

Congenital Internal Hernia Presented with Life Threatening Extensive Small Bowel Strangulation

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Internal hernia (IH) is a rare cause of small bowel obstruction occurs when there is protrusion of an internal organ into a retroperitoneal fossa or a foramen in the abdominal cavity. IH can be presented with acute or chronic abdominal symptom and discovered by accident in operation field. However, various kinds of imaging modalities often do not provide the assistance to diagnose IH preoperatively, but computed tomography (CT) scan has a high diagnostic accuracy. We report a case of congenital IH in a 6-year-old boy who experienced life threatening shock. CT scan showed large amount of ascites, bowel wall thickening with poor or absent enhancement of the strangulated bowel segment. Surgical exploration was performed immediately and had to undergo over two meters excision of strangulated small bowel. To prevent the delay in the diagnosis of IH, we should early use of the CT scan and take urgent operation. (**Pediatr Gastroenterol Hepatol Nutr 2013; 16: 190~194**)

Key Words: Intestinal obstruction, Hernia, Child

INTRODUCTION

Internal hernia (IH) is defined as herniation of viscera through a normal or abnormal aperture within the peritoneal cavity. Acute intestinal obstruction due to IH is quite unusual, representing 0.5 to 5.8% of reported intestinal obstruction [1]. However, IH is associated with high mortality of up to 50% in the certain series [1]. It has been demonstrated that they often remain undiagnosed before emergency laparotomy since the symptoms of IH may be nonspecific

ranging from mild abdominal discomfort to sudden onset intestinal obstruction. Furthermore, it occasionally leads to gangrene necessitating bowel resection of varying extent which may contribute to high mortality [2]. There are several types of congenital IH, which are classified according to their locations as follows; foramen of Winslow, paraduodenal, pericecal, transmesenteric, transomental, and intersigmoid hernias [2]. The most common types that present in children are paraduodenal and transmesenteric hernias [3]. Herein we report a case

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of congenital transmesenteric IH presenting with life threatening extensive small bowel strangulation.

CASE REPORT

A 6-year-old boy visited pediatric emergency department due to sudden onset abdominal pain which developed 1 hour ago. He had complained diffuse abdominal pain and vomited clear fluid twice in the hospital. Vital sign was stable and the physical examination revealed dehydrated tongue and diffuse abdominal tenderness. Laboratory test showed unremarkable except elevated white blood cell count of $24,310/\text{mm}^3$ with normal differentiation. Plain abdominal radiograph showed nonspecific small bowel gas which suggested mild ileus (Fig. 1) and abdominal ultrasound revealed diffuse small bowel wall thickening with ascites (Fig. 2). A contrast-enhanced abdominal computed tomography (CT) was recommended, but his father refused to undergo the further evaluation and discharged against our medical advice.

Fifteen hours later, he was rushed by ambulance to the pediatric emergency department again with

altered mentality. He did not respond to the painful stimuli, his both pupils were dilated to 5mm and showed severe abdominal distension. His blood pressure was uncheckable with weakly palpable femoral pulse, percutaneous oxygen saturation was 77% and blood sugar level was 22 mg/dL. Pretibial intraosseous cannulation was performed urgently due to failure of the intravenous catheterization and fluid and medications were infused through the intraosseous cannulation. He was in shock state and arterial blood gas analysis showed pH of 7.15 and bicarbonate of 7.7 mM. Hemoglobin and hematocrit levels were 10.1 g/dL and 27.7%, but rapidly decreased to 4.7 g/dL and 13% respectively, just one hour later. Also he was in disseminated intravascular coagulation state with antithrombin III of 38.9%, D-dimer of 9.18 mg/L, prothrombin time of 1.61 international normalized ratio, activated partial thromboplastin time of 43.9 seconds and lactic acid of 10.9 mmol/L. After the initial hydration, central venous catheter could be placed via the left subclavian vein and massive fluid, transfusions, and inotropics could be delivered. The contrast-enhanced abdominal CT scan performed immediately after maintaining systolic blood pressure of 90 mmHg. CT scan showed large amount of ascites, bowel wall thickening with

