Complex umbilical cord entanglement

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We describe three cases of complex umbilical cord entanglement seen in the past 5 years at our Placenta Clinic. The cases were associated with intermittent fetal bradycardia, reduced fetal movements, and were all delivered prematurely with multiple umbilical cord loops around the neck, limbs, or body. One intrauterine fetal death occurred. Placental pathology showed downstream effects of cord occlusion and fetal thrombotic vasculopathy.

Keywords: Umbilical cord entanglement, fetal bradycardia, placenta and cord pathology

Introduction

Benign umbilical cord entanglement is a common finding, with one or two loose loops of cord around the neck in up to 25% of term deliveries [1]. Multiple (three or more) loops of umbilical cord entanglement may be found in otherwise unexplained stillbirths and in pregnancies complicated by intrauterine growth restriction [2]. Inter-twin cord entanglement is almost universal in monoamniotic twin pregnancy, where intensive inpatient fetal monitoring and early Cesarean delivery may improve perinatal outcomes [3]. By contrast, the detection of multiple loops of cord around the fetal neck in otherwise normal third trimester pregnancies poses management dilemmas for clinicians, since the positive predictive value for adverse outcome is low. Here we report three scenarios of complex umbilical cord entanglement encountered in the past 5 years at our Placenta Clinic.

Case 1

A 32-year-old primigravida woman was originally referred to plan delivery close to our Pediatric Center for severe fetal pulmonary stenosis and a two-vessel cord. The fetus was otherwise structurally normal and had low impedance umbilical artery Doppler, normal amniotic fluid and biophysical profile score at 32 weeks gestation. Two weeks later she had similar findings (Figure 1A) but 6 days later at 34 weeks and 6 days gestation she presented to clinic with decreased fetal movements. Ultrasound evaluation now showed absent end-diastolic flow velocity in the umbilical artery (Figure 1B). Uterine artery Doppler was normal (Figure 1C) and the placenta looked healthy (Grannum grade 0) with one small basal inter-villous thrombosis (Figure 1D). An extended biophysical profile score was 4/8. She was transferred to a labor and delivery room where a non-stress test (NST) was non-reactive with some decelerations. She had an immediate Cesarean delivery under spinal anesthesia with a male infant weighing 2130 g (5th percentile); the umbilical cord was wrapped around the lower left leg eight times. His Apgar scores were 5 at 1 min and 8 at 5 min with a cord artery pH of 7.1. A Betke–Kleihauer test was negative and he was not anemic. The infant was transferred to our Pediatric Center for treatment of his pulmonary stenosis. Pathology of the placental disc revealed one infarct, confirmed the basal inter-villous thrombus, and showed congested immature third trimester villi consistent with a degree of cord obstruction. No abruption was found. The cord was 31.8 cm long (shorter than average cord length [4]) with increased coiling (2.5 turns in a 5 cm segment) and a single umbilical artery.

Case 2

A 35-year-old G2 P1 woman received ongoing high-risk pregnancy unit care for a history of ulcerative colitis and a previous full proctocolectomy; her antenatal course was uncomplicated, though a recent ultrasound had diagnosed a double nuchal cord. She presented to obstetrical triage at 35 weeks and 4 days gestational age with reduced fetal movements, reporting that she had only felt the baby move once in the previous 24 h. She was experiencing mild contractions, but there was no bleeding or rupture of the membranes. A biophysical profile was 2/8 while an NST showed some variable decelerations, some of which accompanied her contractions. She therefore had a Cesarean delivery of a male infant weighing 2280 g (>10th percentile) that had seven tight loops of nuchal cord. His Apgar scores were four and eight at 1 and 5 min respectively, and the umbilical artery pH was 7.1.4. He was transferred to the NICU due to transient tachypnea, but was discharged home 4 days later in good condition. Placental pathology was normal apart from increased cord length (73 cm) and increased coiling.

