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Congenital transmesenteric internal hernia; A rare cause of bowel ischemia in adults: A case report

Konjenital transmesenterik internal herni; Erişkinlerde nadir bir barsak iskemi nedeni: Olgu sunumu

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Abstract

Congenital transmesenteric hernias represent a very small group of internal hernias which are uncommon and are a rare cause of bowel obstruction and bowel ischemia in adults with few reported cases in published literature. Preoperative diagnosis of the condition is difficult, early intervention and surgical correction goes a long way in preventing high morbidity and mortality associated with cases of internal hernia. We present a case of true congenital transmesenteric hernia in a 31-year-old woman with no previous surgical history or trauma who presented with history of severe abdominal pain and the cessation of both feces and flatus. The abdominal CT scan could not confirm the diagnosis. The patient was operated upon on account of increasing abdominal pain and distention associated with shock. An exploratory laparotomy revealed a congenital transmesenteric defect through which loops of bowel had herniated and become gangrenous, resulting in resection and a stomy type Bouilly Volkmann.

Keywords: Congenital transmesenteric defect, Internal hernia, Bowel ischemia

Öz

Konjenital transmesenterik fıtıklar, nadir görülen ve yayınlanmış literatürde az sayıda bildirilmiş vaka ile erişkinlerde nadir görülen bir barsak tıkanıklığı ve barsak iskemi nedeni olan çok küçük bir iç fıtık grubunu temsil eder. Hastalığın preoperatif tanısı zordur, erken müdahale ve cerrahi düzeltme, içsel fıtık vakalarına bağlı yüksek morbidite ve mortaliteyi önlemede uzun bir yol kat eder. Daha önce cerrahi öyküsü veya travması olmayan 31 yaşında bir kadın hastada gerçek konjenital transmesenterik herni olgusunu sunuyoruz. Şiddetli karın ağrısı öyküsü ve hem dışkı hem de flatusun kesilmesi ile başvurdu. Abdominal BT taraması tanıyı doğrulayamadı. Hasta abdominal ağrının artması ve şokla ilişkili distansiyon nedeniyle ameliyat edildi. Bir keşif laparotomisi, barsak ilmeklerinin fıtıklandığı ve kangrenli hale gelen doğuştan bir transmesenterik defekti ortaya çıkardı, rezeksiyona ve bir stomi türü Bouilly Volkmann'a neden oldu.

Anahtar kelimeler: Konjenital transmesenterik defekt, İnternal fıtık, Bağırsak iskemi

Introduction

Internal hernia is a rare cause of intestinal obstruction in adults. Of internal hernia congenital transmesenteric hernia only constitute an estimated 5–10% of cases [1]. Congenital transmesenteric internal hernia is a very rare but definite acute surgical condition requiring early diagnosis. In almost all cases presentation is acute intestinal obstruction or recurrent pain abdomen due to mesenteric ischemia without definite clinical symptoms or signs. In published literature only 36 patients have suffered from bowel obstruction and 9 from ensuing ischemia secondary to transmesenteric hernia [2,3]. Almost all reported cases are diagnosed intraoperatively.

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Case presentation

A 31-year-old woman was admitted to general surgery emergency with a five-day history of constipation, progressive abdominal pain, nausea and vomiting. The last bowel movement had been two days ago. There was no significant past medical history especially of chronic constipation, psychiatric disease, trauma or abdominal surgery. There was no other relevant past personal or family history.

On examination, the vital signs were: Temperature 39˚C, Pulse 115/min, Respiratory rate 26/min, Blood pressure 90/60 mmHg. Abdominal examination revealed a distension of the abdomen without signs of peritonitis. The abdomen was tympanic to percussion. There were no umbilical or groin hernias. Digital rectal examination demonstrated an empty rectal vault without intraluminal masses. Further systemic examination was unremarkable. The abdominal X-ray revealed few gas distended bowel loops (Figure 1).

Blood investigations showed leukocytosis at 26.000 e/dl, CRP at 148 mg/l, serum sodium and potassium levels were within normal limits. Functional renal failure: serum urea 0.6 g/l, blood creatinine at 12mg/l.

The abdominal computed tomography: showed crowded and stretched mesenteric vessels, dilated and clustered small bowel, thickened intestinal wall (Figure 2).

