



Umbilical Cord True Knot and Overcoiling: A Dangerous Association

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Abstract

The association of true knot and overcoiling of cord is a rare and risky condition during pregnancy. Most knots are loose with no clinical significance, but when becoming tight, they may damage fetal circulation and give a sudden Intrauterine Death (IUD). Coils number/cm identifies Umbilical Coiling Index (UCI). Hypercoiled cords is defined when UCI exceeds 0.3 coils/cm. We report one case of a double rare congenital anomaly occurred at Catania S. Marco's University-Hospital. A 37 years old, para 1: a previous Caesarean Section (CS), gravida 2, was admitted to hospital and an unexplained intrauterine fetal demise at 37th week of gestation was diagnosed. Following CS, a female stillborn of 2,300 grams with large areas maceration signs was extracted. A combination of true knot and spiralization of umbilical cord was observed and identified finally, as death trigger following fetal hyper mobility. Since the movements of the child and the onset of both knot and coiling cord with mechanical ischemic alterations, do not depend on the correct management but often on fortuity, this event is unpredictable, and no fault can be attributed to the medical team.

Keywords: Umbilical cord; True knot; Intrauterine death; Legal problems

Abbreviations: IUD: Intrauterine Death; UCI: Umbilical Coiling Index; CS: Caesarean Section; MCA-F: Middle Cerebral Artery Flowmetry; PI: Pulsatility Index; UA-F: Umbilical Artery Flowmetry

Introduction

The incidence of true knot ranges from 0.3 to 2 % of all deliveries. It is an unpredictable event, but several risk factors are correlated whit this condition: advanced maternal age, multiparity, obesity, previous spontaneous abortion, chronic hypertension and gestational diabetes. Some obstetric conditions, such as genetic amniocentesis, male fetus, mono-amniotic twins, small fetuses, polyhydramnios, long umbilical cord and prolonged gestation, are involved to [1]. As for as funicular coiling, it is considered fisiologic or pathologic depending on the coils and an index has been done depending on number of coils/cm. Undercoiling cord index was defined as less than 0.07 coils/cm and overcoiling as more than 0.30 coils/cm. Regarding to percentility, the umbilical coiling identifies an under coiled cord if the coiling index was below than the 10th percentile, normocoiled if the umbilical coiling index was between the 10th and 90th percentile, or overcoiled if the umbilical coiling index was above the 90th percentile. A correlation between abnormal cord coiling and fetal death, preterm delivery, fetal growth restriction, fetal chromosomal or structural abnormalities, operative delivery due to fetal distress and meconium staining has been found [2]. A very rare case of association of cord overcoiling (spiralization) and true knot in a 37 years old pregnant woman resulting in fetal death is reported.

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Case

A 37 years old woman, in her third pregnancy, with a history of a previous Cesarean Section (CS) at term and a spontaneous abortion at 10 weeks, came into the emergency room at 37 weeks of amenorrhea referring absence of fetal movements for unspecified days. She had no co-morbidities during pregnancy. Blood tests were regular. The pregnant woman had performed routine ultrasounds (US) at the first (6+1 weeks), second (21+1) trimester and they were regular. Third trimester US at 35 weeks gave the following result: monofetal pregnancy, longitudinal situation, cefalic presentation, regular fetal movements and cardiac activity, regular amniotic fluid and placenta with biometric data corresponding to the 35 week 30th percentile. Three vessels were identified by US in the cord: two arteries and one vein. Middle Cerebral Artery Flowmetry (MCA-F) was carried: MCA-F Pulsatility Index (PI) 1.70 (normal range 1.402.32) and Umbilical Artery Flowmetry (UA-F) as well : UA-F PI : 0.93 (normal range 0.44-1.43). After hospitalization, US diagnosis of fetal death was done, however the cause death was not clarified at moment, and no cord turns were found around the neck or the body of the fetus. Due to the previous CS and the current fetal death, although the chance of a vaginal delivery was offered, the pregnant woman chose operative delivery owing to psychological reasons. A transperitoneal CS was performed with a transverse incision of the lower uterine segment, and a stillborn of 2.3kg was extracted. A tight true knot together with spiralization of umbilical cord with 2 coils/cm was detected. A 45cm umbilical cord with ischemia at the knot was observed. The weight of placenta together with umbilical cord was 635gr (Figure 1A). The amniotic fluid was haematic and stained while the amount was regular. The still-born showed large areas of maceration in the site of thorax, abdomen, groin and ankle (Figure 1B).



Figure 1A-B: Left picture: True knot and umbilical cord spiralization. Right picture: Still-born with maceration.

