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Case Report

Be vigilant before you clamp the cord: iatrogenic transaction of ileum - an avoidable catastrophe

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ABSTRACT

Congenital hernia of the umbilical cord (CHUC) is a rare congenital midline abdominal defect as compared to the umbilical hernia which is much more common and occurs post-natally. Despite its recognition as a distinct entity since 1920s, CHUC is often misdiagnosed as a small omphalocele, resulting in its under-reporting. These masses may be easily overlooked at birth, which may result in complications like infections and an intestinal injury due to careless proximal application of the cord clamp. Herein, we present a neonate who sustained accidental transaction of small bowel in congenital hernia of the umbilical cord due to inappropriate clamping of the cord. The case is reported in order to emphasize the potential hazard of clamping the bowel at the time of the handling of the umbilicus. It is imperative that extreme caution should be exercised by medical professionals and nursing staff prior to clamping the cord in order to avoid this type of injury. Such injuries are completely preventable if proper care and utmost precaution is taken during ligation of cord but can result in catastrophes otherwise. For the benefit of the readers the embryology of CHUC and differentiating features from other abdominal wall defects are also discussed.

Keywords: CHUC, umbilical hernia, Omphalocele, Gastroschisis, Ileal transaction

INTRODUCTION

At the time of birth, anterior abdominal wall defects can present either as gastroschisis or omphalocele. Gastroschisis presents with herniation of the fetal abdominal contents into the amniotic fluid without any covering. Omphalocele, on the other hand, is a congenital anterior abdominal wall defect at the base of an umbilical cord with herniation of the abdominal contents covered by the parietal peritoneum, the amnion and Wharton's jelly.[1] Mild forms of omphalocele can present as congenital hernia of the umbilical cord (CHUC) with herniation of either the solitary intestinal loop or persistent omphaloenteric duct (POD). The risk factors in omphalocele as well as CHUC are still not completely known and their association with other birth defects has not always been established.[1]

It is quite different from the post-natally diagnosed umbilical hernia and is believed to arise from persistent physiological mid-gut herniation. The incidence of CHUC is estimated to be 1 in 5000. Unlike an omphalocele, it is not linked with chromosomal anomalies but has been loosely associated with intestinal anomalies, suggesting the need for a complete fetal anatomical ultrasound evaluation.[2]

Traumatic injury to intestinal loops in congenital hernia of the umbilical cord has been reported previously and emphasized so as to prevent such injury at the time of birth.[3] This case is reported because of its very rare occurrence, and as a caution to all that such an accident may take place. In tying or clamping the cord, especially one that is unduly large, it should be tied well away from the abdomen.

CASE REPORT

A 8 days old male neonate with birth weight of 2 kg presented to us with fecal discharge from umbilicus since 10 hours after birth with provisional diagnosis of patent vitellointestinal duct. As per the parents the patient was passing small amount of stool per rectally for initial three days after birth but for the last 7 days the patient was not passing stool. There was no history of vomiting, fever, melena and hematemesis. The mother was not registered and did not take any antenatal visits or nutritional supplementation during pregnancy. The baby was delivered at home by a midwife. It was a full term normal vaginal delivery conducted by a midwife who clamped the umbilical cord stump about 2 centimetres from the umbilical base. The patient started passing stools from the umbilicus just 10 hours after birth owing to injury to the bowel which was herniating through the umbilicus (Figure 1). This was the result of applying the clamp at an improper site on umbilical stump by the midwife.

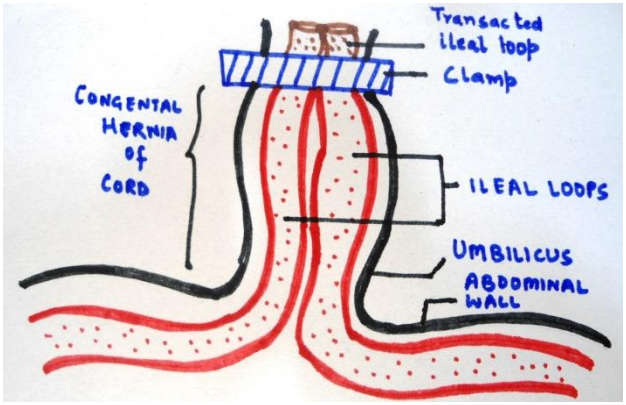

Figure 1: Diagrammatic representation of transaction of ileal loop by cord clamp in CHUC.

