Case Series

Access this article online

Website: www.afrjpaedsurg.org

DOI: 10.4103/0189-6725.170231

Quick Response Code:

Congenital mesenteric hernia in neonates: Still a dilemma

Parkash Mandhan, Noora Alshahwani, Zainab Al-Balushi[1], Anwar Arain[1]

ABSTRACT

Congenital transmesenteric hernia in neonates is a rare cause of intestinal obstruction with devastating outcomes and still remains a challenge to diagnose pre-operatively. Patients are often managed with emergency surgical exploration and may need bowel resection. We present 2 neonates with small bowel obstruction secondary to strangulated transmesenteric hernia through a congenital defect in the small bowel mesentery, which were managed successfully. We have also reviewed the literature about congenital transmesenteric hernia in neonates.

Key words: Congenital mesenteric defect, internal hernia, neonates, small bowel obstruction, transmesenteric hernia

INTRODUCTION

Congenital transmesenteric hernia (CTMH), a herniation of viscera through an anatomic defect in the mesentery, is rare in new born and still a challenge and still a challenge for the paediatric surgeon to diagnosis pre-operatively. It usually presents with intestinal obstruction and is associated with high morbidity and mortality.[1] It represents 5-10% of all cases of internal hernias in children.[2] This entity creates a diagnostic dilemma as the clinical presentation is vague, and initial diagnosis is usually that of acute intestinal obstruction. The actual diagnosis of internal hernia is only established during surgery. A delay in diagnosis is lethal and can lead to death.

Department of Paediatric Surgery, Hamad General Hospital, Hamad Medical Corporation, Doha, Qatar, [1]Department of Paediatric Surgery, Division of Paediatric Surgery, Sultan Qaboos University Hospital, Al Khod, Muscat, Oman

Address for correspondence:
Dr. Parkash Mandhan,
Department of Paediatric Surgery, Hamad General Hospital, Hamad Medical Corporation, Doha, Qatar.
Email: kidscisurg@icloud.com

We report 2 cases of CTMH in neonates (Neonatal-CTMH) with a short history of bowel obstruction and on emergency exploration were found to have strangulated CTMH through a congenital mesenteric defect. We have also reviewed the literature with emphasis on early diagnosis in new born, followed by early surgical management to avoid high morbidity and mortality.

MATERIALS AND METHODS

Case 1

A 5-day-old baby girl was referred for progressively increasing abdominal distension, bilious vomiting, and not passing stool for 1-day. Her birth history was unremarkable she passed meconium on day-1 of her life. Physical examination showed distended abdomen with visible bowel loops in the upper abdomen. A plain X-ray of the abdomen was suggestive of proximal small bowel obstruction. After initial resuscitation, an upper gastrointestinal (GI) contrast study was planned, but due to deterioration of her clinical condition, an emergency laparotomy was carried out considering acute small bowel obstruction secondary to volvulus of bowel. On exploration, there was a 2-cm mesenteric defect in distal ileal mesentery through which a segment of jejunum was herniated, which was mildly dusky [Figure 1]. The rest of her bowel was normal. The entrapped bowel was released, and mesenteric defect was closed. Her postoperative course was uneventful, and she has remained well after surgery.

Cite this article as: Mandhan P, Alshahwani N, Al-Balushi Z, Arain A. Congenital mesenteric hernia in neonates: Still a dilemma. Afr J Paediatr Surg 2015;12:203-7.

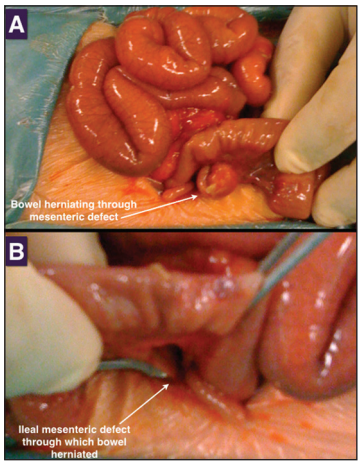

Figure 1: Intraoperative finding (of case 1) showing bowel herniated through a mesenteric defect (a) and a defect in the ileal mesentery through which loops of bowel herniated (b)