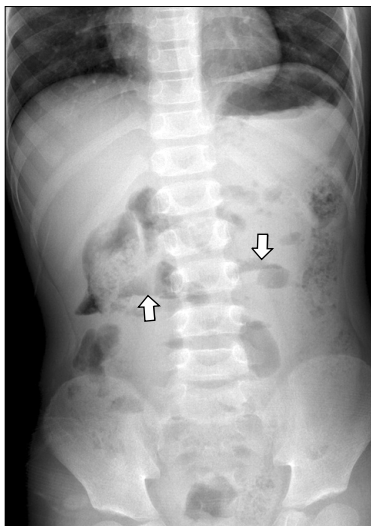


Fig. 1. Initial erect abdominal plain radiograph shows several irregular small bowel air without definite small bowel dilatation. Note two bowel loops with air-fluid level (arrows). These findings are abnormal but not specific.

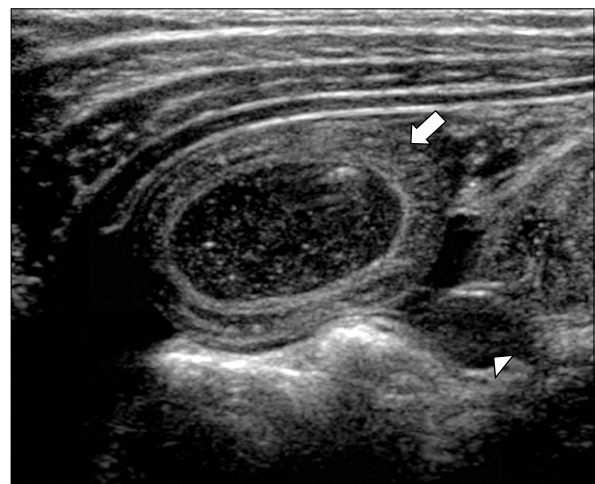


Fig. 2. Initial ultrasonography of the abdomen shows concentric wall thickening of the distended small bowel loop (arrow). Note the ascites (arrowhead).



Fig. 3. Contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis. (A) Coronal reformatted image shows large amount of ascites and extensive involvement of wall thickening and absent enhancement of the small bowel that indicate strangulation (arrows). Note the upward course of the terminal ileum (open arrow). (B) Axial CT scan demonstrated beaklike narrowing at the transition zone (arrowhead: serrated beak sign) and whirl of the obstructed bowel (curved arrow: whirl sign) which may suggest closed loop obstruction. Note the feculent matter mingled with gas bubbles within the lumen of dilated small bowel segment (asterisk: small bowel feces sign).



Fig. 4. Intraoperative photograph revealed herniating gangrenous bowel through small bowel mesenteric defect in operation field.

poor or absent enhancement of the strangulated bowel segment. The serrated beak sign and whirl sign of the obstructive bowel loop were identified at the transition zone which might indicate closed loop obstruction (Fig. 3). Surgical exploration was performed immediately. On the operation field, three quarters of the small bowel was herniated through the two fingerbreadth defect of mesentery located on near the ileocecal valve (Fig. 4). All herniated small bowel showed diffusely hemorrhagic, edematous and inflammatory change. Under the diagnosis of

congenital IH with strangulated small bowel, resection of more than 2 meters of gangrenous small bowel was followed by the anastomosis of proximal jejunum and terminal ileum.

After four-day treatment at pediatric intensive care unit, he was improved enough to move to general ward and started soft diet on the 5th day after operation. He did not fully recover yet due to hypertension after renal ischemic injury and short bowel syndrome, fortunately, his cognitive function is normal and he is gradually improving conditions.

DISCUSSION

IHs are rare cause of acute intestinal obstruction caused by hernias, accounting for only a small percentage of all instances of intestinal obstruction [2]. The autopsy incidence of IH is 0.2-2%, most of them were asymptomatic [4]. In the present case, the patient had no history of chronic abdominal pain or previous abdominal surgery, and symptoms of IH are nonspecific such as diffuse abdominal pain or vomiting.

