CASE REPORT

Neonatal obstructed Treitz's hernia with abdominal cocoon simulating volvulus neonatorum

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Ramnik V Patel,
ramnik@doctors.org.uk**SUMMARY**

A case of congenital obstructed Treitz's hernia presenting with bilious vomiting in a newborn baby girl has been presented. Internal herniation of contents within a peritoneal sac of the right paramesocolic hernia formed abdominal cocoon which simulated volvulus neonatorum. Plain radiographs and contrast studies were helpful in defining the nature and extent of the lesion. The patient underwent exploratory laparotomy, reduction of small bowel contents from the hernial sac forming an abdominal cocoon, Ladd's procedure to correct associated midgut malrotation with incidental appendicectomy uneventfully and recovered well.

BACKGROUND

Treitz's hernia is the eponymous name for a para-duodenal hernia which has been more recently called paramesocolic hernia (PMH).¹ These are rare internal congenital hernias that arise in the potential spaces and folds of the posterior parietal peritoneum adjacent to the ligament of Treitz. They usually present later in life with partial obstruction and presentation in the neonatal period with obstructed right paramesocolic hernia (RPMH) with a sac forming an abdominal cocoon which is an extremely rare event. Clinically, it was suspected as an acute abdomen with palpable mass and we wish to present the clinical and imaging aspect of this rare and interesting internal hernia.

CASE PRESENTATION

A baby girl was born at term after an uneventful pregnancy and normal vaginal delivery. She required no resuscitation at birth. At 1 week of age she presented to the local district general hospital (DGH) with persistent bilious vomiting and fullness in the upper abdomen.

The baby continued to have bilious vomiting and was therefore referred to us after stabilisation. On arrival, she had dark green aspirates in her nasogastric tube but she was alert and active with soft and non-tender abdomen. She was very stable and well clinically and did not require any resuscitation at our hospital.

INVESTIGATIONS

Complete blood count and biochemical investigations including renal and liver functions were normal. Plain abdominal radiograph showed dilated gas filled stomach and normal left colonic and rectosigmoid gas (figure 1A). She underwent upper gastrointestinal series at DGH which showed dilated stomach and proximal duodenum which was sharply

changing calibre to the right and over the spine and a small trickle of contrast in the small bowel loops could be seen (figure 1B). It clearly showed a misplaced duodenojejunal flexure, which is low lying and further to the right than normal. On delayed films at 1.25 and 12 h after the examination, there was prominent gastric and duodenal dilation proximally, further contrast progression into the loops of small bowel lying mostly in the centre of the abdomen outlining the small bowel loops in the RPMH sac forming the cocoon (figure 2A,B).

The plain abdominal radiograph on arrival to our hospital clearly showed most of the contrast in the large bowel and a loop of the terminal ileum to the left of the spine in the left upper abdomen (figure 3A). We felt that it was abnormal but wanted to confirm the dynamic aspect and the position of the duodenojejunal flexure in relation to pylorus. Repeated upper gastrointestinal series examination showed a distended stomach and a proximal duodenum, with total obstruction in the third part of the duodenum (figure 3B,C). An attempt was made for abdominal ultrasound and colour Doppler for the superior mesenteric vessels to see the relationship between artery and vein in the pedicle for midgut malrotation evidence but due to gas and contrast in the bowel, it was inconclusive. A contrast enhanced abdominal CT is a gold standard in adults and helps to confirm the diagnosis. Clear indications of exploration in view of complete duodenal obstruction and associated midgut malrotation; availability of theatre slot for emergency operation and concerns of paediatric radiology team for the radiation dose involved in the procedure led us to proceed with exploratory laparotomy as the next investigation with a view to have therapeutic surgical interventions as required after exploration. All these investigations of plain abdominal films, contrast upper gastrointestinal series and associated follow-through studies and limited ultrasound and colour Doppler studies indicated RPMH with a sac forming an abdominal cocoon and having associated midgut malrotation.

