TRUE UMBILICAL CORD KNOT, NOCHAL CORD AND CORD ROUND BODY WITH FAVOURABLE OBSTETRIC OUTCOME IN AN UNBOOKED ELDERLY NULLIPARA: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Basically, umbilical cord knots in general can be classified as true or loose. True knot of the umbilical cord in contrast to other conditions like nuchal coils and cords around the body, may have significant clinical sequelae. Tightening of umbilical cord knot is a very rare and highly unprecedented complication of pregnancy that can lead to foetal demise or neonatal death. We report a rare case of True umbilical cord knot, nuchal cord and cord round body in Yenagoa, South south Nigeria: A 42 year old unbooked elderly nullipara who presented at 41 weeks and 3 days with Labour pains of 4 hours, Drainage of Liquor of 2 hours and Bleeding per vaginam of 30 minutes. She had an Emergency caesarean section for Abruptio placenta with a live baby and foetal heart irregularity in labour. Intraoperative findings include a bloody liquor, Live female neonate with Apgar score of 4 [1] 8 [5]. Birth weight was 3.4kg. There was no gross congenital anomaly seen. Placenta was fundally located, Placenta weighed 0.7kg. A true umbilical cord knot, nuchal cord and cord round body twice was found. Umbilical cord length was 144 cm. The true knot was 50 cm from foetal insertion and cord was centrally inserted into the placenta with a retroplacenta clot of about 200ml. Currently, our experience on True umbilical cord knot is constrained to incidental postnatal finding. However, until prenatal diagnosis is improved with the use of ultrasonography for True umbilical cord knot, good clinical acumen, intrapartum surveillance and prompt obstetric intervention remain pivotal in the aversion of perinatal mortality.

Keywords:, True knot, Umbilical cord, Nuchal cord, Perinatal mortality

INTRODUCTION

Basically, umbilical cord knots in general can be classified as true or loose. True knot of the umbilical cord in contrast to other conditions like nuchal coils and cords around the body, may have significant clinical sequelae. [1,2] True umbilical cord knot occurs in about 0.3 – 2.1% of pregnancies [1-4] and tightening of umbilical cord knot is a very rare and highly

unprecedented complication of pregnancy that can lead to foetal demise or neonatal death. [4] It has been associated with 6% perinatal mortality. [1]

Despite the rarity of True umbilical cord knot, certain predisposing factors have been identified. These include: male foetuses, monoamniotic twins, process of undergoing genetic amniocentesis, gestational diabetes mellitus, [1] long umbilical cords, [2,4] polyhydramnios, [4] small size foetuses, [2,4] and multiparity. [3] Other maternal factors associated with true knot of the umbilical cord include: advanced maternal age, history of miscarriages, obesity, anaemia and chronic hypertension and then prolonged pregnancy. [2,4,5]

Although the exact pathophysiology of formation of True knots of the umbilical cord is unclear, [6] however, it occurs when the foetus moves through a loop or loops of cord while being active in the uterus. [4,,7,8,9] While most True knots form very early in pregnancy as believed by most investigators due to the relative large amount of amniotic fluid around 9 – 12 weeks period, in contrary, there is reported evidence of knot formation of the umbilical cord during labour. [10]

Being a new training or research centre in the region, no case report or study has been done in the past on True knot of the umbilical cord at the Federal Medical Centre, Yenagoa, South south Nigeria. Also, the rarity of this condition cannot be overemphasized. We therefore present such in this article.

CASE REPORT

Mrs I. G., a 42 year old unbooked G_3P_0 $^{+2}$ trader with secondary level of education who was unsure of her Last Menstrual Period (LMP), however, an ultrasound Scan (USS) done at 27 weeks Gestational Age (GA) puts her Expected Date of Delivery (EDD) at 29/11/2015 and Estimated GA at presentation was 41 weeks + 3 days.

She presented to labour ward on the 9th of December, 2015 with Labour pains of 4 hours, Drainage of Liquor of 2 hours and Bleeding per vaginam of 30 minutes.

Labour pain started 4 hours prior to presentation. She had Induction of Labour (IOL) at a Primary Health Centre for prevention of prolonged pregnancy with one dose of misoprostol inserted per vaginam 3 hours before the onset of labour pain. Pain was intermittent and gradually increasing in intensity, frequency and duration.

Drainage of liquor started 2 hours prior to presentation. It was spontaneous and sudden in onset, copious, initially clear but later became slightly bloody. There was no umbilical cord prolapse and she still felt foetal movement satisfactorily.

