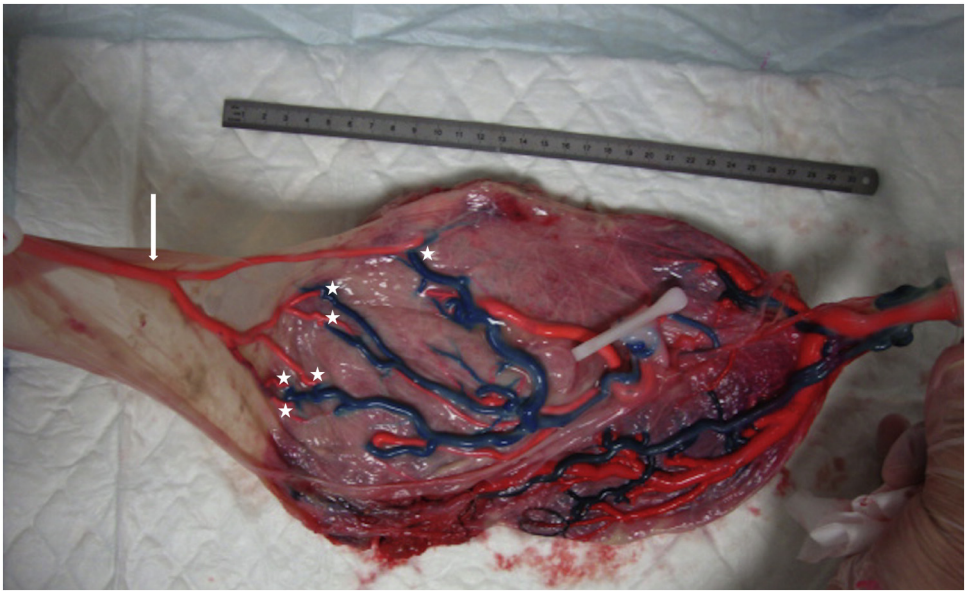

Fig. 3. Fetal placental surfaces after injection of staining fluids: injection of a blue stain into the umbilical artery and injection of a red stain into the umbilical vein of both placentas. The TA placenta (donor) is on the left, the TB placenta (recipient) on the right. The arrow indicates a bulky velamentous vein (6 mm diameter), draining numerous arteriovenous anastomoses indicated by asterisks (mean diameter measured at 3.5 mm).

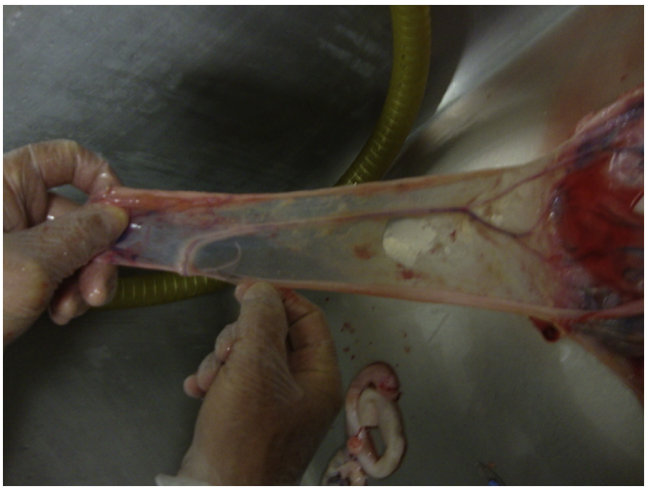

Fig. 4. Velamentous vein going away from TA (without injection).

injected red) (7), it was possible to identify several individual arteriovenous anastomoses, the mean diameter of which was 3.5 mm, draining into a bulky velamentous vein which measured more than 6 mm in diameter, and thus relayed the TA placenta (anemic) to the TB placenta (polycythemic) (Fig. 3). The Fig. 4 shows the velamentous vein going away from TA.

Discussion

In spite of the dichorionic nature of the pregnancy, the case reported here is clearly that of a TAPS based on the diagnostic criteria established by Lopriore et Slaghekke [3,10], corroborated by the PSV differential, difference in hemoglobin levels at birth and reticulocytosis ratio.

Two principal points are noteworthy. Firstly, to the best of our knowledge this is the first reported case of TAPS occurring in the context of a dichorionic diamniotic pregnancy. It is indeed rare to find vascular anastomoses between the two twins in a dichorionic pregnancy [9] and exceptional that these anastomoses can cause a hemodynamic disequilibrium between the twins. It should however be borne in mind that this event can happen and that a case of twin-twin transfusion syndrome has also been reported in a dichorionic pregnancy [11].

Furthermore, the placental architecture is extremely unusual for a TAPS: although arteriovenous anastomoses were detected they were of large diameter. In TAPS, arteriovenous anastomoses are invariably described as being of small diameter (< 1 mm), which is thought to allow slow transfusion of red cells from one twin to the other without the onset of an intertwin hemodynamic disequilibrium, contrary to the twin-twin transfusion syndrome [7]. Unfortunately, the placenta picture is of poor quality and cannot rule out minuscule anastomoses from twin A to twin B in the vascular equator area. The hypothesis of an acute transfusion of red cells, which would be fairly consistent with the clinical presentation and rather sudden onset of TAPS, is contradicted by the negative Kleihauer test and the difference in the reticulocytes rates of the twins at birth.

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