On examination the baby was pale with signs of dehydration and sepsis. On per abdomen examination two openings were visible at the umbilicus due to complete transaction of the bowel and fecal matter was coming out from one of them. Both of the bowel lumen could be catheterized separately. The abdomen was distended; soft, non-tender and fecal staining of finger was observed on per rectal examination. The external genitalia were normal and there were no other associated anomalies on clinical examination. Hemogram revealed anemia with elevated leukocytes. Blood urea and serum creatinine were raised with altered serum electrolytes. C-reactive protein was positive. Blood culture was sent just after admission that was reported later as positive for *Klebsiella* species. After resuscitation and optimizing the baby an exploratory laparotomy was performed by an infra-umbilical incision. The bowel loops were delivered after splitting of umbilical ring. Both the loops could be identified separately and catheterised (Figure 2). Proximal loop was dilated but not hypertrophied and the distal loop was narrow. Once distal patency was ascertained an ileo-ileal anastomosis was performed in single layer in interrupted fashion with 5-0 vicryl. The abdomen was closed in layers. Post-operatively packed cells and fresh frozen plasma was transfused along with ionotropic support. The baby had a stormy post-operative course. Unfortunately, the patient succumbed to sepsis on second postoperative day.

Figure 2: Intra–operative photograph with catheterisation of both ileal loops.

DISCUSSION

During early fetal life, a greater portion of the intestine lies in the proximal part of the umbilical cord, also known as the extra-celomic cavity. In normal course, the intestines withdraw into the abdominal cavity around 10-12 weeks of gestation, the umbilical ring closes and the extra-celomic cavity is thereby obliterated. In rare cases the umbilical ring does not close and in such instances variable portions of the intestines remain in the extra-celomic cavity, which persists as congenital hernia into the umbilical cord (CHUC).[4] The congenital hernia of cord is a distinct entity from other anterior abdominal wall anomalies such as gastroschisis and omphalocele. An intact umbilical ring, absent abdominal wall deficiency, presence of a sac comprising of an outer layer of amnion and inner peritoneum are the features distinguishing this entity from later two conditions. In addition, the contents which vary from loops of intestine to any movable intra-peritoneal organ (depending on the size of the defect) are also different from gastroschisis and omphalocele.[5] As a rule there is a cuff of skin from one half to one inch wide, which extends from the abdominal wall onto the neck of the sac.[4] Like all abdominal defects in which the midgut has not returned to the abdominal cavity before birth to allow for rotation and fixation, these patients also have malrotation, although it is not usually a cause of intestinal obstruction. Umbilical hernia is another distinct entity which needs to be distinguished from these anomalies. The diagnosis of umbilical hernia can be made by two features:

1. The defect is covered by normal skin.
2. It is only rarely present at birth, instead usually becoming apparent in the first weeks or months of life.

The incidence of CHUC is low .Tow had reported incidence of 1 in 5000 births compared to postnatal umbilical hernia, partly because most of these are misdiagnosed as "omphalocele minors."[2] Few early reports had shown male preponderance, association with prematurity and familial occurrence.[4] CHUC is usually a benign pathology, where, meconium discharge from the sac through an associated PVID may be found occasionally.[6] Cases with meconium stained liquor and meconium aspiration syndrome in a newborn with CHUC associated with Type III A ileal atresia and perinatal gut perforation have been reported.[7]

Prenatal ultrasound examinations play a vital role in the diagnosis of fetal anomalies and have thus influenced the management of new-borns at the time of delivery. Accurate prenatal diagnosis of anterior abdominal wall defects is generally possible during routine prenatal ultrasound examinations and these anomalies. Knowledge of characteristic ultrasound features facilitates therapy and prognosis.[1,8,9] Congenital hernia into the umbilical cord occurs at an early embryological stage and can be detectable by fetal ultrasonography as early as the second trimester.[10] Despite advances made in prenatal ultrasound diagnostics, CHUC, which is the minimal form of umbilical cord herniation, can go unnoticed. The presence of a single loop of intestine or POD may be quite challenging at the time of delivery.[1]

Delivery of uncomplicated pregnancies by midwives is common, and new-borns with CHUC pose a high-risk group for herniated structure injuries. These injuries may also go primarily unnoticed. It is only later, after careful evaluation, that their presence may be discovered.

Any unusual thickening of the base of the cord along with the minute fistulous opening to its side should alert the physician to the existence of this anomalies.[6] The cord in such patients must be clamped a safe distance away and an early paediatric surgical consultation must be contemplated. Further medical treatment or surgical intervention should be left to the discretion of paediatric surgeon. Neonatal surgical exploration is indicated to rule out associated intestinal atresia. It is noteworthy that timely surgical exploration and intervention in suspected cases helps to avoid complications and reduce morbidity.

CONCLUSION

Careful inspection of the umbilical cord of every new-born at the time of delivery is essential. Obstetricians, paediatricians and nurses who customarily clamp, manipulate, or shorten the cord in the delivery room or upon arrival in the nursery should be cognizant of this anomaly. Increased awareness and knowledge of such an entity among health professionals is of paramount important in preventing misdiagnosis, inadvertent bowel injury during cord clamping at delivery and associated morbidity and mortality.

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