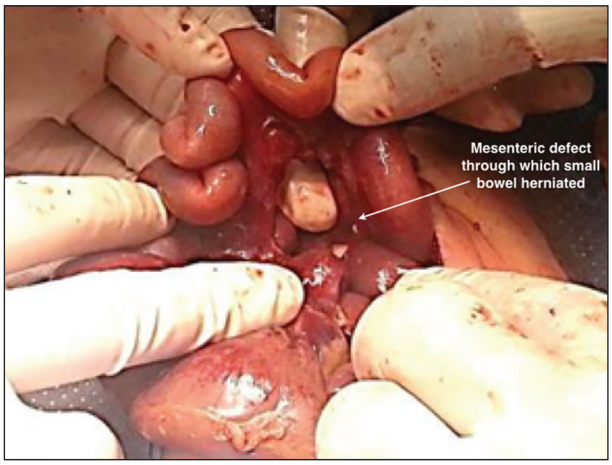

Figure 2: Intraoperative photograph (of case 2) after reduction of the hernia contents showing congenital mesenteric defect in the mid ileum through which loops of small bowel herniated resulting gangrenous bowel

Case 2

A 5-h-old baby boy was referred with tachypnoea and decrease activity. His birth history was unremarkable, and his Apgar score was 8 and 10 at 1 and 5 min, respectively. On examination, baby had stable vital signs and mildly distended abdomen. A plain abdominal X-ray at 8-h of age showed small bowel pushed towards the left side with few air fluid levels. He received initial management with nil per oral, orogastric tube, intravenous fluids and antibiotics. During next few hours, he developed further abdominal distension associated with high and bilious orogastric output and required elective intubation. Further deterioration in his clinical assessment prompted for early surgical intervention. Exploratory laparotomy revealed peritoneal cavity full with meconium and a mesenteric defect in terminal ileal mesentery [Figure 2] through which a segment of the mid-ileum was herniated, which was ischemic and had multiple perforations. After releasing the entrapped small bowel, resection of ischemic bowel segment was carried out followed by primary anastomosis, closure of the mesenteric defect, peritoneal wash and wound closure. Patient's postoperative course was slow and steady. He was discharged home after 3-weeks on full feeds and since then has remained well with normal growth and development.

To conduct the literature review, PubMed search was performed using words "congenital internal hernias," "transmesenteric hernia," "intestinal obstruction," and "mesenteric defect," separately and in different combinations, limiting it to neonates (i.e., <1-month of age). Additional studies were recruited from the reference list cited.

DISCUSSION

The literature review excluding patients older than 1-month or where age was not specified yielded total of 15 cases of CTMH from 1950 to 2014 [Table 1]. Addition of our 2 cases will make a total of 17 cases Neonatal-CTMH.

Preoperative diagnosis of Neonatal-CTMH has remained a challenge as the clinical presentation often occurs as bowel obstruction.[3-8] The bowel obstruction may progress rapidly to strangulation and shock. Bowel ischemia was reported as soon as 6 h after onset of symptoms.[1] The role of imaging modalities in accurate diagnosis of Neonatal-CTMH is limited as there is no specific modality to support its diagnosis prior to surgical exploration. Hence, the definitive diagnosis is reached at surgery as happened in our cases and also reported by others.[3-8]. The role of contrast study is limited, as it will not yield the specific diagnosis, it also has the potential of radiation exposure and hypothermia in neonates and finally it leads to delay in the surgical exploration. In some reported cases of Neonatal-CTMHs, associated bowel anomalies such as jejunal/ileal atresia, intestinal duplication, Hirschsprung's disease, and malrotation have been reported.[9] Even in the presence of associated GI anomalies, diagnostic modalities may prove of little help to diagnose Neonatal-CTMH as the underlying cause of acute bowel obstruction. Hence in presence of bowel obstruction, regular clinical assessment and further progress of symptoms of the new born babies remains the best possible guide to decide further course of management. In our cases, the decision of early surgical exploration was taken due to clinical deterioration of neonates and the absolute diagnosis was established at surgery.

