Reversible arterial redistribution in a fetus with true umbilical cord knot: case report and review of literature

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Abstract
Umbilical cord knot (UCK) affects around 1% of pregnancies and tightening of UCK is a very rare and highly unpredictable complication of pregnancy that can lead to fetal demise or neonatal death. The majority of authors agree that very little could be done to prevent fetal deaths in pregnancies with undiagnosed tight UCK. We herein report the case of a 39-year-old, gravida five, para three, pregnant woman at 40 weeks and five days of pregnancy, whose pregnancy evolved without complications and who was admitted to hospital for the management of the birth. Although the last ultrasound examination before birth showed a reversible arterial redistribution in the fetus dependent on the postural status of the pregnant women and other factors associated with umbilical cord knot were present, the diagnosis was missed because of the factors’ non-specificity. After a spontaneous labor without complications a dead male fetus, weight 3300 g, without heartbeat, Apgar score 0 was delivered. Macroscopic and microscopic findings confirmed that the cause of neonatal death was asphyxia caused by a tight UCK. The aim of our paper is to present the dramatic outcome of a pregnancy with a fetus with a tight umbilical cord knot (UCK), to bring to attention the signs that suggested the diagnosis, and to review the literature on this subject.

Keywords: reversible fetal arterial redistribution, true umbilical cord knot, fetal death.

Introduction
Umbilical cord knot (UCK) affects around 1% of pregnancies [1, 2]. Tightening of UCK is a very rare and very often unpredictable complication of the pregnancy that can occur both before birth or intrapartum leading to fetal demise [3] or neonatal death [4, 5]. The UCK is often diagnosed at birth and escapes frequently to antenatal diagnosis [4, 6, 7]. Some authors report cases of antepartum diagnosis of UCK [8–10] and analyze the antenatal signs that could suggest the presence of an UCK [11–13]. The best way to diagnose an UCK antenatally is the threedimensional color Doppler ultrasoundography [14–17]. Signs that could suggest the presence of a UCK are few and non-specific, such as: changes in recording the fetal movements [13], sinusoid heart rate pattern in non-stress cardiotocography (CTG) [11, 12], notchig of the umbilical artery waveform [18], intrauterine growth restriction [19] or fetal arterial redistribution [6]. Therefore, it is important to report cases and advise about the signs that are associated with a tight knot of the umbilical cord.

The aim of our paper is to present the dramatic outcome of a pregnancy with a fetus with a tight UCK, to bring to attention the signs that suggested the diagnosis, and to review the literature on this subject.

Case report
We herein present the case of a 39-year-old pregnant woman with gravidity five, parity three, 40 weeks and five days age of pregnancy that was admitted to hospital for the management of the birth. The patient was 162 cm height, 52 kg at the beginning of the pregnancy and had a weight gain of 12 kg until birth. The patient had two previous spontaneous abortions and previously delivered two girls each of 2900 g weight at birth, six and four years ago, respectively. We notice that the patient performed a genetic amniocentesis because of a high risk of trisomy 21 calculated in the first trimester of pregnancy. The fetal karyotype was normal 46,XY. No anomalies were found during the scan performed at 12 weeks and at 18 weeks of pregnancy. The course of pregnancy was without complications. The growth scan performed at the time of hospitalization showed a normal growth of the fetus (Figure 1) with a normal volume of amniotic fluid, with a grade three maturity placenta inserted on the posterior uterine wall. A Doppler evaluation of the arterial circulation of the fetus, with the pregnant woman in a posterior decubitus position, showed a normal umbilical artery flow, an increased pulsatility index (IP 1.84) at the level of the right middle cerebral artery with an elevated cerebro-placental ratio (2.92) (Figure 2). Normal values were found in the middle cerebral artery (IP 1.27) and cerebro-placental ratio index (2.02) after turning the pregnant women from dorsal to left lateral decubitus (Figure 2). The non-stress evaluation showed a normal profile. Because of inadequate cervical...
condition (cervical length 27 mm) and a Bishop score of 3, the induction of labor was postponed for another two days. Daily performed non-stress recording showed a normal profile. Two days after admittance and evaluation a spontaneous labor began. No significant CTG changes were found during the labor and complete dilatation was reached five hours after the beginning of labor. No medication for augmentation or inhibition of uterine contractions was necessary. The expulsion had a duration of 15 minutes. Intermittent heart tone auscultation was performed by a Doppler fetal heart rate monitor to prove the fetal condition. No changes were found until birth. Five to six minutes after the last fetal heart tone auscultation, a male fetus, weight 3300 g, without heartbeat, Apgar score 0 was delivered. Intensive care maneuvers were not successful in order to restore the viability of the newborn. A true tight knot was found situated near the placental end of the umbilical cord (Figure 3). Macroscopic and microscopic findings confirmed that the cause of neonatal death was asphyxia caused by a tight UCK. Macroscopic examination of the neonate showed no autolytic changes and no external or internal congenital malformation. An intense cyanotic color of the skin, congested internal organs (occurrence of edema at the level of the lung, liver, spleen and brain) and small hemorrhages were found at the level of the brain and suprarenal gland. The histological study performed in tissues obtained at the level of the kidney (Figure 4), lung (Figure 5), heart (Figure 6), liver (Figure 7), spleen (Figure 8), and brain (Figure 9) showed the occurrence of findings suggesting acute hypoxia.

