

Lean Umbilical Cord – a Case Report

Thin-Cord-Komplex – ein Fallbericht

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Key words

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- thin-cord syndrome
- placental insufficiency
- Wharton's jelly

Schlüsselwörter

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Abstract

The “lean” umbilical cord (also known as thin-cord syndrome) is a comparatively rare anomaly of the umbilical cord, which has seldom been described in the medical literature. We report on a 35-year-old woman who presented to us at 29 + 4 weeks gestation with vaginal bleeding and cervical incompetence subsequently complicated not only by premature rupture of membranes but also acute placental insufficiency requiring emergency caesarean section under general anaesthesia at 31 + 2 weeks gestation. At surgery no obvious cause for the acute placental insufficiency – such as placental abruption, cord prolapse or true knot of the umbilical cord – was found. Other possible causes such as vasa praevia or placenta praevia had previously been excluded sonographically on admission for vaginal bleeding. The only notable intraoperative finding was a macroscopically extremely thin umbilical cord.

Zusammenfassung

Der sogenannte Thin-Cord-Komplex ist eine relativ seltene Nabelschnurpathologie, die bisher in der Literatur nur wenig beschrieben wurde. Wir berichten über eine 35-jährige Patientin, die sich seit der 29 + 4 Schwangerschaftswoche in unserer stationären Behandlung bei vaginaler Blutung sowie Zervixinsuffizienz befand und bei der es im klinischen Verlauf nicht nur zu einem vorzeitigen Blasensprung, sondern auch zu einer akuten Plazentainsuffizienz mit Entbindung per Notsectio in Intubationsnarkose in der 31 + 2 Schwangerschaftswoche kam. Intraoperativ zeigte sich kein Anhaltspunkt für Ursachen einer akuten Plazentainsuffizienz wie vorzeitige Plazentalösung, Nabelschnurvorfälle bei vorzeitigem Blasensprung oder Nabelschnurknoten. Andere Ursachen wie Vasa praevia oder Placenta praevia bei vaginaler Blutung als Aufnahmegrund wurden bereits im Vorfeld sonografisch ausgeschlossen. Lediglich fiel intraoperativ eine makroskopisch äußerst dünne Nabelschnur auf.

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Case History

A 35-year-old gravida 2 para 1 presented to our antenatal clinic at 29 + 4 weeks gestation with mild vaginal bleeding. Speculum examination revealed under period strength, bright red haemorrhage from the cervical canal. On ultrasound examination there was no evidence of retroplacental haematoma, placenta praevia or vasa praevia. A slender fetus was noted with growth parameters on the 5th percentile, normal amniotic fluid and normal umbilical cord (UC) doppler parameters. The cervix was shortened – length 28 mm – without funneling. The patient had previously been admitted to our department at 23 + 0 weeks gestation with brownish coloured spotting and had received steroids for fetal lung maturation

because of a retroplacental haematoma. At this stage a large placental lacuna was considered in the differential diagnosis. Subsequent regular outpatient follow-up at our antenatal clinic showed no further evidence of retroamniotic or retrochorionic haematoma.

At 29 + 4 weeks gestation steroids were repeated (2 doses of 12 mg betamethasone 24 hours apart). A vaginal swab was positive for anaerobic bacteria and GBS (no Gardnerella) which was treated with local fluomizol. On re-evaluation at 30 + 0 weeks, after 48 hours without further vaginal bleeding, the cervix was unchanged at 28 mm and no funneling. However, the fibronectin test was positive (60 ng/ml) and the patient agreed to a further period of in-patient observation. Subsequently, at 30 + 4 weeks gestation, preterm rupture of mem-