Table 1: Characteristics of 17 neonates with CTMH reported in the literature from 1950 to 2014

Author	Age/sex	Presentation	Radiological findings	Location of defect	Operative findings	Intervention	Outcome
May and Brintnall 1953[14]	3 days/male	—	—	—	Meconium peritonitis; mesenteric hernia with gangrenous bowel	Resection and anastomosis	Survived
Tow *et al.*[3]	2.5 h/male	Abdominal distension, difficult delivery; left humerus fracture	—	Proximal ileal mesentery	Meconium peritonitis with herniated jejunum with gangrene and perforation	Resection and loop jejunostomy	At 1-year-Intestinal obstruction requiring resection and re-anastomosis
Murphy[9]	<1-day/male	Multiple intestinal perforation	—	Proximal ileum	—	Laparotomy	Death
	4 days/female	Premature, RDS	—	Terminal ileum	Ileal atresia, with herniated proximal ileum; intestinal duplication	Miculicz resection, excision of duplication	Death
	1-day/female	—	—	Mid intestine	Illeal atresia, with herniated strangulated jejunal loops; incomplete left colon rotation; persistent superior vena cava; double ureter and pelvis; left kidney	Miculicz resection	Death
	2 days/female	Premature	—	Mid intestine	Jejunal atresia with blind end jejunum twisted around the hernia defect	Miculicz resection	Survived
	36 h	—	—	Mid intestine	Jejunal atresia, strangulation and volvulus of herniated distal ileum, and meconium peritonitis	Laparotomy	Death
	4 days/male	Premature	—	Terminal ileum	Ileal atresia with herniating ileal loops; malrotation if bowel; ventral pancreatic anlage; accessory spleen	Ileo-ileostomy, Ladd's procedure	Death
	2 days/male	—	—	Terminal ileum	Ileal stenosis, with herniating proximal ileal loops; Hirschsprung's disease	Ileo—transverse colostomy	Death
	2 days/female	Mongolian features	—	Proximal ileum	Ileal atresia, herniating distal jejunum and ileal loops with the strangulation; incomplete large bowel rotation; VSD; accessory spleen	Operation refused; supportive treatment	Death
Fan *et al.*[7]	1-day/male	Bilious emesis	UGIS: Dilated stomach and duodenum with near complete obstruction	Terminal ileal mesentery	Herniated bowel	Reduction of bowel an repair of defect	Survived
	4 days/male, (twin)	Abdominal distension/ bilious vomitus	AXR: Persistent dilated loops	30 cm from ICV (defect size 4 cm)	Herniated small bowel (viable), dusky terminal ileum (are with mesenteric defect)	Resection of the terminal ileum, appendectomy, ileostomy and mucous fistula created	Survived
Nouira *et al.*[5]	1-day/male	Abdominal distension/ tachypnoea	AXR: Nonspecific abdominal gas distribution	30 cm from ICV (defect size 5 cm)	Herniated bowel, gangrene	Resection and anastomosis, and repair of defect	Survived
Sato *et al.*[4]	1-day/male	Bilious vomiting, abdominal distension	AXR: Dilated small bowel US: Fluid filled small bowel loops	—	Herniated small bowel, with torsion, thrombosed mesenteric veins	Resection of 20 cm of small bowel with end-to-end ileal anastomosis. Closure of the defect with abs sutures	Survived

(Continued)

Table 1: (Continued)

Author	Age/sex	Presentation	Radiological findings	Location of defect	Operative findings	Intervention	Outcome
May and Brintnall 1953[14]	3 days/male	—	—	—	Meconium peritonitis; mesenteric hernia with gangrenous bowel	Resection and anastomosis	Survived
Mandhan *et al.* (current cases)	5 days/female	Bilious vomiting, abdominal distension	AXR: Nonspecific bowel gases	15 cm from ICV (defect size 2 cm)	Segment of jejunum herniating through defect, dusky in colour	Release of entrapped bowel segment	Survived
	5 h/male	Bilious vomiting, abdominal distension	AXR: Nonspecific bowel gases	30 cm from ICV (defect size 3 cm)	15 cm of the mid-ileum herniating through defect, with torsion, gangrene and perforation	Resection of affected bowel and end-to-end anastomosis	Survived

RDS: Respiratory distress syndrome; VSD: Ventricular septal defect; UGIS: Upper gastrointestinal series; AXR: Abdominal X-ray; ICV: Intra-cerebroventricular; US: Ultrasound; CT: Computed tomography, CTMH: Congenital transmesenteric hernia

The outcome of Neonatal-CTMH depends greatly on index of suspicion of the diagnosis, followed by surgical intervention as a rapid progression from incarceration to strangulation of the bowel can happen, which may lead to sepsis and death. Janin *et al*. have reported high morbidity and mortality in their series[1] and in Murphy series of seven neonates; six new born died [Table 1].[9] In our cases, our early surgical intervention was helpful, but yet in one case, the herniated bowel segment was already infarcted, and baby required bowel resection. This highlights that a high index of suspicion and an early surgical intervention may be helpful to avoid the high-risk morbidity and mortality in such cases.