4 hours after admission, the patient worsened her abdominal pain and her hemodynamic became unstable. After initial resuscitation with intravenous fluids, analgesics and antibiotics, decision was taken to proceed for an emergency laparotomy.

Intra operative findings approximately 1 m 50 cm of gangrenous small bowel, proximal ileum herniating (Figure 3) through a congenital small bowel mesenteric defect (Figure 4). The hernia was reduced, the mesenteric defect was repaired, small bowel resected and stomy type Bouilly Volkmann performed.

The postoperative course was uneventful. Discharge from hospital was five days following admission. Histopathology revealed hemorrhagic infarction of 145 cm of small bowel and mesenteric vessels showed no evidence of vasculitis or thrombosis. The restoration of intestinal continuity was done two months later with good evolution.

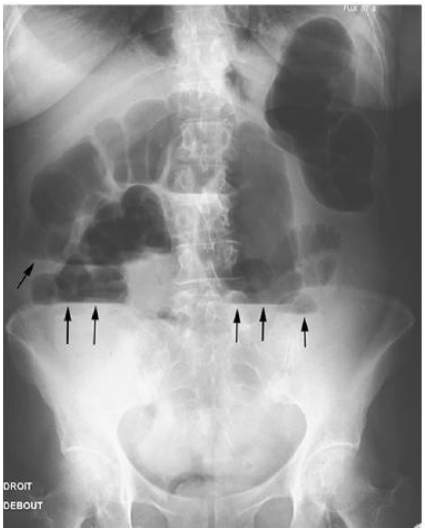

Figure 1: Dilated digestive loops, many seats hydroaeric levels

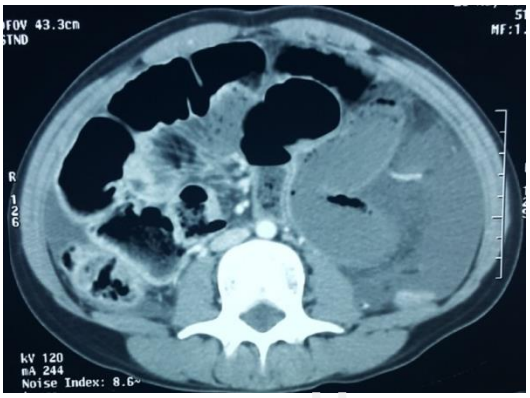

Figure 2: CT scan showed crowded and stretched mesenteric vessels, dilated and clustered small bowel, thickened intestinal wall

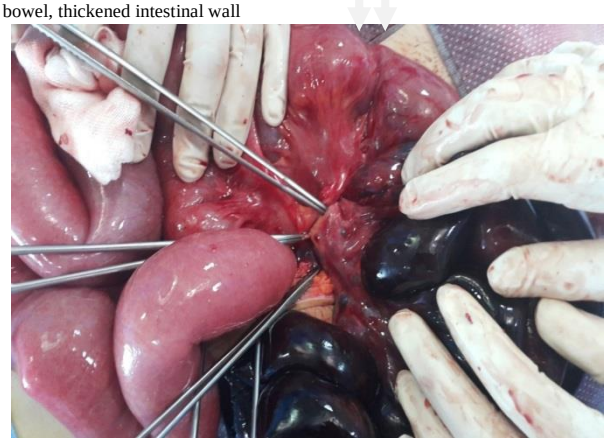

Figure 3: Intraoperative view demonstrating gangrenous small bowel, proximal ileum herniating through a congenital small bowel mesenteric defect

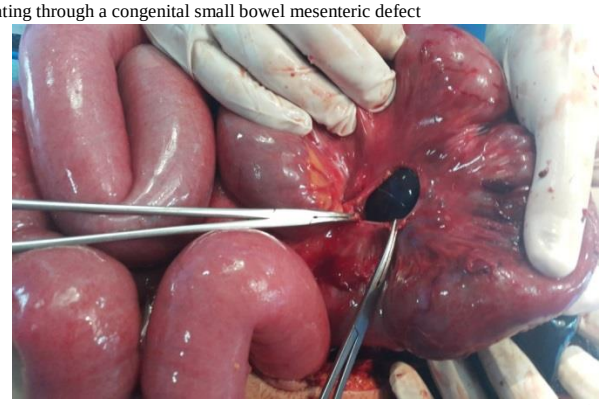

Figure 4: Intraoperative view demonstrating a congenital small bowel mesenteric defect

Discussion

An internal hernia is a protrusion of viscera through a defect or aperture, either mesenteric or peritoneal, and may be either congenital or acquired. Most internal hernia are paraduodenal (53%) and are acquired postoperatively, resulting from incomplete closure of surgically created mesenteric defects [4].

