OBSTRUCTED CONGENITAL HERNIA OF UMBILICAL CORD IN A NIGERIAN NEWBORN AND ITS DIAGNOSTIC CONUNDRUM

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ABSTRACT

Background: **Congenital Hernia of umbilical cord (CHUC) is a rare presentation. The failure of return of intestinal loops following physiological herniation has been postulated as the embryological basis of this condition. This is usually mistakenly termed as omphalocele minor. We aim to highlight this rare finding, the clinical features, diagnostic dilemma, and management of CHUC.**

Case Presentation: **He is a 4-day old male with obstructed CHUC. He had an oedematous umbilical stump, with circumferential normal abdominal wall skin folds in the form of a collar wrapping round the proximal aspect of the protrusion. The umbilical defect measured about 4 cm in diameter. Subsequently, surgical reduction and repair of defect was done, with resolution of symptoms.**

Conclusion: **CHUC though a rare entity and easily missed, accurate diagnosis and management offer good prognosis. Also, recognition will prevent iatrogenic bowel injury from careless clamping of the cord with associated morbidity and mortality. It also helps to prevent mis categorization and prevent unwarranted anxiety with misdiagnosis.**

Keyword: Congenital hernia, Physiologic herniation, Umbilical defect, Bowel injury

INTRODUCTION

Congenital hernia of the umbilical cord (CHUC), also known as congenital hernia of the cord or umbilical cord hernia, is a rare anomaly which is often misdiagnosed as omphalocele minor and an under-reported entity.[1–3] It is the rarest form of the anterior abdominal wall defects and results from failure of return of the physiologically herniated bowel loops. These bowel loops ideally should return into the abdominal cavity around 10 - 12 weeks of gestation. However, persistence of a patent umbilical ring predisposes to herniation of bowel into the umbilical cord. This is unlike omphalocele which results from failure of fusion of the four body wall folds and progression of the lateral body wall folds to the midline.[4,5] CHUC usually is not associated with any chromosomal or other organ anomalies unlike in omphalocele that could be syndromic.[4,5] It could be confused with cord hematoma, cord cyst, urachal cyst, giant cord or cord teratoma during antenatal scans and postpartum.[1,4] However, it is characterised by a proximal cord swelling, covered by a circumferential strip of normal skin which always enwraps the umbilical ring and a variable length of the proximal part of the cord.[4]

A number of complications can be associated with CHUC which include intestinal malrotation and obstruction.[4–7] Also, trauma to the intestines from accidental clamping of the cord over a herniating bowel loop due to uncharacteristic appearance or lack of awareness of CHUC among health care workers.[1,2] It is also thought that misdiagnosis could also cause unwarranted anxiety among parents. We report a case of obstructed congenital hernia of umbilical cord in a new-born male, highlighting its characteristic presentation thus creating awareness to prevent misdiagnosis and untoward complications of iatrogenic bowel injury.

CASE PRESENTATION

A 4-day-old male referred from a neighbouring state with protrusion from the umbilical area which was noted at birth with visible intestinal peristalsis. There was associated postprandial vomiting which was bilious, there was no fever and child had passed meconium on the first day of life. He is a product of term gestation, delivered via spontaneous vaginal delivery with good APGAR score and a birth weight of 3.8 kg at 38 weeks of gestation. Antenatal obstetrics ultrasound scans done noted umbilical anomaly suspected to be omphalocele and did not classify it further.

The child was afebrile, not pale, anicteric, but dehydrated at presentation. His vital signs were as follows: heart rate - 138 beats/minute, respiratory rate

- 48 /minute, oxygen saturation (SpO_2) – 98% in room air and temperature was 37.4^{0}C. The heart sounds were S_1 and S_2 only with no murmur, and the chest was clinically clear. The abdomen was distended, move with respiration; the umbilical stump was swollen with circumferential normal abdominal wall skin wrapping round the proximal aspect of the umbilical protrusion. The umbilical defect measured about 4 cm in diameter.

