Case Report

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Congenital hernia of the umbilical cord associated with extracelomic colonic atresia and perforation of gut in a newborn

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SUMMARY

Congenital hernia of the umbilical cord (CHUC) is a rare congenital entity compared to more common post-natally occurring umbilical hernia. Although recognized as a distinct entity since 1920s, CHUC is often misdiagnosed as a small omphalocele, resulting in its underreporting. We present the first case report of CHUC associated with extracelomic colonic atresia, complicated by perinatal perforation in a newborn. We also discuss the differentiating features from other anterior abdominal wall defects such as omphalocele and gastroschisis including its embryogenesis.

Key words: Colonic atresia, hernia of umbilical cord, neonate, perforation

INTRODUCTION

Congenital hernia of umbilical cord (CHUC) has been reported sparingly compared with more commonly occurring postnatal umbilical hernia. We report the first case of congenital hernia into the umbilical cord associated with extracelomic colonic atresia and perinatal perforation of gut in a newborn. We also discuss the differentiating features from other anterior abdominal wall defects such as omphalocele and gastroschisis including its embryo-pathogenesis.

CASE REPORT

This was a case report of a male preterm (35 weeks) baby weighing 1800 g ,born to 26-year-old Saudi mother (G3P2) by normal vaginal delivery. There was no history of maternal illness or any congenital anomalies in the family. On examination, the baby was found to have a swelling (5 cm × 4 cm) at the base of umbilical cord [Figure 1] with meconium exuding from a small defect on the swelling. Umbilical cord had normal vessels and attachment to the abdomen with a cuff of skin carried over to the sac. Loops of bowel could be seen through the membrane of the sac. The umbilical ring appeared complete and intact, but somewhat widened. There was no other gross congenital anomaly. Initial impression of congenital hernia into the cord with patent vitello intestinal duct (PVID) was made and was decided for surgical correction. On exploration sac was found to contain loops of distal ileum, ileo-cecal junction, appendix and ascending colon [Figure 2]. There was a perforation on ascending colon with hypoplastic distal bowel. A catheter passed through the perforation got stuck within 2 cm of the ascending colon suggesting a Type I colonic atresia [Figure 3]. Rest of gut was normal. Limited resection and ileo-ascending anastomosis was done after confirming distal bowel patency. Post-operative recovery was uneventful and the child is doing well at 2 years follow-up. Histopathology confirmed the existence of a Type I diaphragm in the ascending colon.

DISCUSSION

Embryologically, during early fetal life, a greater portion of the intestine lie in the proximal part of the umbilical cord, this is called the extracelomic cavity. Normally, the intestines withdraw into the abdominal cavity at about 10-12 weeks gestation, the umbilical ring closes and the extracelomic cavity is thereby obliterated. In rare cases the umbilical ring does not close and in these instances variable portions of the intestines remain in the extracelomic cavity, which persists as congenital hernia into the umbilical cord. Therefore hernia of cord is an entity distinct from other anterior abdominal wall anomalies such as gastroschisis and omphalocele. An

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Figure 1: Hernia into the umbilical cord with meconium discharge

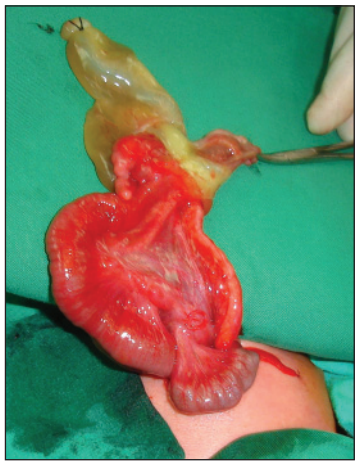


Figure 2: Sac containing dilated distal ileum, ileo-cecal junction, ascending colon with perforation and hypoplastic distal colon

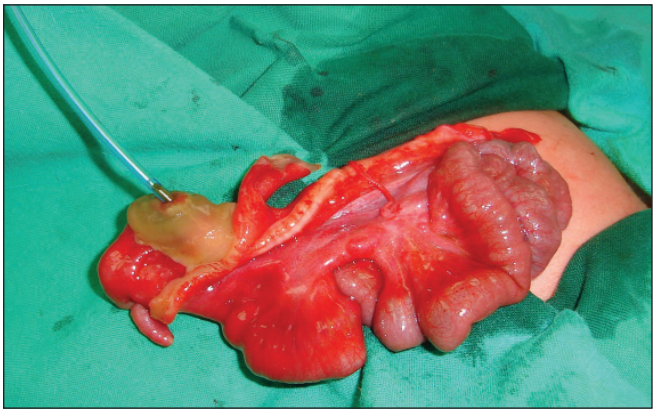


Figure 3: Perforation of the ascending colon with catheter stuck at Type I colonic atresia

intact umbilical ring, absent abdominal wall deficiency, presence of a sac comprising of an outer layer of amnion and inner peritoneum and contents varying from loops of the intestine to any movable intraperitoneal organ (depending on the size of the defect) are the features distinguishing this entity from later two conditions.[1]

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 as seen in our case [Figure 1]. The incidence of CHUC is low (Tow[2] had reported incidence of 1 in 5000 births) compared to postnatal umbilical hernia, partly because most of these are misdiagnosed as "omphalocele minors." Few early reports in 1920s to 1930s had shown male preponderance, association with prematurity and familial occurrence.[2,3] CHUC, usually a benign pathology, may occasionally have meconium discharge from the sac through an associated PVID.[4] We have reported a case with meconium stained liquor and meconium aspiration syndrome in a newborn with hernia of cord associated with Type III A ileal atresia and perinatal gut perforation.[5]

In our current case, there was meconium discharge from the sac at birth suggesting perinatal perforation of the gut. We had made an initial impression of PVID until exploration revealed a perforated Type I colonic atresia. We belief this rare association is being reported for the first time.

Frequent clinical instances of intestinal atresias due to intrauterine vascular accidents such as volvulus, intussusception, internal hernia, constriction of the mesentry in a tight gastroschisis or omphalocele defect and even callous umbilical cord clamping have been observed.[6-9] Therefore, we hypothesize that an early mesenteric vascular accident in the extracelomic herniated bowel could have caused this colonic atresia preventing the loops of the bowel to return to the celomic cavity and thus giving rise to congenital hernia of the cord.

Congenital hernia into the umbilical cord is a distinct anomaly occurring at an early embryological stage compared to omphalocele and gastroschisis and can be detectable by fetal ultrasonography as early as the second trimester.[10] One should be cognizant of such anomaly especially when there is a swelling at the base of the cord so as to avoid any iatrogenic injury to gut due to close umbilical clamping. Neonatal surgical exploration is indicated to rule out associated intestinal atresia.

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