Transmesenteric hernia is a protrusion of viscera through a defect in mesentery of small bowel, transverse colon or sigmoid colon. Congenital internal hernia although rare, they can present at any age, though they are more common in the pediatric population [2]. Is an important cause of small bowel obstruction and its diagnosis is a challenge to both surgeons and radiologists. Congenital internal hernia is usually published as case reports rather than case series.

In 1836 Rokitansky reported first instance of trans-mesenteric internal hernia in an autopsy. Incidence of mesenteric defects is about 0.5% in all autopsies.

The pathogenesis of mesenteric defects is uncertain with one popular hypothesis suggesting the cause may be prenatal

intestinal ischemia and subsequent thinning of the mesenteric leaves, because prenatal intestinal ischemia is associated with bowel atresia in 5.5% of the pediatric population [5]. Alternatively a genetic etiology has been suggested given the association between transmesenteric hernia and other anomalies including cystic fibrosis and Hirschprung's disease [4].

Three main types of transmesenteric internal hernias are seen. The first and most common is the transmesocolic, which has been documented to occur in 0.7–3.25% of patients after laparoscopic Roux-en-Y gastric bypass surgery [6]. The second type of transmesenteric internal hernia occurs when bowel prolapses through a defect in the small-bowel mesentery, it is the same case in our patient. Finally, the third type, known as the Peterson type, has also been described and involves the herniation of small bowel behind the Roux loop before the small bowel eventually passes through the defect in the transverse mesocolon [7].

Transmesenteric hernias are more likely than other subtypes to develop volvulus and strangulation or ischemia, the incidence of which is reported to be as high as 30% and 40%, respectively, with mortality rates of 50% for the treated groups and 100% for the nontreated subgroups [8,9]. Volvulus and strangulation or ischemia may be partly caused by the usual small aperture of the defect (2–5 cm) in addition to the lack of encapsulation of the herniated loops, allowing a large length of small bowel to herniate through the mesenteric defect [8,9]. That's what happened in our case.

Transmesenteric hernias presentation can vary from asymptomatic to simple intermittent episodes of abdominal pain, nausea, diarrheas to acute abdomen with vomiting and even unexpected death [10,11].

X-rays and CT imaging may help in identifying signs of obstruction, volvulus and/or strangulation in patients with internal hernias [11]. Certain CT features represented by clustering of dilated small bowel peripherally without overlying omentum and leading to colonic displacement centrally may be suggestive of transmesenteric hernias [11]. However, final diagnosis will only be confidently achieved intraoperatively [12,13]. In our case, CT findings were suggestive of bowel obstruction with midgut volvulus to be the likely cause.

In literature, majority of transmesenteric hernias present as emergency cases, and surgical intervention with or without bowel resection and mesenteric defect closure is often required [10,12,14]. An early intervention is recommended to prevent unnecessary bowel resections and even death [10-12]. Our patient unfortunately was operated on in the gangrene phase probably due to a delay of management.

Conclusion

Preoperative diagnosis of transmesenteric hernia is difficult due to a lack of specific radiological or laboratory findings to confirm a surgeon's clinical suspicion.

We report a rare case of a 31-year-old woman with a spontaneous transmesenteric hernia of proximal ileum with associated gangrene of bowel caused by a congenital mesenteric defect. The insidious onset of this surgical emergency reaffirms the importance of surgeons maintaining a high index of suspicion for a transmesenteric hernia in patients with non-specific clinical and radiological signs.

Misdiagnosis and subsequent delayed exploration may lead to bowel ischemia and subsequent mortality, prognosis being directly correlated with the delay in diagnosis and treatment.

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