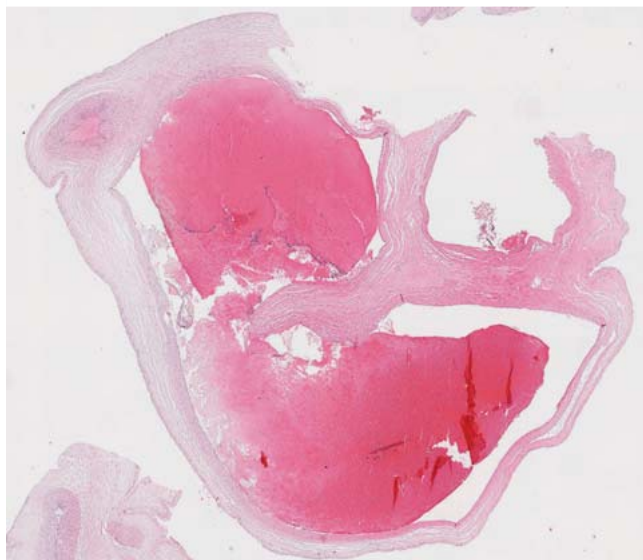


Fig. 1 Histological cross section of umbilical cord showing essential absence of Wharton's jelly; cord otherwise normal with three blood vessels.

branes (PROM) occurred. In view of positive group B streptococcus status intravenous antibiotics (infectocillin) and daily monitoring of infection markers were commenced. Laboratory parameters including CRP levels remained normal and repeat doppler ultrasound at 30 + 5 weeks was unremarkable. Following the positive PROM test, PROM ultrasound did show oligohydramnios. At 31 + 2 weeks gestation a routine CTG revealed multiple, recurrent decelerations with fetal heart rate (FHR) down to 80 bpm (beats per minute) in the absence of uterine contractions. The situation did not improve on changing the patient's position but FHR recovered following a partusisten (fenoterol hydrobromide) bolus. The patient was immediately transferred to our delivery room where tocolysis was continued with a partusisten infusion. In addition to CTG monitoring the fetal heart rate was verified repeatedly by ultrasound; umbilical artery doppler examination was performed repeatedly (resistance index [RI] 0.51, positive end diastolic flow [EDF]). The placenta was sonographically unremarkable and there was no vaginal bleeding. In our delivery room fetal bradycardia recurred (FHR 60 bpm) and fetal heart rate recovered again after a new partusisten bolus. Due to this repetitive situation of fetal bradycardia and decelerations it was decided to perform urgent caesarean section in spinal anaesthesia. On arrival in the operating theatre, however, fetal bradycardia was noted once again and emergency caesarean was then performed under general anaesthesia. A male infant with mild respiratory depression was delivered from a normal cephalic presentation (Apgar at 1/5/10 minutes: 7/8/8; arterial pH: 7.26; venous pH: 7.43; BE: -5.2). Intraoperatively there were no signs of placental abruption. The only notable finding was that the umbilical cord appeared, macroscopically, unusually thin. There were no postoperative complications. On consultation with the patient, and on her request, the placenta and umbilical cord were sent for histological examination.

The anatomical pathology findings (macroscopic) were as follows: Placenta disc-shaped, weight 315 g, size 13 × 11 × 2,8 cm with centrally inserting, 18 cm long section of three-vessel, macroscopically lean (diameter: 0.4 cm) umbilical cord. No umbilical cord knots. Placental surface on incision unremarkable.

Umbilical cord histology: Wharton's jelly essentially absent. UC otherwise normal with three blood vessels (► Fig. 1).

In summary: the placenta was assessed as eutrophic for the gestational age (32nd week of pregnancy) but with premature placental aging consistent with the clinical suspicion of placental insufficiency; the thin UC, with a maximum diameter of 4 mm, was consistent with a diagnosis of lean umbilical cord (or so-called thin-cord syndrome).

Discussion

Lean umbilical cord is an UC anomaly characterised by reduced or completely absent Wharton's jelly. The fetal umbilical cord consists of one vein, transporting oxygen-rich blood from the placenta to the fetus, and two arteries, returning nutrient-poor, carbon dioxide-rich blood back to the mother. Wharton's jelly surrounds the three UC vessels providing a flexible protective layer against vessel compression, kinking and other mechanical forces, thus assuring the fetal blood supply and removal of metabolic waste products.