Shortly after spontaneous rupture of foetal membranes, she started bleeding actively per vaginam, bright red with some blood clots which was initially contained with 3 well soaked comfit pads. There was no trauma to the abdomen or instrumentation. She is not a known hypertensive or diabetic and does not take tobacco products in any form. There is no known family history of twinning or exaggerated pregnancy symptoms.

There was no warning bleeds in this pregnancy, fainting attacks, dizziness. At the onset of bleeding PV, she was asked to come to our facility for expert management.

Index pregnancy was conceived spontaneously and booked at the referring health centre at a GA of 25 weeks. She could not remember her booking parameters but they were all said to be normal. Obstetric USS done at 27 weeks GA showed active singleton foetus in longitudinal lie, cephalic presentation, posteriofundal placenta. Doppler ultrasound was not done as it was not routinely done in the referring health centre. She had 2 doses of Intramuscular Tetanus Toxoid and Oral Sulphadoxine Pyrimethamine for Intermittent Preventive Treatment (IPT) for malaria both of them at 25 and 29 weeks GA respectively. Pregnancy was uneventful until recent event.

She had 2 Termination of pregnancy in 1999 and 2000 at GA of 6 weeeks. Pregnancies were confirmed with biochemically, unplanned and undesired. Had Manual Vacuum Aspiration (MVA) at private clinic by medical officers. There were no post abortal complications.

Examination at presentation revealed a young woman in intermittent painful distress, not pale, afebrile, anicteric, acyanosed, not dehydrated, no pedal edema.

Her respiratory rate was 20c/m and chest was clear clinically. Pulse rate was 104 b/m, regular, full volume blood pressure was 110/70mmHg, only 1st and 2nd heart sounds were heard.

Abdomen was enlarged, moved with respiration, no area of tenderness. Liver, spleen and kidneys were not palpably enlarged. She had 3 moderate contractions in 10 minutes. Symphisiofundal (SFH) was 37cm compatible with term gestation. Foetal parts difficult to palpate, however, Foetal Heart Rate (FHR) was 152b/m, irregularly irregular.

Vaginal examination revealed a blood smeared female external genitalia with active bleeding PV. Cervical os 5cm dilated. Foetal membrane was absent. Gloved examining finger stained with fresh blood.

A diagnosis of unbooked elderly nullipara with abruption placenta with live baby and foetal heart irregularity at term.

She and her husband were counselled on her condition and the need for an emergency caesarean section. IV access was secured with 2 wide bore cannulae.

Samples were taken for urgent PCV= 43%, RVS= negative, bedside clotting time = 5mins, Urinalysis= negative for ketones, glucose and protein.

Intravenous fluid 1litre of normal saline was commenced. Informed consent was taken. Theatre was booked. Consult was sent to neonatologist. She was prepared for theatre. She was in theatre 18 minutes from the time the decision was made to operate on her (i.e 25 minutes from presentation)

Intraoperative findings include; a well formed lower uterine segment, bloody liquor, Live female neonate delivered, Apgar score of 4 ^{1} 8 ^{5}. Birth weight was 3.4kg. There was no gross

congenital anomaly seen. Placenta was fundally located, Placenta weight was 0.7kg. A true umbilical cord knot, nuchal cord and cord round body twice was found. Umbilical cord length was 144 cm. The true knot was 50 cm from foetal insertion and cord was centrally inserted into the placenta, Retroplacenta clots of about 200ml. Grossly normal fallopian tubes, ovaries and urinary bladder.

Figure 1: Demonstration of a true knot, nuchal cord and cord round body twice following Emergency caesarean section

Figure 2: Clamping of the cord

Figure 3: True knot of the umbilical cord

DISCUSSION

This is the first reported case of True umbilical cord knot, nuchal cord and cord round body in Yenagoa. From our literature search in Nigeria, Ikechebelu JI et al reported a case of True umbilical cord knot in Nnewi, [11] South East Nigeria. Unlike our case with favourable obstetric outcome, theirs led to foetal demise. This may be explained by the fact that our patient was managed as a case of Abruptio placenta with a live baby and foetal heart irregularity and subsequently had an Emergency Caesarean Section 25 minutes into presentation (ie 4 hours 25 minutes from onset of labour) while theirs had a spontaneous vertex delivery following 13 hours of labour. Labour may give room for tightening of the umbilical cord which may occlude the foetal circulation leading to foetal demise. Although this is a possibility of the cause of foetal demise, autopsy was not done to rule out other foetal anomalies. Hence, True umbilical cord knot may also be an incidental finding.

