Clinical Brief

Congenital Hernia of the Cord

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ABSTRACT

Several embryopathies involve umbilicus including midgut herniation, omphaloceles, urachal and vascular anomalies. Although described earlier, hernia into umbilical cord has not found enough mention in the literature. Poor understanding of its clinical characteristics has made to miscategorization of this entity as "omphalocele minor" by many. Some of these undergo spontaneous reduction or epithelialisation if left alone. The present study describes a series of four consecutive cases of this entity and describe their clinical characteristics, associated GI anomalies and possible embryogenesis.
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Umbilicus is a site of numerous embryopathies involving vessels, urachus, midgut herniation, anterior abdominal wall defects and congenital cysts. Hernia into the umbilical cord is an entity, has been poorly understood and often miscategorized as 'omphalocele minor'. Few small and reducible congenital hernias of cord are left as such by the clinicians which finally get epithelialised as 'cutis navel'. To the worst, inadvertent clamping of cord leads to iatrogenic gut injury in a situation of hernia of cord. We present a spectrum of clinical presentations and associations of this distinct entity in a series of four neonates managed in our department. We highlight the distinct clinical features and associated anomalies of this entity and explain its possible embryogenesis.

CASE REPORTS

Case 1

A preterm (34wk) male baby weighing 2100g born to a primigravida mother by normal vaginal delivery , noted to have a fleshy mass at the base of the cord measuring 1X0.5cm2 at 4 days of life (transferred from another hospital). There were no other gross congenital anomalies. Exploration of umbilicus revealed a 4 cm long Meckel's diverticulum herniating into the cord (Fig 1a, b). Patient was cured after excision of diverticulum.

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Case 2

A full term female baby weighing 2870g was noted to have a 2x2cm2 swelling at the base of the umbilicus at birth (Fig.2). There was no umbilical discharge. Also there was no other gross congenital anomaly. Initially a conservative management was maintained as the child was having neonatal sepsis. However the swelling did not subside and was explored on 10th postnatal age. A 3cms long Meckel's diverticulum was excised. Child went home after cure of sepsis and is doing well.

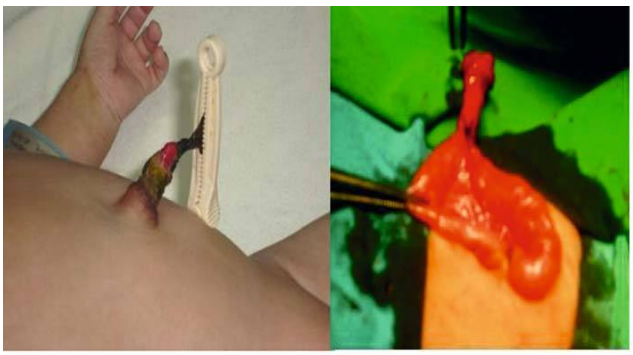

Fig. 1. Herniation of Meckel's diverticulum in case no. 1 undergoing excision.

Case 3

A preterm (35wk) male baby weighing 1800g born to a multipara women by normal vaginal delivery noted to have a 5x4cm2 swelling into the umbilical cord with a narrow base(2cm) and meconium discharge from a small opening on the swelling (Fig 3a). There was no other gross congenital anomaly or any maternal illness. Surgical exploration revealed herniation of distal ileum, ileo-cecal junction, ascending colon with type I colonic

atresia and peroration of proximal colon in the umbilical cord (Fig 3b).Child was treated by limited resection and ileo- ascending anastomosis.

Case 4

A male baby weighing 1750g at 33wk gestation was born to a multigravida mother with meconium stained liquor and meconium aspiration syndrome. Baby was noted to have hernia into cord with peforation of extracelomic gut and type IIIb ileal atresia in the hernia sac(Fig.4), reported by the authors earlier[1].Baby is doing well at 18months follow up following resection and anastomosis.

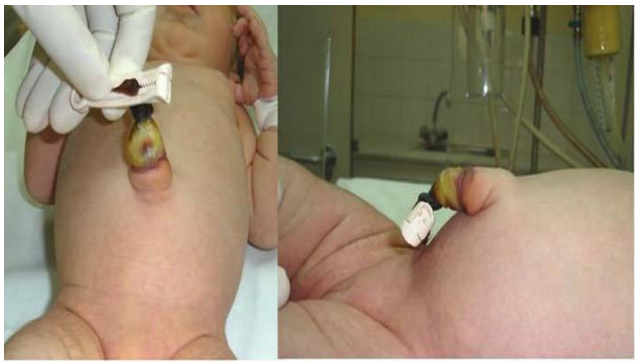

Fig. 2. Nonreducing hernia of cord in case no. 2 (day 5 of life) . Exploration on day 10 revealed a Meckel's diverticulum.