In CTMH, bowel loops herniate through a defect in the mesentery in the absence of a hernia sac. In majority of cases a mesenteric defect of about 2-5 cm in diameter has been noted in the ileal mesentery, but defect may also occur in the fold of treves,[10] and at or near to the duodeno-jejunal junction[1] leading to internal hernia and its sequel. In literature review of 13 Neonatal-CTMHs, the anatomical site of congenital transmesenteric defect included proximal jejunum (1), proximal ileum (3), mid ileum (3) and terminal ileum (7) as showed in Table 1. The association of small bowel atresia and other systemic anomalies in Neonatal-CTMH supports the congenital aetiology of this defect, which may happens during embryogenesis possibly due to a vascular insult, incomplete tabularization, or defect occurring during the return of bowel and rotation process. The existence of a small or large congenital mesenteric defect leaves behind a potential risk of bowel herniation and its sequel, which may happen early in neonatal period as observed in few reported cases including ours, or it may result in internal hernia later in life.[11-13]

In summary, it is difficult to make an accurate preoperative diagnosis of Neonatal-CTMH. The definitive diagnosis and localization is made intra-operatively. An early surgical consult is of great importance to reduce the risk of further morbidity and mortality.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Janin Y, Stone AM, Wise L. Mesenteric hernia. Surg Gynecol Obstet 1980;150:747-54.

2. Page MP, Ricca RL, Resnick AS, Puder M, Fishman SJ. Newborn and toddler intestinal obstruction owing to congenital mesenteric defects. J Pediatr Surg 2008;43:755-8.
3. Tow A, Hurwitt ES, Wolff JA. Meconium peritonitis due to incarcerated mesenteric hernia: Recovery following operation for intrauterine rupture of intestine. AMA Am J Dis Child 1954;87:192-203.
4. Sato T, Abe S, Tsuboi K, Iwata M, Tamura A, Tsuchihashi H, *et al*. Sudden death of a child because of an intestinal obstruction caused by a large congenital mesenteric defect. Leg Med (Tokyo) 2012;14:157-9.
5. Nouira F, Dhaou BM, Charieg A, Ghorbel S, Jlidi S, Chaouachi B. Small bowel obstruction caused by congenital transmesenteric defect. Afr J Paediatr Surg 2011;8:75-8.
6. Malit M, Burjonrappa S. Congenital mesenteric defect: Description of a rare cause of distal intestinal obstruction in a neonate. Int J Surg Case Rep 2012;3:121-3.
7. Fan HP, Yang AD, Chang YJ, Juan CW, Wu HP. Clinical spectrum of internal hernia: A surgical emergency. Surg Today 2008;38:899-904.
8. Elmadi A, Lechqar M, Biache I, Tenkorang S, Khattala K, Rami M, *et al*. Trans-mesenteric hernia in infants: Report of two cases. J Neonatal Surg 2014;3:39.
9. Murphy DA. Internal hernias in infancy and childhood. Surgery 1964;55:311-6.
10. Chan KT. Transmesenteric hernia. Singapore Med J 1963;3:34-7.
11. Umaparan G, Nallusamy K, Abdul Wahab MH, Mohammad Nor AM, Mat Arif NA. Congenital trans-mesenteric herniation: A rare cause of small intestine strangulation in adults. J Surg Case Rep 2012;2012(11). pii: rjs003. doi: 10.1093/jscr/rjs003.
12. Llore N, Tomita S. Apple peel deformity of the small bowel without atresia in a congenital mesenteric defect. J Pediatr Surg 2013;48:e9-11.
13. Butterworth J, Cross T, Butterworth W, Mousa P, Thomas S. Transmesenteric hernia: A rare cause of bowel ischaemia in adults. Int J Surg Case Rep 2013;4:568-70.
14. May SC, Brintnall ES. Strangulated transmesenteric hernia in an erythroblastotic newborn; (report of a case treated by resection and anastomosis with survival). Surgery 1953;33:312-4.