Vasa previa and postpartum hysterectomy in maternal Rh alloimunization

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Summary

Velamentous insertion of the cord, or vasa previa, is a malady where fetal vessels tranverse membranes ahead of the fetal part. The incidence of vasa previa is 1: 2000-3000 deliveries. Fetal mortality is over 50-75%. Early diagnosis is needed because these deliveries require emergency cesarean section; it is especially more common with placenta percreta, uterine atony and hemorrhage. Intravascular infusion of red blood cells (RBCs) into the fetus is one of the most successful means of in utero therapy for severe fetal anemia caused by RBC alloimmunization. We performed four fetal intrauterine intravascular transfusions (IVT) as therapy for severe fetal anemia. The patient underwent elective cesarean section. After delivery, profound uterine atony and vaginal hemorrhage were noted and the patient underwent hysterectomy. Pathological examination of the placenta and umbilical cord documented velamentous insertion of the cord. Before intrauterine IVT a detailed US examination is necessary to exclude vasa previa or placenta previa. Uterine atony may be result after a diagnosis of placenta previa or vasa previa. Intrauterine IVT is an irreplaceable diagnostic procedure in the treatment of severe fetal anemia

Key words: In-utero intravascular transfusion; Placenta previa completa; Vasa previa; Total abdominal hysterectomy.

Introduction

Velamentous insertion of the cord, or vasa previa, is a malady where fetal vessels transverse membranes ahead of the fetal part. The incidence of vasa previa is 1: 2000-3000 deliveries and it is more common in multiple gestations. Fetal mortality is over 50-75%. Early diagnosis is needed because these deliveries require emergency cesarean section, especially in cases with rupture of the membranes, which are complicated by signs of fetal distress due to hypoxia and bleeding. Maternal risk is small, especially if the velamentous insertion is associated with anomalous insertion of the placenta, e.g., percreta. Fetal exchange transfusion of about 250 ml has also been described [1]. The discovery of the rhesus (Rh) factor by Landsteiner and Wiener [2] in 1940 led to further findings of the condition by Levine and colleagues [3] who established that erythroblastosis fetalis was caused by immunization of an Rh negative mother by the red blood cells (RBC) from an Rh-positive fetus. A major cause of fetal or neonatal hemolytic disease is an incompatibility of the Rh blood group between the mother and fetus. The D antigen most commomly triggers hemolytic disease, although other Rh antigens, such as c, C, E, e can also cause problems. As a consequence of the hemolytic process, anemia, extramedullary hematopoesis and neonatal hyperbilirubinemia sometimes result in fetal loss or neonatal death or disability. In 1961, Liley [4] demonstrated the prognostic value of amniotic fluid spectrophotometry in identifying infants at risk and then showed intrauterine transfusions could prevent fetal death. Antenatal diagnostic methods identify fetuses at risk of developing hemolysis and assess disease severity in affected fetuses. Severely affected fetuses who in the past died before birth, secondary to severe anemia and hydrops, are now saved by antenatal monitoring and intrauterine transfusions.

The next major advance was in 1981 when Rodeck *et al.* [5] performed intravascular transfusion (IVT) by inserting the transfusion needle directly into a fetal vessel on the placental plate. The intravascular technique offers precise diagnostic evaluation of the fetal status and is effective, even in hydropic fetuses. The shorter procedure time associated with direct simple intrauterine IVT has made it the procedure of choice at most centers [6]. At the Institute of Gynecology and Obstetrics of Belgrade this procedure is generally timed, so that delivery can be carried out at about 36 or 37 weeks. Using this approach, neonatal survival in the nursery approaches 100% and long-term morbidity from prematurity is exceedingly low.

Previous cesarean section, velamentous umbilical cord insertion, an existing uterine scar and manual removal of the placenta increased frequencies of obstetric complications such as the most difficult postpartum uterine atony treated with total abdominal hysterectomy [7].

Case Report

A 32-year-old, gravida 2, was referred to our department at 26 weeks/3 days of gestation. Maternal blood type was A, Rh-negative anti D positive, with a titer 1: 128. Paternal blood type was A, Rh-positive. Weekly ultrasound (US) evaluation was important for early detection of fetal hydrops especially polyhydramnios as the first sign. Serial US and peak middle cerebral artery (MCA) velocities using Doppler advancements have indicated that IVT could prolong pregnancy and improve the