Discussion

If an UCK is diagnosed before birth, the pregnancy must be monitored very carefully and cesarean section is the best way to deliver the fetus [8, 20, 21]. The fetus of pregnant women with a non-diagnosed UCK is in danger to die [3–5].

Only a few studies in PubMed present case reports of pregnancies with UCK [22] or perform an analysis of both the outcome of the cases with UCK [1, 2] or the main signs associated with UCK [23–26].

Figure 1 – Growth chart of the fetus with umbilical cord knot. The estimated weight expressed in grams (ordinate) and the gestational age expressed in weeks of gestation (abscissa) are represented at the time of prenatal visits (blue points) and birth (red point). The estimated weight of fetus fits in the normal nomogram of a eutrophic fetus.

Figure 2 – Reversible arterial redistribution in the fetal circulation. The cerebro-placental ratio (abscissa) and the gestational age expressed in weeks of gestation (ordinate) at the last prenatal visit before birth are represented with the pregnant women in dorsal (red point) and left lateral decubitus (blue point). A reversible elevation of the cerebro-placental ratio was found when the pregnant woman was explored in dorsal decubitus (red point).

Figure 3 – Umbilical cord knot. The true umbilical cord knot that became visible after the birth and caused the fetal asphyxia.

Figure 4 – Microscopic image in the asphyxiated neonate at the level of the kidney showed blood stasis in the vessels, tubular vacuolization and dilatation, glomerular dilatation. Hematoxylin–Eosin (HE) staining, ×200.
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The strangulation of the vessels of the umbilical could stop the materno-fetal blood circulation and cause different grades of fetal asphyxia [22]. A single experimental study analyzes the main factors that increase the risk of venous occlusion in a tied umbilical knot [23] and found that they are the decreased cord mass index, the decreased hydration index and the low venous flux capacity [23]. Because these risk factors are more frequent in fetuses with low body weight and intrauterine growth restriction, it is expected that these categories of fetuses be more exposed to the risk of strangulation of maternal–fetal circulation due to UCK [23].

Five studies from Räisänen et al. [1], Airas & Heinonen [2], Joura et al. [24], Blickstein et al. [25] and Hershkovitz et al. [26] analyze the maternal and fetal factors associated with UCK and found the following: multiparity, previous abortion, obesity, maternal anemia, diabetes mellitus, chronic hypertension, hydramnios, prolonged gravidity, long cord, patients who undergone genetic amniocentesis, and patients that carry a male gender fetus.

Seven studies analyze the obstetric outcome of pregnancies with UCK. The studies showed a four to eightfold higher risk of fetal death, 1.5 × higher risk of neonatal intensive care, 3.9 × higher risk of preterm birth, 3.1 × higher risk for small for gestational age (SGA) fetuses, a longer hospitalization stay and lower Apgar score at birth in fetuses with UCK compared to fetuses without UCK [1, 2, 24, 26–29].
The pregnancy presented in our report showed some of the antenatal risk factors of UCK, such as multiparity, prolonged pregnancy, genetic amniocentesis, previous spontaneous abortion, a male fetus, and reversible arterial redistribution in the fetus. Such arterial Doppler flow changes could appear in other conditions, too. Although described to appear in fetuses with UCK [30], their disappearance after turning the pregnant women to the left lateral decubitus position in the absence of pathological changes in the non-stress test suggested that the arterial Doppler changes are caused by a postural position of the pregnant women. Since all the signs described above were unspecific, the diagnosis of UCK was missed.

The pregnant woman was admitted to the hospital because of a late-term pregnancy to induce the labor. Since the Bishop score was unfavorable and the cervical length was 27 mm, the labor induction was postponed.

The surveillance of labor and expulsion was performed according to regular guidelines and the occurrence of an UCK after birth explained the fetal demise.

The dramatic outcome in our patient raises questions about legal issues regarding the surveillance and the guideline in such a case. Although the quality of ultrasound machines improved, the antenatal detection of fetuses with UCK is in our opinion complicated.

For third trimester pregnant women, whose fetus displays transient hemodynamic [30] or non-stress changes, we should consider the existence of a UCK. Even if a detailed ultrasound scan is performed, the diagnosis in the third trimester of pregnancy is very difficult because UCK could be undistinguishable from multiple loops of the umbilical cord. The best way to diagnose UCK remains the detailed ultrasound scan performed in the first or second trimester of pregnancy.

Conclusions

The majority of authors agree that very little could be done to prevent fetal deaths in pregnancies with undiagnosed tight UCK. If an UCK is diagnosed before birth an intensive surveillance of the fetus during pregnancy and a cesarean delivery are recommended. The prenatal signs that suggest an UCK are unspecific. We herein report the prenatal presence of a reversible redistribution of the arterial circulation, in a fetus that died at birth because of asphyxia caused by tight UCK. It is crucial to recognize signs that could suggest the presence of an UCK prior to birth, to try to diagnose this situation and to prevent a disastrous outcome.

Conflict of interests

The authors declare that they have no conflict of interests.

Ethical issue

We confirm that our study meets the ethical guidelines. Permission to perform this study was given by the Ethical Commission of the “Victor Babeş” University of Medicine and Pharmacy, Timișoara, Romania (Reference No. 14/27.06.2015). Informed consent was obtained from the patient.

References

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