DIFFERENTIAL DIAGNOSIS

A spectrum of congenital duodenal obstructions including duodenal web, duodenal stenosis and Ladd's bands as intraluminal, intramural and external compressions and internal herniation other than the PMH and through the mesenteric defects were all considered in the differential diagnosis. Midgut malrotation with volvulus neonatorum was considered but clinically the baby was so well and mass so soft, it appeared unlikely. The detailed

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Figure 1 (A) Plain abdominal film showing dilated stomach (S) bubble and proximal duodenum (D) in the upper abdomen, central conglomeration of bowel loops with less dense step ladder pattern indicated by arrows and gas in the rectum, sigmoid colon (SC), left colon (LC) and distorted dilated abnormally placed caecum (C) after correlating with intraoperative findings. (B) Contrast study film at 15 min showing extrinsic obstruction to duodenum with dilated stomach (S) and proximal duodenum (D) leading to a trickle of contrast into proximal small bowel—long thick arrow followed by negative air contrast bowel loops in the hernial sac indicated by thin arrows finally leading to distorted and displaced bubble of air in the caecum (C) leading to splenic flexure (SF).

review of the investigations and repeat upper gastrointestinal series confirmed the malrotation; the internal herniation with peritoneal sac forming an abdominal cocoon and complete obstruction needed an exploration to correct it and therefore a contrast-enhanced CT was not performed.

TREATMENT

Examination under anaesthesia showed soft mass in the central abdomen. At exploratory laparotomy, partial peritoneal encapsulation of upper small bowel owing to herniation of jejunal loops into the RPMH sac was evident. There was incomplete rotation of the midgut with the caecum lying in the left upper quadrant with severe congestion and angulation of the proximal jejunum lying in the hernial sac posteriorly under the mesocolon. There was lymphatic congestion of the mesentery and little free chylous fluid at the base of the mesentery.

Small bowel loops were reduced into the main peritoneal cavity which corrected the lymphatic congestion. Apart from the lymphatic congestion rest of the bowel was well perfused and no resection was required. Ladd's procedure was performed with widening of the base of the mesentery which opened up the neck of the hernia widely. Small bowel was returned on the

right side of the abdomen, the colon on the left side in a non-rotation fashion with an incidental inversion appendectomy concluded the operation.

OUTCOME AND FOLLOW-UP

Her postoperative period was uneventful and she made a good recovery. She was discharged home on fifth postoperative day. Histopathology confirmed normal histology of the appendix and that of the cocoon wall showed the peritoneal sac with congestion at the mesenteric root. At 30-month follow-up she is asymptomatic and thriving well.

DISCUSSION

Treitz's hernias arise in the potential spaces and folds of the posterior parietal peritoneum adjacent to the ligament of Treitz. The PMH are unfamiliar complex variety of intra-abdominal congenital internal hernias.² They are classified into RPMH or left (LPMH) hernia, depending on embryological origin and anatomical features. They result from abnormal rotation and fixation of the midgut as an intraperitoneal internal congenital hernia. RPMH results from the absence of rotation of prearterial segment of the small bowel anterior to the superior

Figure 2 (A) Delayed supine 1 h and 15 min film showing dilated stomach and proximal duodenum with afferent loop (downward thick arrow), the opacified bowel loops in the hernial sac (outline with thin arrows) and an efferent loop (upward pointing thick arrow) followed by combined double contrast-positive contrast and negative air contrast into the splenic flexure, left colon (LC) and sigmoid colon (SC) and rectum (R). (B) Delayed supine 12 h film showing dilated stomach (S), proximal duodenum (D), almost gasless cocoon and very prominent and clear terminal ileal loop finally emerging as the efferent loop (EL) in left upper quadrant and complete opacification of the left-sided large bowel loops.