Discussion

The umbilical cord is the first means of communication and protection between both mother and child. Through the blood contained in the umbilical cord the child receives nourishment and oxygen during gestation [3,4]. It consists of a gelatinous tissue called "Wharton's Gelatin" which contains three umbilical vessels:

- i. An umbilical vein that carries oxygen-rich blood and nutrients to the fetus.
- ii. Two arteries that carry catabolites from the fetus to the placenta.

The gelatin is made up of mature, irregularly distributed mucous connective tissue, which can create false knots. The true knots are unusual (1%) and rarely became fatal, because the gelatin usually prevents the total occlusion of the vessels. The umbilical cord true knot formation may be linked to its length, such as to cause its wrapping after increasing of sudden fetal movements. While single nuchal cords, which occur in 15.8-30% of singleton fetuses at term of pregnancy, have not been associated with significant adverse perinatal events, true knots of the umbilical cord are unusual occurring in 0.04-3% of deliveries and have been associated with perinatal morbidity (11% of cases) and the risk of stillbirth is 4-10-fold higher [5-7].

Moreover a true knot of umbilical cord may cause fetal hypoxaemia (ischemic hypoxic encephalopathy). Various abnormalities are observed in the morphology and pathology of the umbilical cord, but knowledge of them is rather poor.

Conclusion

This case report offers to consider the following points:

- a) Correlation with the true knot and the overcoiling cord with the fetal death.
- b) Predictability
- c) Psychological impact
- d) Medico-legal issues

Regarding the correlation between true knot and overcooling cord, in Literature we have found no reports because the association of this pathological conditions is very uncommon and it has not been described so far. Since the true knot was found to be cause of stillborn, as well the overcooling, it is evident to suppose that the incidence of fetal death in such a dangerous association of both pathologies is increased proportionally. The unforeseen fetal demise depends on the tightness of the knots and UCI. The tighter the knot the more frequent is the fatal jeopardy of the unborn; concerning spiralization the hyper coiled cord with high UCI increases the risk of the fetus interfering with the well being and in some cases may lead to fetal death. Coiling of the umbilical cord and adverse pregnancy has been reported. Undercoiling (umbilical coiling index below the 10th percentile, using references values from uncomplicated pregnancies) was associated with fetal death (odds ratio [OR] 3.35, 95% Confidence Interval [CI] 1.48-7.63), spontaneous preterm delivery (OR 2.16, 95% CI 1.34-3.48), trisomies (OR 5.79, 95% CI 2.07-16.24), low Apgar score at 5 minutes (OR 3.14, 95% CI 1.47-6.70), velamentous cord insertion (OR 3.00, 95% CI 1.16-7.76), single umbilical artery (OR 3.68, 95% CI 1.26-10.79), and dextral coiling (OR 1.80, 95% CI 1.02-3.17). Overcoiling (umbilical coiling index above the 90th percentile) was associated with asphyxia (OR 4.16, 95% CI 1.30-13.36), umbilical arterial pH<7.05 (OR 2.91, 95% CI 1.05-8.09), small for gestational age infants (OR 2.10, 95% CI 1.01-4.36), trisomies (OR 9.26, 95% CI 2.84-30.2), single umbilical artery (OR 8.25, 95% CI 2.60-26.12), and sinistral coiling (OR 4.30, 95% CI 1.52-12.2) [8]. Predictability is very difficult. Many times, as reported, these events are unpredictable. It was seen by a study conducted on a sample of 18.500 deliveries that among the whole amount of cases, 133 could go against the real issue, and only a very short percentage (0.08%) could be diagnosed with antepartum US. Despite the knowledge of risk factors and modern diagnostic techniques, this condition is not possible to be prevented, especially in the pregnant before the onset of labour [9,10]. Investigation, however, cannot predict potential future tightening of true knots and, therefore, cannot foresee possible adverse perinatal outcome associated with this condition. Particularly, the umbilical artery Systolic/Diastolic velocity (S/D) ratios in many cases are found within a normal range [11]. The Psychological impact is very strong particularly when the pregnancy woman is on the first pregnancy and worse when the pregnancy arises in aged women in which a further pregnancy is rarer [3,5,12-17]. Medico-legal issues are not to be under-evaluated. Although the physician follows a correct management, the perception of the woman is to find in many cases the reason of this fatal event on the malpractice [18]. Since the movements of the child and the onset of both knot and coiling cord with mechanical ischemic alterations, do not depend on the correct management but often on fortuity, this event is unpredictable, and no fault can be attributed to the medical team.

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