

Severe antenatal strangulation and sudden fetal death occurs in term: case report

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Summary

The authors report a case of a sudden antenatal death, by severe strangulation, unlikely related in a term pregnancy; multiple loops of nuchal umbilical cord (UC) (ten), rarely describe in literature, were observed around the fetal neck. The in utero fetal death (IFD) was suspected by the non-attendance of fetal movements and confirmed by US scan. The tight nuchal cord around the neck (tCAN) diagnostic was made during caesarean delivery, as it was not discovered in pregnancy US scan monitoring nor in the US scan made in emergency. The newborn examination shows severe fetal strangulation by the presence of many spires of a too long UC (1.50 m). Autopsy was not been accepted by the family. Through this reported case the authors wanted to show the difficulties of its diagnosis in less developed Sub-Saharan country where US scan practice is not usual.

Key words: Antenatal fetal death; Multiple loops; Nuchal cord; Strangulation.

Introduction

Intrauterine fetal death (IFD) is a devastating complication of pregnancy, an exceptional painful event from whom any parent is prepared. The overall incidence of stillbirth is reported to be around 1.2% with an incidence after 20 weeks of 1%, after 28 weeks of 0.4% and at term of 0.2% [1]. The underlying reasons can be divided in maternal, fetal, and placental causes. Concerning the fetus, umbilical cord (UC) complications seem to be the most common cause for fetal demise in the third trimester [2]. Multiple nuchal cord loops of UC around the neck (CAN) are common with an incidence reported to be between 15.8% and 30% [3]. Unfortunately these complications are regarded as unpredictable and unpreventable [4]. Tight nuchal umbilical CANs (Figure 1) are relatively frequent findings and some times associated with a negative fetal outcome. During pregnancy, strangulation is exceptional (Figure 2) but can occur at anytime during second or third trimesters. Strangulation and constriction of UC can be associated and lead to acute intrauterine asphyxia. Fetal deaths occurred frequently (15-30 %) during trial [4].

Herein clinical case the authors report a sudden IFD in third trimester, due to a severe strangulation of tight umbilical cord around neck (tCAN) with multiple UC turns. Through it the authors would like to show difficulties of its diagnosis and management in low socio-economic African countries.

Case Report

A 16-year-old primiparous, with no particular individual antecedents, was admitted for a broken right femur by firearm. She was carrying a progressive pregnancy estimated at 39 weeks. The investigation found no pelvic pain or vaginal discharge, and fetal active movements were well felt by the patient. Hemodynamic and general state of health were good. Obstetrical examination found a soft uterus height measuring 32 cm. Fetal heart beats were regular at fetal Doppler at 148 beats / minute. Fetal presentation was vertical but higher. X-ray radiography found a closed break of right femur. Ultrasound scan found a fetus in breech presentation with regular cardiac activity regular. No diagnosis of nuchal cord (NC) was evoked. Placenta was well adhered and had a normal insertion, with no placental hematoma. A cesarean section had been planned and right leg was placed in splints awaiting for surgery. Fetal movement assessment and fetal Doppler were performed twice in the day. The clinical course was satisfactory but three days later the patient indicated difficulties in perceiving fetal movement without any other signs. Fetal Doppler showed no fetal heart activity, confirmed by US scan. Cesarean section was performed and showed a stillborn with intraoperative multiple torsions of UC around the neck, with severe entanglement (Figure 2). The stillborn had weight of 3,100 grams and did not present any external abnormalities or dysmorphic signs (Figure 1). The UC had a length of 143 cm and contained three blood vessels (Figure 3). No constriction, knots or any others abnormalities (cupula nor abruptio placentae) were found (Figure 3). No histological examination of placenta and autopsy were done. Treatment of broken femur was performed in the same surgical procedure. The postoperative course was simple with healing of cesarean wound section observed two weeks later. The remaining of the follow-up was made by a trauma specialist.

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Figure 1. — Tight cord around the neck (tCAN)



Figure 2. — Severe strangulation.

Discussion

NC is defined as loop of UC 360° or more around the fetal neck. “Tight” is defined as the inability to manually reduce the loop over the fetal head, and loosen as the ability to manually reduce the loop over the head. [5]. In the present case, the authors found multiple NC loops which is a very uncommon finding.

According to Lal Neena *et al.*, NC come and go during gestation, but become more frequent towards term [6]. It is most frequently seen among UC abnormalities and the prevalence has been reported to be 15-24% at delivery [6-8]. The incidence increases with advancing gestation from 12% at 24 to 26 weeks to 37% at term [9]. They are not associated with perinatal morbidity and mortality but in some fetuses and newborns, CAN may cause problems, especially when the cord is tightly wrapped around the neck (Figure 1). The cluster of cardiorespiratory and neurological signs and symptoms associated with unique physical features that occur secondary to tight cord around-the-neck has been referred to as tCAN syndrome [9].