Figure 3 Repeat plain and upper gastrointestinal contrast study in supine, anteroposterior and lateral views. Note sharp cut-off of the afferent loop (thick right arrow), small bowel mass effect without any gas in it (the small arrows), trickle of contrast in efferent loop (thick left arrow), caecum in left upper abdomen and large bowel pushed to the left. Note the afferent loop to the right of the spine and the level well below the pyloric plane on the oblique lateral view.

mesenteric vessels. In LPMH, the entire intestine rotates normally but there is failure of fixation of the left mesocolon to the posterior abdominal wall in normal fashion.

Despite the congenital nature, diagnosis is usually made in adulthood, with a mean age of 38 years.² They are more common in men. LPMH is three times commoner than RPMH in both sexes. Treitz's hernias present potential risk for entrapment, incarceration, obstruction and strangulation. Specific clinical signs are often absent, leading to the frequent delay of correct diagnosis; with bowel compromise resulting in up to 20% of patients. The presentation may vary from subtle and easily overlooked findings on physical examination to massive abdominal distension with respiratory distress and cardiovascular collapse.³ Presentation may be acute with small intestinal obstruction, strangulation or perforation, or with chronic intermittent abdominal pain. Although internal hernias may have a congenital aetiology, most of the reported cases have occurred in adulthood; presentation in childhood is uncommon in general and exceptional in neonates.

Abdominal cocoon is an unusual form of presentation of the PMH in children and adolescents. Abdominal cocoon is an unusual small-bowel obstruction caused by a membrane encapsulating the small intestine in the manner of a cocoon in the internal hernia with a peritoneal sac.²⁻⁴ The primary cocoon is congenital in origin and is very rare, most cases are secondary to disease or drugs and our case lends support to congenital origin in neonates and children. A rare presentation with obstructive jaundice has been described.⁵ Internal hernias are uncommon in children and their clinical and imaging findings have not been widely addressed in the literature.⁶

Early diagnosis depends largely on the prompt detection of obstructive manifestations by the clinician and subsequent accurate interpretation of radiographic findings and other investigations including pain and contrast studies and ultrasound with colour Doppler studies, leading to definitive treatment, which should always be preceded by appropriate resuscitation/preparation of the infant/child.³

There is a wide spectrum of imaging findings of PMH as outlined below in children, and paraduodenal hernias were easy to appreciate. One must have a high index of suspicion for right

paraduodenal hernia if the upper gastrointestinal series shows the duodenum and proximal small bowel to the right of the spine.⁶ In neonates, plain film provides very good background negative contrast of air and although based on features of haustra it is difficult to identify the large bowel but the contrast of air in stomach and duodenum and colon and rectum is darker as compared with small bowel (figure 1). We follow the anatomical landmarks of the stomach and duodenum and that of the rectum and left colon which are more consistent at any age.

In our case only the first film of contrast was performed under fluoroscopy which showed very clearly the afferent loop, the 1.25 h film demonstrated clear contents of the hernial sac in the form of a cocoon and the delayed 12 h film has clearly outlined the efferent loop and all of the large bowel was on the left side of the spine with the caecum and appendix in the left upper quadrant. Repeat dynamic upper gastrointestinal tract contrast demonstrated that the duodenojejunal flexure was over the spine and it was very much lower than the pyloric level.

Treatment is by surgery, owing to the high (50%) lifetime risk of obstruction. Laparoscopic repair has been reported in adults.⁷ Operative mortality can exceed 20% especially in strangulation and gangrene.⁸

Learning points

- ▶ Neonatal presentation of right paramesocolic hernia is very rare and has not been reported.
- ▶ Formation of abdominal cocoon by internal herniation forming a soft non-tender mass simulated volvulus neonatorum with malrotation which was a unique feature in our case.
- ▶ Initial radiological study at the referring hospital was reported having no malrotation but review of films and repeat study helped resolve the problem.
- ▶ Abdominal cocoon with internal herniation should be suspected if there is soft non-tender mass and there is associated midgut malrotation with partial or complete obstruction on radiographs.

Contributors All the authors have been involved actively in the clinical management of the case and in the collection, analysis and interpretation of data as well as preparation, criticism and finalisation of the manuscript of this article.

Competing interests None.

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