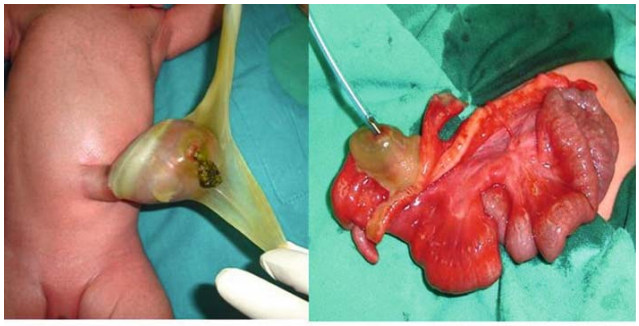

Fig. 3. Herniation of distal ileum, ileocecal junction and ascending colon with type I colonic atresia and perforation in case no.3

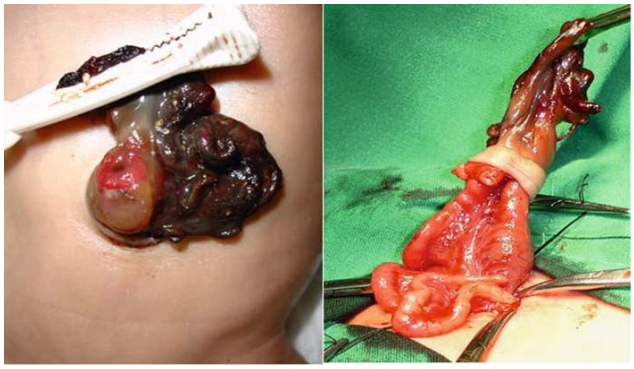

Fig. 4.Herniation of distal ileum with type IIIb atresia and perforation in case no. 4

DISCUSSION

During early fetal life there is physiological herniation of a greater portion of the intestines into the proximal part of the umbilical cord, which is called extracelomic cavity. At about 10-12 wks gestation, intestines withdraw into the abdominal cavity, the umbilical ring mostly closes and the extracelomic cavity disappears leaving behind Wharton's jelly and umbilical vessels in the cord. In rare instances, the umbilical ring does not close and variable portions of the intestines remain in the extracelomic cavity which present at birth as congenital hernia into the umbilical cord (CHUC)[2].

Unlike omphaloceles and gastroschisis, CHUC has an intact abdominal wall, a complete umbilical ring, a sac comprising of outer amnion and inner peritoneal lining and contains contents varying from loops of intestines to any movable intraperitoneal organs. Distinctively a cuff of skin is seen extending from abdominal wall onto the neck of the sac (Fig.1a, 3a). We encountered these distinctive features in all of our cases clearly establishing the diagnosis of CHUC.

Historically, in 1929,for the first time Hempel-Jorgensen[3] had reported two cases of this entity in a family and had coined the term 'familial congenital umbilical hernia'. Subsequently Tow in 1937[4] and Burn in 1938[2] described the characteristics and embryogenesis of this entity in couple of cases. Ever since only few case reports[1, 5, 6] have been published clearly describing this entity. Possibly poor understanding of CHUC has led to it's under reporting.

We have found a male preponderance (3:1) and association of prematurity (3 out of 4 cases) in our series akin to previous reports[3, 4]. Unlike omphaloceles, CHUC is believed to be a simple anomaly without any associated chromosomal or other organ involvements.

Persistent vitello intestinal duct (PVID) in one[5] and cloacal anomaly in another case[2] are the only reported associations so far. We have found two cases of Meckel's diverticula, one case of type I colonic atresia and one case of type IIIb ileal atresia as the associated GI anomalies in the series. Surgical exploration was done in all cases .

CHUC occurs at a specific embryological stage and the cause of failure of return of gut into celomic cavity is still obscure.Animal experiments and clinical scenarios have clearly described the etiopathogenesis of intestinal atresias. Intrauterine mesenteric vascular accidents due to volvulus, intussusception, and internal hernia, constriction of the mesentery in a tight gastroschisis or omphalocele defect have been observed[7-11]. It is quite possible that in two of our cases, such a vascular accident could have led to intestinal atresia and

persistence of intestinal loops in the extracelomic cavity as CHUC. In other two cases a persistent vitello intestinal duct structure (*e.g.,* Meckel's diverticulum) could have been the cause of CHUC. Therefore we hypothesise that the varied clinical presentation of CHUC may be multifactorial in causation (of arrested withdrawal of physiological herniation of gut).

Achiron *et al*[6] has demonstrated that CHUC occurs at early embryological stage and can be detectable at early 2nd trimester on antenatal USG. The present study did not have any of our cases being diagnosed by antenatal USG.

Cord hematoma, cyst, omphaloceles and gastroschisis etc. form the differential diagnoses for antenatally detected hernia of the cord. Few of these might disappear before term and rest may persist as CHUC. No specific antenatal intervention has been advocated although a regular follow up is required as cord hematoma carries increased risk of fetal death.

Therefore CHUC is a distinct entity with varying clinical presentations. Clinically it can be easily distinguished from omphaloceles and gastroschisis. Except in spontaneously reducing varieties, rest would require surgical exploration to rule out atresia or remnants of vitello-intestinal duct. Outcome is excellent due to lack of association with other congenital or chromosomal anomalies.

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