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Case Report

Fatal Small Bowel Occlusion on a Congenital Transmesenteric Internal Hernia: Report of an Autopsic Case

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Abstract

Transmesenteric internal hernia is a clinically difficult diagnosis due to intermittent and non-specific symptoms, and scant findings on clinical examination. In the case of strangulation or incarceration, the miss-diagnosis of the condition can lead to death. We report a fatal case of trans-mesenteric internal hernia, complicated with occlusion, and discovered only at autopsy. The 16-year-old patient was misdiagnosed with simple abdominal pain by two different doctors. True diagnosis was made only at autopsy. Transmenteric internal hernia can be easily missed and needs to be considered when diagnosing patients with abdominal pain associated or not with an occlusive syndrome, mainly in the young population.

Keywords: Autopsy; Abdominal pain; Internal hernia; Transmesenteric

Introduction

Internal hernias are defined by the protrusion of one or more viscera through a normal or abnormal opening in the peritoneum or in the mesentery, or through a depression of the abdominal cavity that [1-3]. They can be congenital or well acquired after trauma or surgery of the abdomen, and may cause strangulation or incarceration. This report describes a rare case of transmesenteric internal hernia complicated with small bowel occlusion, discovered only at autopsy, occurring in a 16-year old adolescent without any past medical or surgical history.

Case Report

A 16-year-old child without notable pathological or surgical history presented an acute abdominal pain. The symptomatology began ten days before his death, made of peri-umbilical pain associated with vomiting, without stopping of materials and gases. His parents reported the regular occurrence of similar crises during the previous days, spontaneously resolved after a few hours. He presented initially to the Emergency Department where explorations were made. Blood counts, serum creatinine, and blood glucose levels were normal. He was discharged to home under symptomatic treatment. In the following days, there were no signs of improvement. The patient consulted the family practitioner. The clinical examination and the abdominal ultrasound performed were considered normal and the patient was sent home again under antispasmodic treatment.

One day before his death, the patient continued to deteriorate clinically and presented hematemesis associated with excruciating abdominal pain. He died during his transportation to the hospital despite resuscitation measures. At the autopsy, abdominal examination revealed an abundant bloody peritoneal fluid in the abdominal cavity, with the incarceration of ileal loops through a defect in the mesentery measuring 3cm in diameter (Figures 1-3). The incarcerated ileum was necrotic over a length of 120 cm (Figure 4). Death was attributed to the strangulation of a transmesenteric hernia, considered as congenital regarding the lack of surgical history or traumatic context.

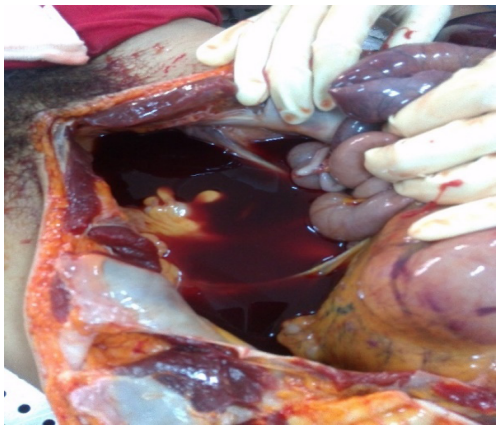
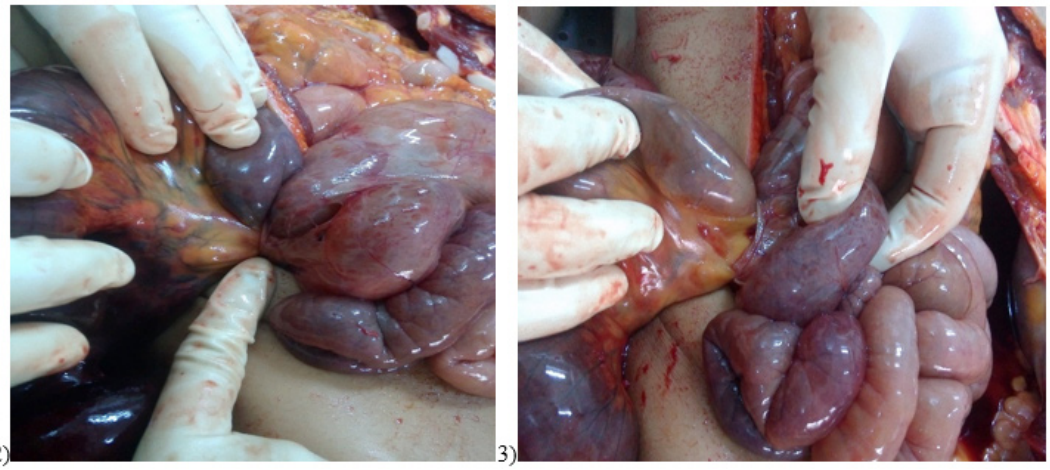


Figure 1: Peritoneal bloody fluid.