Case 3

A 25-year-old G 3 P1 woman was referred with a history of recurrent vaginal bleeding during the first trimester and echogenic bowel on a recent ultrasound. At 19 + 6 weeks gestation the ultrasound examination was noted for several episodes of fetal bradycardia to around 60 beats per minute. The fetal anatomy was normal including a cardiac screen. Fetal size, amniotic fluid, and placental size and morphology were normal. Both the uterine artery (pulsatility index 0.85) and umbilical artery (pulsatility index 1.6) Doppler studies were normal during
periods of normal heart rate (140–145 beats/min). A normal middle cerebral artery peak systolic velocity (31 cm/sec) for gestation excluded fetal anemia and there were no signs of non-immune hydrops. Maternal blood was taken for anti-Ro and La antibodies (subsequently negative) and a fetal echocardiogram was arranged for the following week. Unfortunately at this 20 + 2 week examination an intrauterine fetal demise was noted. She returned to our service for an induction of labor, where at delivery the cord was found to encircle the neck, shoulder, and body for a total of four loops. Two of the loops were wound tightly around the left arm. Pathology showed that the umbilical cord measured 42.5 cm in length (longer than average [4]) and was hyper-coiled with up to five complete turns in a 5 cm length (normal is one coil for every 5 cm of length [2]). Fetal autopsy showed mild asymmetrical intrauterine growth restriction and early hypoxic-ischemic changes in the brain. The heart was anatomically normal thereby excluding non-compaction of the myocardium [5]. Some old hemorrhage within the decidua with adjacent villous infarction was noted consistent with her repetitive first trimester bleeding.

Discussion
Cord length positively correlates with the risk of cord entanglement [6], and while cord entanglements may pose a risk of intermittent cord obstruction, this phenomenon may also protect against cord prolapse during labor [1].

The prenatal diagnosis of cord entanglement has been found during ultrasound scans as early as 10 weeks gestation [1], due to increasing use of the nuchal translucency assessment as part of trisomy 21 screening. A recent study demonstrated degrees of cord entanglement, involving the neck, hand, leg, body and shoulder, in 63% of fetuses at 13–16 weeks gestational age [7]. Most of these nuchal cords will resolve in later gestation [2].

These data suggest that cord entanglement may be considered a normal characteristic of early fetal development [7].

Animal models (using fetal sheep) have demonstrated that a 50% occlusion of the umbilical cord is required to induce variable decelerations in the fetal heart rate [8]. These data may be interpreted clinically to suggest that non-stress testing is useful in discriminating between benign and serious cord entanglement. Indeed, this rationale is used in the surveillance of monoamniotic twins. In addition, the cord entanglement may either directly limit fetal activity, or else induce a variable hypoxic stress, that collectively will be sensed as reduced fetal movements. Two of our cases were assessed in response to complaints of reduced fetal movements. In one case, the dramatic alteration in umbilical artery Doppler prompted evaluation of other causes, such as fetomaternal hemorrhage, as we were reluctant to deliver a premature infant with congenital heart disease. The sudden development of absent-end-diastolic flow velocity in a single umbilical artery, in the absence of uteroplacental vascular pathology, is very rare. In a prospective series of 212 high-risk patients, 0/19 cases of absent-end-diastolic flow velocity occurred in the majority of women with normal placental function. We therefore suspected cord obstruction, but did not find a nuchal cord. In the future, we will check the lower limbs in this rare context.

Placental and cord pathology examinations are important to perform when cord entanglement is suspected of causing stillbirth or acute fetal compromise. The distal placental lesion associated with persistent cord occlusion is fetal thrombotic vasculopathy caused by chronic fetoplacental vascular stasis [9]. One study found clinical or pathological evidence of cord occlusion in 16 of 23 cases with this placental histologic lesion [10]. Therefore submitting the placenta and cord for examination in this context may reveal evidence of cord obstruction that is likely to have been functionally relevant to the clinical outcome. Fortunately these
forms of cord pathology have a low recurrence risk in future pregnancies.

The third case was also unusual, since it presented before either the maternal perception of fetal activity, or the utility of non-stress testing. A common cause of intermittent fetal bradycardia during routine fetal anatomical ultrasound is probe pressure, especially when examining the fetal intra-cranial structures. On this occasion the bradycardia was repetitive despite light pressure and no placental cause was found. In view of the history for recurrent losses, the possibility of evolving congenital heart block was considered. A fetal echocardiogram was arranged to specifically look for this and the possibility of occult congenital heart disease. We did not search for cord entanglement at this gestational age, in part as the umbilical artery Doppler was normal. While the fetus can be manipulated in-utero at this stage, for example during the placement of chest shunts, to our knowledge we are unaware of any reports describing the dis-entanglement of complex loops of umbilical cord diagnosed prenatally. Technically this seems feasible though the ongoing risk of subsequent re-entanglement is likely to be high. Therefore the ability of prenatal diagnosis and therapy to improve the outcome in the context of this case seems improbable.

Declaration of Interest: The authors report no conflicts of interest.

References