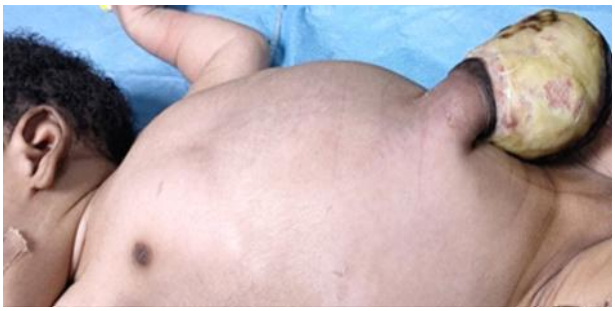

Figure 1: Showing normal abdominal wall skin enwrapping the proximal aspect of the umbilical stump with hernia contents

Bowel sounds were hyperactive. Perineal and digital examination were essentially normal. (Figure 1).

Blood count and chemistry revealed essentially normal levels of analytes.

He was commenced on intravenous fluid resuscitation with 4.3% dextrose saline, while on nothing by mouth. Empirical broad-spectrum antibiotics (ceftriaxone and metronidazole) were commenced. Sterile gauze soaked in warm normal saline, layered with Sofra-tulle® was used to wrap the umbilical stump. The patient subsequently had an emergency surgical repair under general anaesthesia after obtaining an informed consent from parents. Intra-operative findings were a 4cm umbilical defect which was extended by about 2 cm transverse incision to permit the reduction of the herniating bowel. The herniating viscera were loops of ileum, caecum, and ascending colon. The bowel loops were matted together by adhesions and amnion. (Figure 2). Adhesiolysis was done, the herniating viscera was found to be grossly normal with no atretic segment and was reduced into the abdomen. Anatomical closure of the ring was done with vicryl 0 sutures in a continuous fashion. (Figure 3).

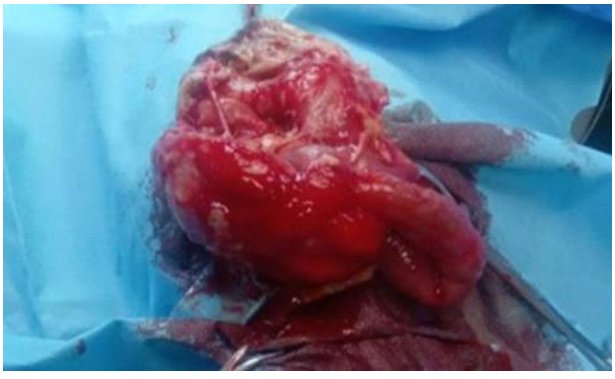

Figure 2: Intra-operative picture showing herniating bowel loops that are matted together with adhesions.

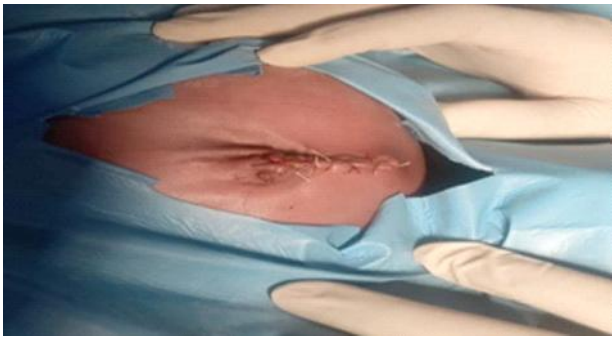

Figure 3: Post-operative picture after repair of CHUC.

He had good recovery from surgery, had satisfactory return of bowel function by post-operative day 3, was commenced on graded oral intake which was well tolerated and was discharged home on 5th post operative day.

The patient was followed up in the outpatient clinic and showed satisfactory wound healing with no complaints.

DISCUSSION

Power and D'Arcy were the first to report a case of CHUC in1888, and Tow in 1937 described the characteristics and embryogenesis of CHUC[4]. Compared to post-natal umbilical hernia, its congenital form (CHUC) is less common, with an incidence rate of 1 in 5000 newborns.[8] Till date only few cases have been reported, this may possibly be due to poor understanding and detection rate of CHUC.