In a case report published in 1961 Samuel Pike Hall states his conviction that the absence or reduction of Wharton's jelly could be responsible for many unexplained intrauterine deaths, and that too little significance was attributed to this pathology in routine clinical practice [4]. Raio et al. provide a detailed description of UC diameter as it develops through the course of a pregnancy [8]. Our patient delivered in the 32nd week of pregnancy. According to their data normal average cord diameter at this gestation is 16.59 mm, and the histopathologically determined cord diameter of 4 mm in our case would correlate with the normal average cord thickness in the 13th or 14th week of pregnancy. Raio and colleagues further describe an increase in UC thickness up until the 33rd and 34th weeks of gestation to a maximum diameter of 16.72 mm, and a subsequent decrease in thickness to an average of 15.59 mm at term [8]. Weissman et al. also describe the changes the umbilical cord undergoes during pregnancy using nomograms, observing a maximum UC diameter of up to 18 mm between the 38th and 39th weeks of pregnancy [10]. Both publications (Raio et al. and Weissman et al.) provide detailed tables of UC diameter for the respective gestational ages [8, 10]. Weissman et al. use measures of cord diameter taken in the longitudinal plane, whereas Raio et al. measure the cord in cross section (transverse plane). Both publications recommend taking measurements as close to the fetal abdominal UC insertion as possible.

Another feature of our case that was typical of a lean umbilical cord was the ultrasound finding of a slender fetus with growth along the 5th percentile in the presence of normal UC doppler parameters (small for gestational age, SGA). Bruch et al. describe an association between reduced Wharton's jelly, hypoplastic UC blood vessels and fetal intrauterine growth retardation due to resulting placental insufficiency without pathological doppler parameters [1]. Raio et al. also conclude that fetuses with thin umbilical cords are at increased risk of growth below the 10th percentile (SGA or IUGR) compared to those with normal UC diameters and more often show signs of stress at birth [6, 7]. They state that fetuses shown to have thin umbilical cords on ultrasound from 20 weeks gestation onwards have a 4.4 times increased risk of being SGA at birth [7]. When ultrasound was conducted after 25 weeks gestation the risk was increased to 12.5 times [7]. In addition, they report that babies with thin UCs were significantly

more likely than those with normal thickness cords to have a 5-minute Apgar score < 7, and oligohydramnios was more common [7].

It should however be noted that a lean umbilical cord does not necessarily always result in complications. Raio and Ghezzi et al. highlight that many pregnancies remain uncomplicated despite a lean UC on ultrasound examination, and that additional criteria should be sought in order to identify higher risk situations. For example, according to Ghezzi et al., the strongest predictor of poor outcome is the umbilical vein cross-sectional area, i.e. the diameter of the vein with respect to UC cross-sectional diameter [3], and report significantly increased numbers of perinatal fetal death and increased neonatal intensive care admission rates when the umbilical vein area is below the 10th percentile [3]. There is also a significant association between increased IUGR rate and lean umbilical cord where the UC area is below the 2.5 percentile. Other studies agree with these results and recommend the measurement of UC area as a screening parameter to detect patients with values below the 10th percentile, allowing monitoring to be intensified as appropriate [1, 7, 9].

Conclusion

Today it is known that a lean umbilical cord due to reduced or completely absent Wharton's jelly is associated with a worse neonatal outcome in affected children [2, 3, 5, 7, 9]. In the case presented here the combination of a lean UC and reduced amniotic fluid secondary to PROM was most likely responsible for reduced fetal perfusion, which in turn produced recurrent decelerations on CTG prompting urgent delivery. Of note, the CTG only became pathological relatively late, which could be explained by the combination of PROM, lean UC and the fetal position, resulting in mechanical UC compression. This case illustrates that in the presence of oligohydramnios, SGA or IUGR it is not only important to assess fetal doppler parameters and the number of UC vessels, but also cord morphology, since in rare cases this can play an important role. It would seem sensible to assess UC morphology sonographically on a routine basis, or at least in the presence of SGA or IUGR of uncertain cause, thus enabling detection

of women who might benefit from more intensive antenatal monitoring. In our case prior knowledge of the presence of a lean UC would not have altered obstetric management since close observation and monitoring were already indicated due to SGA and PROM. Notably, however, postpartum knowledge of this diagnosis was important for the patient involved as it provided an explanation for her preterm delivery, and the low recurrence risk was significant from a psychological point of view with respect to the planning of possible future pregnancies.

Conflict of Interest

None.

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