Certain predisposing factors of True umbilical cord knot have been identified. These include male foetuses, monoamniotic twins, process of undergoing genetic amniocentesis, gestational diabetes mellitus, [1] long umbilical cords, [2,4] polyhydramnios, [4] small size foetuses, [2,4] and multiparity. [3] Other maternal factors associated with true knot of the umbilical cord include advanced maternal age, history of miscarriages, obesity, anaemia and chronic hypertension and then prolonged pregnancy. [2,4,5] Our patient, Mrs I.G., had some of these factors. She is 42 years (advance maternal age) with history of induced miscarriages and umbilical cord was 144 cm which was very long. Although cord length is usually variable, the mean cord length as reported by Agwu UM et al [12] was 57.87 +12.6 cm. Going by this, the umbilical cord length of this index case was more than double of the normal. However, unlike what is reported in other literatures, Mrs I.G., was a nullipara, gave birth to a female baby. The history of polyhydramnios could not be ascertained because she was unbooked, had drained liquor and there was no prior ultrasound evidence of polyhydramnios. Other aforementioned risk factors were ruled out.

There is no robust literature on the diagnosis of True knot of the umbilical cord prenatally and is regularly an accidental discovery. [13,14,15,16,17] Its prenatal qualities may not be obvious. The visualization of the knot is usually lost in screening through prenatal ultrasound amid the second and third trimesters. [17] Albeit early observations suggest that the knot of umbilical cord is formed very early in pregnancy due to the relative large amount of amniotic fluid around 9 – 12 weeks period, in contrary, there is reported evidence of knot formation of the umbilical cord during labour. [6,14] Reasons for failure of diagnose have been adduced. The interpretation through two-dimensional ultrasound images of a complex three-dimensional object, such as a knot of the umbilical cord, is extremely difficult. [17] Moreover, the interposition of foetal parts and reduction of amniotic fluid could make the diagnosis a challenge. For this reason, true knot of umbilical cord is an obstetric complication usually with postnatal diagnosis. The complementary ultrasound techniques, as colour Doppler, Doppler velocimetry, and four-dimensional ultrasound, may improve the detection rate of true knot and differentiate it from false or loose knot. [3,15] Nevertheless, prenatal diagnosis of true knot of the umbilical cord has been demonstrated using ultrasound which will show “cloverleaf pattern” on gray scale images. [7, 17] For Mrs I. G., the diagnosis

was made postnatally, as she was unbooked and presented in labour. Even though Obstetric USS was done at 27 weeks, the diagnosis was missed. In addition, routine colour Doppler or four – dimensional ultrasound is not recommended because the cost may be prohibitive.

Although the majority of true knot of the umbilical cord does not present with clinical significance, mainly because it is loose, there is an association between this finding and foetal intrauterine death. [1] It is known that true knot of the umbilical cord is associated with a four to ten times higher risk of foetal death. [2,3,5,19] According to Spellacy et al, [1] there was an increased incidence of abruptio placentae with cords around the body. There was a lower one minute Apgar score with these cord complications, but no difference was found in the 5 minute Apgar scores or in the one year neurological examination in comparison with the control group. The Apgar score was lower at one minute when the cords were pulled tightly around the neck than when they were loose. [1] In addition, 5% of all stillbirths are associated to true knot of the umbilical cord, [4] It can lead to serious complications for the foetus due to possible changes in intrauterine foetal circulation with subsequent intrauterine growth restriction or foetal death. [19] The risk of foetal distress is 7%. Prenatal diagnosis of true knot of umbilical cord is also associated to serious perinatal complications such as meconium-stained amniotic fluid. [5] Even though the obstetric outcome of Mrs I. G. was favourable, there was Abruptio placenta, foetal heart irregularity and low one minute Apgar score of 4. The Abruptio placenta could be from the nuchal cord and cord round body, mimicking a short cord which is a risk factor. Hence, the Emergency caesarean section was prompt and merited.

CONCLUSION

Currently, our experience on True umbilical cord knot is constrained to incidental postnatal finding. However, until prenatal diagnosis is improved with the use of ultrasonography for True umbilical cord knot, good clinical acumen, intrapartum surveillance and prompt obstetric intervention remain pivotal in the aversion of perinatal mortality.

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