Few studies have shown that NC and or tCAN can affect the outcome of delivery and may have long-term effects on the infant [10]. Meanwhile, their role as risk factor of stillbirth is always discussed [10, 11] and this reported case proved the contrary. However, some case reports of post-mortem findings on stillbirths show negative pathology reports and the present authors agree that tCAN is the only cause of death [12]. CAN represent 15-30% of deliveries and are observed enough during labour [13] and responsible for about 50% of fetal cardiac beat anomalies [14]. It is the unique physical features of tCAN syndrome that distinguishes it from birth asphyxia even though there are many similarities. According to Bendon *et al.* [15], UC abnormalities are considered as one of the causative factors for birth asphyxia and NCs can be sub-classified by the number of nuchal wrapping, by complex wrapping of other body parts, and by the type of



Figure 3. — Placenta examination.

loop [15]. In type A we have a simple 360° rotation that must end with the placental end of cord on top, versus type B in which the fetus rotates through the loop creating an incipient knot with the fetal end of the cord. Other studies classified tCAN (those cut to enable delivery) versus loose cords. In general the more complicated the nuchal wrapping, the more likely that there will be complications [15].

The clinical significance of NC is controversial. Many factors contributed to the controversy such as tightness of the NC, cords wrapped around the neck in a “locked fashion” or not, and multiple cord entanglements around the fetal neck [3]. It is suggested that a moderate tight CAN would impair cephalic venous blood flow only, whereas a very tight would compromise the umbilical circulation and produce systemic hypoxia, hypercapnia, and acidemia [3].

During pregnancy, the indentations of the neck with NC can be deep, but there is little evidence that the cord causes

significant tracheal obstruction. A few case reports suggest that the cord interfered with the fetal swallowing and caused polyhydramnios [16]. In tCAN, there may be jugular venous obstruction especially if the NC tightens one [17]. The presence of NC alone is not a cause of stillbirth. In an otherwise unexplained fetal death, assigning cord wrapping as cause of stillbirth is supported by finding a complicated wrapping or a very tight cord as evidence of tension. A short length of free cord from the neck to placenta or from the neck to the umbilicus would support cord compression or torsion as mechanism of death. If the cord is still attached to the infant, the type A or B of wrapping can be determined simply by which segment of cord is on top (cephalad). Type B may be more likely to result in cord occlusion [15].

During labour, the manifestation of tCAN symptomatology seems to occur both in the presence of normal and depressed Apgar scores [18]. UC strangulation due to tCAN may cause obstruction of blood flow first in thin umbilical vein wall, while infant's blood continues to be pumped out through the thicker walled umbilical arteries, thus causing hypovolemia and hypotension resulting in acidosis [19]. Anemia [18] and mild respiratory distress may occur. Case of facial and conjunctival petechiae [19] and rarely petechiae of the neck and upper part of the chest and skin abrasion of neck (Figure 2), have been described [20] where the cord was tightly wrapped and facial suffusion [21], as in the described case. Some born alive infants may also be somewhat obtunded with a low tone and have transient feeding difficulties. These findings raise the possibility of transient encephalopathy, which may lead to long-term complications [9]. A stillbirth attributed to a cord problem should have evidence of cord obstruction or circulatory compromise. Other potential causes of stillbirth need to be excluded prior to labelling cord abnormalities as causative factor, since they be seen in more than a third of all normal live births [9].

tCAN syndrome may conceptually be considered as strangulation which may result in non-lethal problems or death as in the present observation. The pathophysiological mechanisms of strangulation injuries (lethal and non-lethal) involves venous, arterial obstruction (arterial spasm due to carotid pressure) in the neck and vagal collapse (increased parasympathetic tone) [9]. This can lead to cerebral stagnation, hypoxia, and unconsciousness and at last loss of muscle tone. A study on potentially asphyxiating conditions and spastic cerebral palsy in infants of normal birth weight showed evidence of association of tCAN in children with quadriplegia [22].

Rocha *et al.* showed that intermittent UC occlusion in preterm and near term sheep, caused a decline in pO₂ and pH, and higher PCO₂ and altered brain protein synthesis/degradation [12]. According to Parast *et al.*, significant correlation of placental changes of "minimal histologic criteria" are associated with cord accidents (as tCAN is part of cord accidents), using specific placental histologic criteria by restriction of umbilical blood flow in unexplained stillbirth [10].

