A Rare Combination Causing Fetal Demise: Isolated Single Umbilical Artery and Long Umbilical Cord

Fetal Ölüme Neden olan Nadir Bir Kombinasyon: İzole Tek Umbilikal Arter ve Uzun Umbilikal Kord

Çoklu Kord Anomalileri / Multiple Cord Abnormalities

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Özet

Gerçek düğümler nadir kord anomalileridir, ancak kord kazalarının önde gelen nedenleridir. Tek umbilikal arter ve uzun umbilikal kord gerçek düğüm ve nukhal kord oluşmasına yatkınlık yaratmaktadırlar. Biz bu vakada fetal etkilenmeye neden olabilen diğer kord anomalileri ile tek umbilikal arter arasındaki ilişkiyi göstermeyi amaçladık. Bu raporda tek umbilikal arter ve diğer kord anomalilerinin eşlik ettiği, fetal ölümle sonuçlanan bir gebelik vakası sunulmuştur. Doğum sonrası umbilikal kord uzunluğu üzerindeki dört gerçek düğümle beraber 167 cm ölçülmüştür. Bu bulgular gerçek düğümlerin fetal ölüm konusunda ciddi bir risk faktörü olduğunu ve tek umbilikal arter ve uzun umbilikal kord ile ilişkili olabileceğini vurgulamıştır. Bu nedenle detaylı ultrasonda tek umbilikal arter bir kez saptandığında diğer kord anomalilerinin saptanması ve kord kazalarından sakınılabilmesi için inceleme genişletilmelidir.

Anahtar Kelimeler

Fetal Ölüm; Uzun Umbilikal Kord; Nukhal Kord; Tek Umbilikal Arter; Gerçek Düğüm

Abstract

True knots are rare cord abnormalities but they are leading cause of cord accidents. Single umbilical artery and long umbilical cords exhibit a tendency to form true knots and nuchal cords. We aimed to show the association between single umbilical artery and other umbilical cord abnormalities which may lead to fetal compromise as in this case. We presented a pregnant women with intrauterine fetal demise associated with isolated single umbilical artery and other cord abnormalities. After delivery, the length of umbilical cord was noted to be 167 cm with four true knots on it. Those findings once again emphasize that true knots are serious risk factors for fetal death and could be associated with single umbilical artery and long umbilical cord. Therefore once single umbilical artery is detected on detailed sonography, examination must be extended to rule out other cord abnormalities and avoid cord accidents.

Keywords

Fetal Demise; Long Umbilical Cord; Nuchal Cord; Single Umbilical Artery; True Knots

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Introduction

True knots and nuchal cords are the leading causes of cord accidents. The reported incidence of true knots range from 0.3 to 1.2%[1]. Umbilical cord abnormalities such as long umbilical cord (LUC) exhibit a tendency to form true knots and nuchal cords. There is also an association between LUC and non-reassuring fetal status, lower Apgar scores, and fetal demise[2]

Case Report

Thirty-one years old gravida 2 parity 1 woman admitted to our clinic with decreased fetal movements for several days at 38 weeks of gestation. Ultrasonographic examination revealed absent fetal cardiac activity with appropriate fetal measurements for its dates and normal amniotic fluid index. Antenatal follow up was unremarkable except isolated single umbilical artery without any associated malformations detected in second trimester detailed ultrasonography.

Her laboratory results were all normal and her vital signs were within normal limits. After detection of absent cardiac activity labor induction was initiated. She delivered vaginally a 3180 gr male fetus. Baby was macerated with an umbilical cord measuring167 cm (figure 1a). On gross examination the cord was edematous with one vein and one artery. There were also four true knots along the cord. Tightly coiled nuchal cord (three times) was also noted (figure 1b).

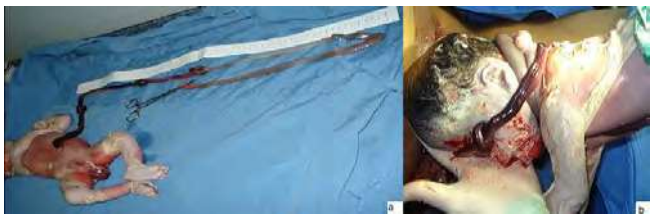


Figure 1. Macereted male fetus with long umbilical cord (167cm) and accompanying four true knots (A), Three times tightly coiled nuchal cords (B).