In children with diagnosed IH, 85% of the neonates and infants had congenital transmesenteric

hernias and 82% of the reported congenital paraduodenal hernias presented in older children [3]. However, the other review of the literature showed that transmesenteric hernia was the most common type in older children as well as in neonates [3,5]. In adult, most frequent IH is paraduodenal, resulting from incomplete closure of surgically created mesenteric defects, and usually acquired resulting from previous abdominal surgery especially Roux-en-Y anastomosis [2,5]. Upper gastrointestinal (UGI) with small bowel follow-through contrast examination has a good detection rate for paraduodenal hernia, and can be diagnosed when a cluster of bowel loops is associated with loss of the usual interdigitation between the loops [3]. Transmesenteric hernias can occasionally be seen as a cluster of bowel loops on GI studies, but the detection rate was low on previous study [3]. Butterworth et al. [5] report the case of a 26-year-old female with spontaneous transmesenteric hernia of jejunum and proximal ileum due to a congenital mesenteric defect resulting in bowel gangrene, presenting initially with no hemodynamic abnormalities became to septic condition. Kakimoto et al. [6] described the case of 2.5-year-old girl with transmesenteric hernia resulting in death, which went undiagnosed during a recent hospital visit. The preoperative diagnosis of transmesenteric hernia is much more challenging than paraduodenal, but the risk of volvulus and bowel ischemia within a transmesenteric hernia is much more critical [3]. While it remains controversial, an UGI is contraindicated with and acutely presenting obstruction because it can cause a partial obstruction to become complete or may further complicate a total obstruction and took more time than CT scan. The ultrasound are limited in identifying an IH, but they are useful in assessment of bowel obstruction and other pathology that can cause vomiting, abdominal pain and distension. The great part of transmesenteric hernia cases, they are arriving in hospital with fatal condition, and that is the reason why CT should be performed urgently. CT findings in closed-loop obstruction, such as IH or volvulus, depend on the length, degree of distension, and ori-

entation of the closed loop in the abdomen [7]. Main difference of volvulus is result from embryologic failure of fixation and rotation of the gut [8]. Nowadays, the benefits of CT have led to an important role in suspected small bowel obstruction, both in the initial diagnosis and in making a treatment plan. CT scan has a high diagnostic accuracy not only become aware of obstruction sign but also in estimating its severity and range. The present case has shown that delayed treatment could be life threatening condition in transmesenteric IH. The principles of treatment in IH with sign of bowel obstruction include volume resuscitation, correction of electrolyte imbalances, transfusion, and last but not least, surgery should be necessary within 24 hours [9].

We report a case of congenital transmesenteric IH of jejunum and ileum with associated gangrene of bowel caused by a congenital mesenteric defect. To prevent the delay in diagnosis of IH, we should early use of the CT scan and consider urgent operation.

REFERENCES

1. Cazejust J, Lafont C, Raynal M, Azizi L, Tourabi AC, Menu Y. Internal hernia through the omental foramen. Answer to the e-quiz "Epigastric pain with sudden onset". *Diagn Interv Imaging* 2013;94:663-6.
2. Jain SK, Kaza RC, Garg PK. Incidental congenital transmesenteric hernia in an adult. *Eur Rev Med Pharmacol Sci* 2011;15:461-2.
3. Tang V, Daneman A, Navarro OM, Miller SF, Gerstle JT. Internal hernias in children: spectrum of clinical and imaging findings. *Pediatr Radiol* 2011;41:1559-68.
4. Zissin R, Hertz M, Gayer G, Paran H, Osadchy A. Congenital internal hernia as a cause of small bowel obstruction: CT findings in 11 adult patients. *Br J Radiol* 2005;78:796-802.
5. 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.
6. Kakimoto Y, Abiru H, Kotani H, Ozeki M, Tsuruyama T, Tamaki K. Transmesenteric hernia due to double-loop formation in the small intestine: a fatal case involving a toddler. *Forensic Sci Int* 2012;214:e39-42.
7. Boudiaf M, Soyer P, Terem C, Pelage JP, Maissiat E, Rymer R. CT evaluation of small bowel obstruction.

- Radiographics 2001;21:613-24.
8. Kim JS, Chung JY, Park DC, Kim SW, Kim HJ, Kim YH. A case of intestinal malrotation with midgut volvulus presenting with intermittent vomiting and abdominal pain. Korean J Pediatr Gastroenterol Nutr 2002;5:79-82.
 9. Bass KN, Jones B, Bulkley GB. Current management of small-bowel obstruction. Adv Surg 1997;31:1-34.