Being both anatomically and embryologically different from omphalocele minor, umbilical cord hernia has the lowest incidence among the abdominal wall anomalies. Umbilical cord hernia appears later during embryonic development, usually after peritoneal cavity formation, unlike omphalocele that happens around the folding of the lateral folds in the earliest stages of embryonic development.[9]

During early fetal life, portion of the intestine lies in the proximal part of the umbilical cord by herniating into the extra coelomic cavity. The herniating intestine return into the abdominal cavity at about 10 to 12 weeks of gestation, and the umbilical ring narrows. Rarely, the umbilical ring remains wide, and portions of the intestine remain in the extra coelomic cavity,

Table 1: Differential diagnosis of congenital hernia of umbilical cord

Domains	Umbilical cord hernia	Omphalocele minor	Post-natal Umbilical he
Aetiology	Persistence of physiological mid-gut herniation beyond 10 - 12 weeks gestation	Small umbilical defect resulting from a failure of closure of the ventral abdominal wall	Herniation of the conten the abdomen secondary t delayed or failure of closu the umbilical ring
Stage of acquisition	12 - 14 weeks	3 - 5 weeks	Postnatal
Incidence	1 in 5000 Under-reported	1 in 4000	Very common; true incid unknown; increased incid in premature and African descent infants
Genetics	Sporadic	Mostly sporadic; few familial cases	Sporadic
Associated anomalies	Rare case reports of associated bowel anomalies and umbilical cord cysts	50 - 70% of them have other major anomalies	Rare
Abnormal karyotype	No association of abnormal karyotype with isolated presentation	30 - 40%	No association of abnorn karyotype with isolated presentation
Prenatal ultrasound findings	Small bowel extending into the base of the normally inserted umbilical cord	Foetal midline abdominal mass with the umbilical cord attaching at the apex of the mass	No herniation seen on prenatal ultrasound
Prognosis	Isolated presentations are considered to have a good prognosis	Depends on associated malformations and karyotype abnormalities	Good prognosis

thus presenting as CHUC. CHUC is different from omphalocele in that, omphalocele results mainly from failure of the four folds to meet at the midline during the formation of the anterior abdominal wall. The characteristic features of CHUC are: an intact umbilical ring and absent abdominal wall defect, extension of abdominal wall skin over the umbilical ring, enwrapping the proximal portion of the umbilical stump.[4,5] Omphalocele minor, on the other hand, is a condition in which a true abdominal wall defect is present.[10,11.]

There are four types of umbilical cord congenital hernia: Type 1 is a simple hernia into the cord without any associated complications; Type 2 is related to intestinal obstruction, and this typifies our index case; Type 3, a distinct variety associated with mucosal prolapse; and Type 4 is associated with evisceration.[12,13.] Associated anomalies in CHUC are rarer compared to omphalocele, identified anomalies are; congenital short gut, intestinal atresia, Meckel's diverticulum, volvulus and patent Vitello intestinal duct.[1,4,5,7,14] Till now, no underlying genetic disorders has been reported to accompany the cord hernia.[15] Table 1 highlights differences between CHUC, omphalocele minor and post-natal umbilical cord hernia. There were no associated anomalies in this index case, and this report aims to highlight the need for early recognition and management of CHUC as it was thought from the referring centre to be omphalocele minor. There are similar reports of misdiagnosed CHUC, and even inadvertent cord clamp placement on the hernia resulting in bowel ischemia and injury. Gasparella *et al.* reported a case of bowel injury from clamping of an unrecognized CHUC in a newborn.[16] In this index case, prenatal ultrasound scan noted an umbilical anomaly even though not classified and this knowledge may have aided caution in preventing accidental clamping of the herniating bowel at delivery, and prompt referral of the new born. The adhesions within the herniating bowel loops noted at surgery has been suggested by previous authors to probably be the reason for failure of return of physiologically herniated bowel loops into the abdominal cavity[1,4]. Also, the adhesions may account for the usual associated bowel obstruction noted with CHUC. Our patient had bilious vomiting

preoperatively with impression of obstructed CHUC, however this resolved following surgery.

CONCLUSION

The congenital umbilical hernia is a rare occurrence, with a good prognosis after surgical repair. If undiagnosed at delivery, bowel injury can occur by clamping the cord over the herniating bowel hence, adequate knowledge of this condition will prevent iatrogenic bowel injury and facilitate prompt intervention and prevent the sequelae of bowel obstruction as seen in this case.

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