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outcome. Amniocentesis was performed to confirm fetal anemia. The amniotic fluid sample had on optical density (the Δ OD 450-0.250 reading) of 7.18 (lower portion of zone 2 of the Liley graph). The sample of amniotic fluid was not contaminated by meconium. These findings indicated the first intrauterine IVT fetal as therapy of fetal anemia. The mother was sedated by diazepam during the procedure. First US assessment of the placental position was completed. A 22 gauge spinal needle was inserted into the umbilical vein at the level of cord insertion into the placenta, under US guidance. The initial fetal hematocrit (Hct1) was 35%, the donor hematocrit (Hct2) was 81% and the final fetal hematocrit (Hct3) was 40%. The volume of infusion was 20 ml of freshly ultra packed O Rh-negative red blood cells. Fenoterol (Partusisten Boehringer Ingelheim) was administered via an infusion pump. The initial infusion was 0.5-1 μg/min, with the rate increased when necessary every 30-60 min to a maximum of 3.0 μg/min. Cephalexin (2 g per os every 12 hours) was started after the procedure until 27 weeks/3 days. At 28 weeks of gestation indirect antiglobulin (Coombs/test) indicated that her titer was 1: 128 repeatedly and 50 ml of ultra packed RBCs were intravascularly transfused (Hct1 = 24%, Hct2 = 82%, and Hct 3 = 40%) in the free umbilical cord loop, respectively. There were not any complications. US diagnosed anterior placenta previa and Doppler assessment of velocity in MCA diagnosed intermediate fetal anemia, but the biophysical profile determined fetal well-being. After four weeks her titer was 1: 256. The third IVT was performed and 82 ml ultra packed RBC was intravascularly transfused (Hct1 = 22% Hct2 = 82%, and Hct 3 = 42%) at the level of cord insertion into the placenta and umbilical vein. The non-stress test before and after the procedure was reactive.

In the workup no sign of hydrops or ascites was detected, but amniotic fluid was increased (amniotic fluid index = 20.5 cm) at the 35th week. Screening tests for gestational diabetes or TORCH were negative. The final IVT was done at the 35th week of gestation. Ultra packed RBC (70 ml) was transfused at the level of cord insertion into the placenta in the umbilical vein repeatedly, without any complications. Fetal Hct was 28% before IVT and at the end it was 39%. The procedure time was approximately 10 min for every intervention.

The patient's first pregnancy was cesarean section and finally at the 36th week of this pregnancy she underwent elective cesarean section. A male fetus weighing 3250 g was born with an Apgar score of 7 at the first minute and 8 at the fifth minute. The cord Hct level was 39%, direct bilirubin 120 mol/l and direct Coombs test was positive. He required two exchange blood transfusions and was discharged on the 17th day of life.

Four hours after delivery postpartal uterine atony and vaginal hemorrhage were diagnosed in the patient. Administration of oxytocin 5 IU IV slowly, 10 IU IM and F2α dinoprost trometamol 500 μg/ml IM every 15 min, with compression of the uterus was not successful. The patient was tachycardic and hypotensive (TA 90/60 mm Hg pulls > 100 beats/min), anemic (Er 2.06 Hgb 59 g/l Hct 18.4 Tr 77) and needed blood as quickly as possible (7 units). Administration of clotting factors should be based on coagulation results (aPTT 30.5 and PTT 54% fibrinogen 4.09 g/l and decreased values of clotting factors II, V, VII). The patient was treated by ten doses of cryoprecipitate used to replace fibrinogen and three units of fresh frozen plasma.

Finally, cesarean hysterectomy with ligation of the bilateral internal iliac arteries had to be considered. The pathological examinations of the placenta excluded placenta percreta, but documented a velamentous insertion of the umbilical cord.

Discussion

Previous cesarean section and manual removal of the placenta increase the frequency and size of fetomaternal transplacental hemorrhage, increasing the risk of immunization if the fetus is Rh positive. Amniocentesis for the determination of degree of fetal anemia or pulmonary maturity carries a 2% risk of immunization if performed under constant US guidance [1].

There is yet no agreement on the hematological criteria for IVT. In most fetal medicine units, as in our Institute, a Hct value between 25-30% or less is usually used as the indication for IVT [8]. The fall in Hct is rapid in fetuses with severe hemolytic disease, often necessitating a second transfusion within 7-14 days. The interval between subsequent transfusions usually is 21-28 days [9].

The intravascular method requires fewer procedural attempts, has fewer failures, results in better pregnancy outcomes and reduces the number of traumatic deaths. We performed all IVT via cordocentesis. From 1990, Plecas *et al.* performed 592 IVTs complicated by neonatal death in 9/161 (5.6%) versus 19/161 (19%) cases of fetal demise in utero. Four interventions of 592 intrauterine IVT were complicated by fallout of the needle, and in one case the IVT was complicated by tamponade of the umbilical cord and need for urgent cesarean section.

Our patient had not had any episodes of vaginal bleeding throughout the pregnancy.

Doppler and color Doppler with endovaginal imaging when necessary detected vasa previa in asymptomatic women as early as the second trimester. Other situations that might mimic vasa previa probably include a normal cord loop [10].

We use our complete protocol for treatment of uterine atony before hysterectomy. Placenta previa and vasa previa particularly in a patient with a previous uterine scar may be associated with uncontrollable hemorrhage at delivery and hysterectomy may be necessary. This is a momentous decision in an atonic uterus consuming clotting factor faster than can be transfused.

Intrauterine IVT can be considered a safe procedure in the hands of an experienced perinatologist with continuous US control of fetal well-being. It is a nessessary to review all relevant data (anamnestic and obstetric/related) in pregnancy to decide the time of delivery.

Although placenta previa and vasa previa may be independent etiological factors for uterine atony, we are not sure that repeated procedures such as IVT can be sufficient trauma for the uterus.

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