Discussion

Umbilical cord knots particularly true knots may represent a hazard to the fetus. Fetuses with true umbilical cord knots are more likely to have intrauterine growth restriction(IUGR) and have four fold increased risk of stillbirth, lower 1 minute Apgar scores, higher rate of meconium stained amnion and nonreassuring fetal heart rate patterns in antenatal period and also in the course of intrapartum period [1, 3, 4]. There are many reports demonstrating an association between cord complications and LUC particularly with true knots[5]. LUC also increases the risk of IUGR and associated with neonatal morbidities. Poor neurological outcome is another important complication of LUC which is related to cord complications or alteration of blood flow dynamics causing intrauterine hypoxia [2]. In a recent study by Rasionen male fetuses had longer umbilical cords than female fetuses. Moreover higher incidence of cord knots were observed in male gender. Also maternal age, multiparity, history of spontanous abortion, polihydramnios and diabetes mellitus were associated with cord knots [6].

Table 1 summarizes published reports regarding multiple cord anomalies and neonatal outcomes [7,8,9,10,11]. As seen, presence of LUC and SUA causes significant deterioration in heart rate patterns, an increase in rate of urgent cesarean sections and NICU admissions as well.

In the review of Baergen et al about placental pathologies SUA was more likely to be associated with LUC [2]. Hence association of LUC and SUA might increase the risk of a possible cord accident. SUA carries relatively normal blood volume with respect to three vessel cord but to compensate this alteration vascular resistance and compliance decrease and consequently such an alteration in vascular resistance make the cord vessel vulnerable to compression[12,13].As a result of an absence of a second protective umbilical artery, compression of single umbilical artery may affect fetoplacental circulation more severely than a normally structured umbilical cord . SUA was blamed on the pathophysiology of sirenomelia with the same reason like hypoperfusion [14]. Besides detailed fetal ultrasonography to rule out major malformations additional umbilical cord abnormalities such as true knots or nuchal cords should be scanned particularly in third trimester. Fetuses with more than one umbilical cord anomalies should be closely followed up.

There are only few studies regarding antenatal diagnosis and obstetrical outcomes of true knots. A significant number of ultrasonographic markers related to true knots have been defined such as 'four leaf clover' pattern, 'hanging nose' sign and 'bizarre multicolored' pattern[15,16]. But none of them has been shown to be specific [17]. However persistence of four leaf clover pattern all through the follow up examination should be use to support the diagnosis.

If a true knot is suspected in antenatal ultrasonography, close follow up with umbilical artery Doppler velocimetry studies until term is warranted[17]. If there is no deterioration in fetal blood flow patterns there is no indication for delivery[18]. But in case

Table 1. Published studies regarding multiple cord anomalies and neonatal outcomes.

Author	Maternal Age	Parity	Fetal gender	Cord Abnormality	IUGR/SGA	NICU admission	Mode of delivery	Heart Rate Pattern	Apgar1and 5 minute
Ugianskiene [7]	30	M	Female	SUA+LUC+4Knots	IUGR	+	Urgent C/S at 39w	diminished variability, absence of accelerations	5-10
Sirinivasan[8]	40	M	Male	LUC+4Knots	SGA	+	Vaginal	Prolonged bradycardia	8-9
Ogbonmwan[9]	27	N	Male	3Knots	IUGR	+	Urgent C/S at 28 w	Late decelerations, diminished variability, absence of accelerations	2-4
Ulm[10]	22	M	Female	2Knots	IUGR	+	Urgent C/S at 39 w	Mild variable decelerations	8-8
Jones[11]	24	M	Female	4Knots	AGA	-	Vaginal	No abnormality	9-10
Our case	31	M	Male	SUA+LUC+4Knots	AGA	-	Vaginal	Fetal Demise	0-0

M:Multipar, N:Nullipar, SUA: Single Umbilical Artery, LUC: Long Umbilical Cord, IUGR: Intrauterine Growth Retardation , SGA: Small for Gestational Age, AGA: Apropriate for Gestational Age, C/S: Cesarean section

of a significant IUGR with abnormal Doppler indices close follow up and timely delivery are warranted[5]. Regarding labor induction, continuous cardiotocography is highly recommended. [17].

Besides its rarity our case demonstrates a situation in which coexistence of multiple cord abnormalities leaded to poor obstetric outcome. Presence of one cord abnormality warrants detailed sonography to rule out other cord abnormalities.

In conclusion when a single umbilical artery is detected, it is important to rule out accompanying cord anomalies. In the presence of multiple cord anomalies fetal surveillance is indicated in order to avoid potential umbilical cord compression and cord accidents.

Competing interests

The authors declare that they have no competing interests.

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