True knot of the umbilical cord in advanced weeks of pregnancy: a case report

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Abstract

True knot of the umbilical cord is a very rare condition. Usually a true knot is noticed after delivery and does not lead to problems. It is relatively less common than the other umbilical cord anomalies. Most authors agree that there is little that can be done to prevent fetal death in undiagnosed true knot. In our case, the patient gave birth to a healthy male baby weighing 2700 grams by normal vaginal delivery after an ultrasound was suggestive of intrauterine growth restriction (IUGR). The baby's Apgar scores were 8 at one minute and 9 at five minutes. The umbilical cord measured 125 cm and contained two loose true knots. It was surprising that the baby could survive until term with these two knots and we hypothesize that this was the cause of the IUGR. Our case was interesting as neither neurologic sequela nor intrauterine death developed, despite two true umbilical cord knots.

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Introduction

True umbilical cord knots may occur in approximately 0.3-2% of all births.[1] The placenta is a discoid reddish-brown structure attached to the uterine wall that binds the fetus to the mother via the umbilical cord. A normal umbilical cord contains two arteries and one vein. Wharton's jelly is a special substance contained within the umbilical cord which serves to protect the vessels from trauma by acting as a cushion. A true umbilical knot develops when a fetus slips through a loop in the umbilical cord.

The most common symptom of a true knot is reduced fetal activity after 37 weeks. Complications may develop during delivery, although they do not increase the risk of fetal death. Prognosis may be very good with proper delivery, care and management. Risk factors for a true umbilical knot include:

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multiple pregnancy including twins, a long umbilical cord, fetuses that are small for their gestational age, polyhydramnios, and monozygotic twins.

Diagnosis of a true umbilical cord knot includes routine ultrasound examination during pregnancy, color Doppler ultrasound, examination of the placenta and umbilical cord during delivery. The best method for antenatal diagnosis of a true umbilical cord is three-dimensional Doppler ultrasound.[1,2]

Case

Our patient was a 24-year-old woman G2P1, at 39 weeks gestation. However, ultrasound measurements showed intrauterine growth restriction (IUGR) consistent with 36 weeks gestation. Additional ultrasound findings showed the umbilical artery Doppler (S/D) was 2.80, uterine artery Doppler (S/D) was 1.71, middle cerebral artery Doppler (S/D) was 4.21. She was hospitalized due to labor and a spontaneous vaginal delivery was planned. She had regular prenatal care with ultrasound examinations; however, an umbilical knot was not detected.

She gave birth to a healthy male baby weighing 2700 gram by normal vaginal delivery. The baby's Apgar scores were 8 at one minute and 9 at five minutes. The umbilical cord measured 125 cm and contained two loose true knots. (Figure 1) It was surprising that the baby could survive until term with these two knots, and we hypothesize that this was the cause of the IUGR.

Figure 1: Umbilical cord with two knots

Discussion

Wiedersberg et al.[3] describe the umbilical cord as the "life line of a fetus". Many abnormalities have been observed in morphology and pathology of umbilical cords.[4] Risk factors include relatively large uterine size and fluid volume (hydramnios, polyhydramnios), which may lead to a true umbilical knot resulting from excessive and exaggerated fetal movements. For example, prevalence of umbilical knot is high among patients with gestational diabetes that has led to hydramnios development. Moreover, grand multipara mothers, who have a loose abdominal and uterine wall, have sufficient space for movement of the fetus and true knot development. In addition, true umbilical knot is seen more frequently in patients who have undergone amniocentesis. A possible pathophysiology of this phenomenon is that, during amniocentesis, the uterus may contract and the fetus move more, contributing to knot formation.[5] Guzikowski et al. detected 10 true umbilical knots in 2864 deliveries (3.5%).[6] Most of the patients were

multiparous (80%). Four of the patients were diagnosed with knots in the prenatal period while the remaining three were not examined with ultrasound as knots were found while they were in the delivery process.[6]

Definitive diagnosis and ongoing monitoring of true umbilical knot may not only provide an explanation for UIGR but will also reduce abrupt and unexpected fetal distress.[6] A literature review suggests that a true umbilical knot does not have a specific ultrasound image.[7] However, the pairing of anomalous images with one or more other factors may suggest a need for weekly follow-up monitoring, especially when the patient is a grand multipara or when amniotic fluid volume is above normal. Gembruch et al. and Räisänen et al. believe that careful ultrasound and Doppler examination provide the best means of diagnosing umbilical cord complications.[5,8] True umbilical cord knots develop in 0.04-3% of all births, and have a perinatal morbidity rate of 11%.[1,9,10] Clearly color Doppler ultrasound and three-dimensional ultrasound play an important role in the diagnosis of true umbilical cord knots.[1,2,10,11]

Hershkovitz et al. found the rate of a true umbilical knot as 1.2% (841/69,139).[5] They also present demographic and clinical characteristics according to the presence or absence of a true knot. For example, women with fetuses that had a true umbilical cord knot were significantly older and had significantly more deliveries. In addition, the proportion of male newborns was found to be higher among the babies who had a true umbilical cord knot.[5]

Umbilical cord length may also be an important indicator of the possibility of a true umbilical knot. A normal mature cord length is 50-60 cm and 12 mm in diameter at birth. A long cord is defined as >100 cm while a short cord is defined as <30 cm. Cord complication rates related to operative intervention incidence, intrapartum complications, fetal heart rate and cardiac anomalies, and perinatal asphyxia are higher in cases of both long and short cord.[12]

In our case, cord length was 125 cm, the number of true knots was two and amniotic fluid volume was normal. The baby was healthy, and, fortunately hypoxia-related cerebral palsy did not develop. These risks should not be neglected. Furthermore, we believe that umbilical cord knot may be one of the causes of IUGR. Our case was interesting as neurologic sequela and intrauterine death did not develop despite the presence of two true knots.

Other indicators of potential true umbilical knot include the occurrence of IUGR or chronic hypertension, grand multiparity, gender, hydramnios and, history of amniocentesis. Prenatal diagnosis of a true knot in a fetus with IUGR has rarely been reported. Among these groups, true umbilical cord knot appeared to occur more frequently for male fetuses.[14] The relationship between a true umbilical knot and hydramnios may provide insight into the pathogenesis of umbilical knots and the increased morbidity and mortality in pregnancies with idiopathic hydramnios.

All pregnant women should be examined for pregnancy-induced hypertension (PIH), low fetal movement rate, gestational diabetes, prolonged

pregnancy, fetal hypoxia, and IUGR during their prenatal care. The fetus should be evaluated carefully with regard to fetal weight, placenta, insertion site of the umbilical cord, nuchal cord and true umbilical cord knots.

Conclusion

Accurate and timely diagnosis of a true umbilical knot may reduce abrupt and unpredictable fetal distress. It should be emphasized that a true knot does not have a specific ultrasonographic image associated with it. Moreover, this pathology may be diagnosed in the presence of either normal or increased amniotic fluid volume.

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