Figures 2 and 3: Trans mesenteric internal hernia.

Figure 4: Necrotic intestinal loops.

Discussion

Internal hernia is a rare cause of acute intestinal obstruction accounting for only 0.5% to 4.1% of the etiologies of acute intestinal obstruction [4]. In the broad category of internal hernias, there are several main types based on their location, among them we find the trans- mesenteric hernia. Trans-mesenteric hernia is defined as a protrusion of viscera through a defect in the mesentery of the small bowel, transverse colon, or sigmoid colon [5]. As no hernia sac is involved, a considerable length of bowel can protrude through the defect [6]. The mesenteric gap, normally between 2 cm and 3 cm in size, is generally the result of a surgery, but it can also be congenital as for our case. Other etiologies such as traumatic or inflammatory are much rarer [6,7]. Trans-mesenteric hernia is an uncommon variant of internal abdominal hernia [8].

It represents only 5 to 10% of internal hernias [6]. 30% of the cases remain without symptoms for lifetime, and the first case of transmesenteric hernia reported in 1836 was found at autopsy as was the case of this report. In fact, transmesenteric hernia is a rare cause of small bowel obstruction and is seldom diagnosed preoperatively, partly because of unfamiliarity with this type of internal hernia, and the lack of specificity of clinical symptoms [6,9,10]. In almost all cases, presentation is an acute intestinal obstruction or recurrent pain abdomen due to mesenteric ischemia without definite clinical symptoms or signs. In children, the most common presenting symptom is a sudden onset of abdominal pain located frequently in the epigastrium and the peri-umbilical area. As loops of bowel pass in and out through the defect, intermittent obstructive symptoms of abdominal pain, distension, nausea, vomiting, and constipation occur [6]. Moreover, the complications have often been described in children, about 35% of cases occuring during childhood [11].

In general, transmesocolic hernias are more likely than other subtypes to complicate, by developing volvulus and strangulation, or ischemia. The reported incidence of which is as high as 30 and 40%, respectively, with mortality rates of 50% for treated groups and 100% for non- treated subgroups [12]. Park CY reported a mortality rate associated with incarceration of the bowel of about 15%, but in the presence of gangrene of the bowel, the mortality rate is more than 50% [10]. These high morbi-mortality rates can be explained by the peculiarities of congenital transmesenteric internal hernias, which are the absence of a limiting hernial sac, large herniating bowel loops, volvulus of herniated bowel and bowel gangrene on both sides of the mesenteric defect [5,7]. Despite its severity, the clinical symptoms of internal hernia may be intermittent and nonspecific, making the diagnosis extremely difficult and challenging.

A high degree of clinical suspicion is needed, and trans-mesenteric hernia should be suspected in all patients presenting with recurrent pain abdomen, spontaneously resolving subocclusive syndrome, or small bowel occlusion without any history of previous abdominal surgery or trauma [13]. Because of the difficulty of clinical diagnosis, imaging studies might play a crucial role. Several diagnostic tools have been proposed, among which CT with injection is currently recommended as the most potent [14]. It usually allows early diagnosis, showing a magma intestinal loops in an abnormal position, possibly pushing the viscera of the neighbourhood. The hernial neck can also be visualized [1,14]. Only early intervention and surgical correction go a long way in preventing high morbidity and mortality. Delayed diagnosis leads to necrosis of the incarcerated loops and subsequent death of the patient.

Conclusion

Transmesenteric congenital internal hernias represent a rare cause of acute intestinal obstruction in adults. Diagnosis should be suspected in patients presenting with recurrent pain abdomen or small bowel occlusion without any history of previous abdominal surgery or trauma. Because of the lack of specificity of clinical symptoms, imaging mainly CT scan, finds its place in the diagnosis. As strangulation of the bowel is associated with high morbidity and mortality, early diagnosis and Exploratory Laparotomy should be the gold standard to prevent lethal complications in congenital transmesenteric intraabdominal hernias.

Conflicts of Interest

None.

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