With CAN, the UC measured over 70 cm (6-7%) or more than one meter for others (1%). In a series of 104 cases with CAN (7.5%), Gasser *et al.* [23] found macerated fetus received at the laboratory by showing obviously a constriction of the neck or a limb, or fetal body by a stretched cord, which is abnormally too long. The excess length of UC, as explained, can be risk factor of this funicular disease. Keckstein *et al.* [24] found that this risk can be multiplied when UC is longer than 70 cm.

The diagnosis of CAN is possible during pregnancy but not simple, particularly in undeveloped countries. Indeed US scan machines use is not readily available. The best moment for the present research was third trimester of pregnancy. Color Doppler Ultrasound is a good investigation when it detects the presence of a NC if the UC could be followed 360 degrees around the fetal neck [25], with a sensitivity in diagnosing of 37.5%, specificity of 80%, and positive and negative predictive values respectively estimated at 29% and 85%, according to Peregrine *et al.* [26]. In poor countries, US scan is rarely employed during pregnancy, firstly for its high cost and secondly for its unavailability in most maternities. Otherwise, many pregnant in poor countries are illiterate and came to maternities only when they are ready to deliver. CAN diagnostic is mostly utilized during labour or at birth in poor countries. According to Peregrine *et al.*, the sensitivity of the US diagnosis of a NC is low prior to induction of labor at term. Therefore NC does not appear to be important for many authors in choosing of way of birth. The low US detection rate of a NC may limit its use in decision of induction of labour in high-risk pregnancies [26]; however, during labour, diagnosis may be suspected in front of abnormal fetal heart rate (FHR) occurring during a uterine contraction, or a lack of progression of the presentation. The immediate threat of CAN presence is fetal distress which presents during labour via two types of changes: an alarming slowdown of FHR (detected by monitoring) during the first contractions or altering of amniotic fluid after membranes rupture. These two problems require acceleration of delivery either by classical vaginal delivery with systematic episiotomy or by performing a cesarean section. Cesarean section planning is recommended as mode of delivery in the present center because of impossibility of fetal monitoring during labor, besides the too long of a delay to perform a caesarean section.

Antenatal or perinatal fetal death represents a tragedy or negative experience sometimes incomprehensible regarded as failure of pregnancy monitoring. This kind of death has variable frequency according to socio-economic level of population. This frequency is low in developed countries (4% to 10%), and high in poor countries (15% to 50%). When umbilical cord is coiled around the neck, it accounts for 70% to 80% of all umbilical cord complications noticed at deliveries. For World Health Organization there was 4.1% of in utero fetal death in Africa and 37% of these deaths occur during labour [27]. Fetal death etiologies are multiple and varied and can be caused by fetal, adnexal or maternal reasons. Nowa-

days, tCAN is increasingly blamed in the adverse issue of these pregnancies. Indeed tCAN can cause fetal asphyxia by strangulation even if there are no uterine contractions. In the current case, there were no uterine contractions but the authors observed many loops around neck, which explain the sudden onset of fetal death by severe strangulation. In literature, tCAN is often related to decreased placental oxygen transfer with signs of fetal acute hypoxia by fetal perfusion disorders. The coiled cord, while blocking blood circulation, induces placental parenchymal infarction, edema, congestion, and / or funicular thrombosis. No maternal disease was diagnosed prenatally. In case a fetal anemia occurs systematically at birth due to fetal-maternal hemorrhage can also be observed. It may be necessary to cut UC between two clips before performing the release of fetal shoulders in deliveries [28].

Newborn with a tCAN were slightly more likely to be admitted to neonatal intensive care unit (6.6% vs. 5.9% admission rate, $p = 0.000$). The subset of very low birth weight neonates with a CAN, compared with those with no NC, were of the same gestational age and birth weight, with same Apgar scores, and were not more likely to have severe intraventricular hemorrhage, retinopathy of prematurity or periventricular leukomalacia, or to die. NC in normal pregnancies at term associated with an increased rate of caesarean delivery in nulliparous was described. The presence of a NC results in slightly lower Apgar scores at one and five minutes, mainly as a consequence of higher caesarean delivery rates [28].

Conclusion

Antepartum fetal death by tight NC around the neck is not uncommon. Its diagnosis is difficult in poor countries particularly because of the lack of US scan and its high cost. However NC must be suspected at term in pregnancy to avoid fetal antepartum or perpartum complications. A prospective study of the fetal income must be performed according to the way of birth modalities in poor countries.

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