CASE REPORT

Two knots in an umbilical cord with seventy centimeter length: A case report

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Abstract

Our case was a newborn with two umbilical knots that delivered 39 weeks gastation and did not experience any developmental disruptions. The risk factors of fetus for true knot formation in the umbilical cord were gender and the umbilical cord size.

KEYWORDS

color Doppler ultrasonography, placenta, pregnancy, umbilical cord

1 | INTRODUCTION

The umbilical cord is a conduit between the developing embryo or fetus and the placenta. Any sort of disorder in the umbilical cord can be life-threatening to the fetus. Indeed, the presence of one single true knot in the naval string is a rare disorder occurring in roughly 0.3%-2% of pregnancies, and accordingly, the formation of two knots in a birth cord seems essentially impossible. While the presence of Wharton jelly surrounding the vessels reduces the probability of collapse of vessels and, consequently, loss of perfusion to fetus, true knots may tighten and decrease or even stop blood flow. Thus, infants with umbilical cord true knots often suffer extents of hypoxia in their prenatal course, which may cause fatal stress or even intrauterine death. 1,2 In fact, even the process of delivering these babies is precarious, and in case of fatal distress and severe hypoxia of the fetus, it can evolve from a normal delivery to a high-risk urgent cesarean section.^{2,3}

In this article, the case of a fetus is reported that despite two true knots in the 70-cm umbilical cord have an uneventful perinatal and postnatal course with no need to NICU admission. The Appar scores are 9 and 10 at minutes of 1 and 5, respectively. The mere risk factors of this fetus for true knot formation in the umbilical cord are the gender and the long size of the umbilical cord.

2 | CASE REPORT

The patient is a thirty-four-year-old pregnant $G_2L_1P_1$ woman with normal screening and no history of any disease. The patient's last menstrual period (LMP) was dated 20/11/2018 according to which the date of delivery was estimated at 27/8/2019. Due to her previous cesarean section, she is admitted in the 39th weeks gastation for a cesarean delivery. Nonstress test (NST) is normal, and cesarean is performed. The boy infant weighs 3300 g with Apgar scores of 9/10. The placenta and umbilical cord are removed. The placenta is normal, though the umbilical cord with the length of 70 cm has two loose knots. Regardless of this finding, interestingly, the infant does not require NICU admission (Figure 1A,B).

This is a report on a rare case of newborn with two umbilical knots that survived 39 weeks of gastation and did not experience any developmental disruptions.

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FIGURE 1 A, The newborn with two true umbilical cord knots. B, Removal of the two true umbilical cord knots from the surgery field

3 | DISCUSSION

In embryo, the umbilical cord is responsible for nourishing the fetus and also removing fetal waste. Therefore, in order to have a full-term and uneventful pregnancy that leads to the birth of a healthy baby, the umbilical cord must have both proper structure and function. The umbilical cord has two arteries and a vein and is normally about 50-60 cm long. 4 Cords longer than 100 centimeter and shorter than 30 cm are considered abnormally long and abnormally short, respectively.⁵ Generally, umbilical malformations cause various complications for both mother and fetus. Therefore, timely diagnosis is of vital importance in preventing dilemmas. The most common disorder that occurs for the umbilical cord is its prolapse, which can lead to stillbirth. An abnormally short or long cord is the second prevalent disorder. In fact, long cords increase the risk of cord entanglements, emergency deliveries, fatal placental vasculopathies, intrauterine fetal death, and neurological disorders, whereas short cords account for complications such as fetal malformations, fetal distress, and placental abruption.⁵ Other abnormalities such as a single-artery cord, cord cysts, and true and false cord knots are less common. Among these abnormalities, a cord true knot is very rare, with a probability of occurrence of approximately 0.3%-2%. This occurs when the fetus flips in the amniotic sac and slips through a loop in the umbilical cord. With this in mind, the possibility of formation of two true knots in the birth cord is roughly zero. In addition, even if it occurs, it is almost impossible for the embryo to survive without a developmental disruption or other disorders.^{1,7}

These true knots potentially prove fatal. More specifically, true knots can be loose and cause little disturbance to the blood supply to the fetus and only lead to SGA or IUGR, while tight true knots can cause abortion, IUFD intrapartum death, and stillbirth. Another possible complication resulting from these knots in the cord is fetal distress or abruption. In a study published in 2016, the prevalence of complications in 340 pregnancies with a knotted umbilical cord was investigated. It was observed that, in these cases, the prevalence of need for neonate intensive care unit (NICU) admission was 13.5%, of fatal death was 0.9%, of premature birth <37 weeks

was 8.6%, of low birth weight (BW) <2500 g was 6.5%, of small for gestational age (SGA) <90% was 12.1%, of minute 1 Apgar score <7 was 9.4%, and of minute 5 Apgar score <7 was 5.3%.8 These true knots might present no symptoms prior to complications and only decrease utero fetal movements at week 37, which is not very noticeable. Unfortunately, they cannot be diagnosed through screenings, and their antenatal identification is only accurate with color Doppler sonography of the umbilical artery, which is not routinely performed during pregnancies. Although the root cause of formation of these true knots has not yet been elucidated thus far, some risk factors have been suggested in various studies including male gender, abnormally long umbilical cord, excess of amniotic fluid (polyhydramnios), small infant, monoamniotic fluid, increased fetal movement, multiparity, maternal gestational diabetes mellitus(due to the possibility of hydroamnios), amniocentesis (due to more accelerated contractions of the uterus resulting in increased fetal movements), and loose abdominal wall and uterine that are common in grand multipara women.⁷⁻¹¹ Thus far, no clinical tests or laboratory data have been reported in the literature for the definitive diagnosis of a problematic umbilical cord. However, Marco Scioscia et al suggested 4-dimensional and color Doppler sonography as a diagnostic modality in this regard. 12

This case was a neonate who had an uneventful prenatal, perinatal, and postnatal course, despite having two true knots in the navel string. However, in two other similar cases, published in 1998¹³ and 2017, ¹⁴ both embryos experienced IUGR and the fact that their umbilical cords showed two true knots is believed to be the probable cause of this situation.

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CONFLICT OF INTEREST

There is no conflict of interest to declare.

Consent for Publication: Written informed consent was obtained from the mother for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

AUTHOR CONTRIBUTIONS

LH: involved in conception and design of the work. PD and FJ: involved in manuscript preparation and drafting the manuscript and searching the literature. LH and FJ: involved in critical revision of the manuscript for content and supervision. All authors read and approved the final manuscript.

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