Acute Small Bowel Obstruction and Small Bowel Perforation as a Clinical Debut of Intestinal Endometriosis: A Report of Four Cases and Review of the Literature

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Abstract

Endometriosis is a quite common pathology, however, intestinal endometriosis is a rare condition, which typically occurs with chronic symptoms. Its acute presentation is very infrequent. We herein report four cases of intestinal endometriosis, in which the clinical debut occurred acutely: two as an acute small bowel obstruction and two as a small bowel perforation. None of the cases had a preoperative diagnosis of endometriosis. The interest of these cases lies in this exceptional form of presentation, such as a surgical acute abdomen. Therefore, intestinal endometriosis should be taken into account in the differential diagnosis of an acute obstructive or perforative process of the small or large bowel.

Key words: intestinal endometriosis, small bowel perforation, small bowel obstruction, acute abdomen


Introduction

Endometriosis is a quite common pathology. It is estimated to affect 4-17% of menstruating women (1). It typically presents as foci of endometrial glands and stroma outside its anatomical sight in pelvic organs, such as the ovaries, peritoneal cavity of the pelvis and rectovaginal septum. However, intestinal endometriosis is a rare condition, affecting between 3-37% of all cases of endometriosis (2-4). The most common location of intestinal endometriosis is the rectosigmoid area (50-90%), followed by rectovaginal septum (13%), small bowel (7%), appendix (3-18 %), caecum (4%) and Meckel’s diverticulum (2 cases described in the literature) (2-6). In the small bowel, the terminal ileum has the propensity to host the lesion, because it is in the vicinity of the fallopian tubes and ovary (4).

When the small bowel is affected, it is generally asymptomatic. If symptoms are present, then they are a result of a chronic obstruction in a majority of cases caused by strictures and adhesions. Additionally, this disease presents a challenge for clinicians because its symptoms can be easily mistaken for Crohn’s disease or tumors (7). Moreover, it usually affects the deeper layers of the intestinal wall, and colonoscopy and biopsy features can be absolutely normal or nonspecific.

We herein report four cases of small bowel endometriosis, diagnosed using surgical specimens, during a seven-year period (between June 2005 and October 2012) in our hospital. None of these cases had a preoperative diagnosis of endometriosis. We also review the literature on this subject.

Case Reports

Case 1

A 39-year-old woman was admitted at our hospital due to a 10-month history of abdominal pain, diarrhea, nausea, vomiting and weight loss of 10 kg. The patient refused cyclical or periodic intestinal bleeding. These symptoms had worsened in the last 3 months, and the initial clinic suspicion was an inflammatory bowel disease or, less likely, an
intestinal tumor. During her stay in the hospital, the patient developed symptoms and physical examination findings compatible with an acute abdomen. An abdominal CT scan was performed, which showed signs of a distal ileal obstruction with marked dilatation of handles and a preterminal ileum obstruction point at the pelvic level (Fig. 1A, 2). At this time, the clinic suspicion was an acute complication of an intestinal tumor or an inflammatory bowel disease in the form of an acute intestinal obstruction. There were no hernias upon abdominal palpation, the patient had no previous surgical history and the CT image was not compatible with an intestinal volvulus or invagination. There was no clinical suspicion of ingestion of a foreign body or a complication of a biliary calculus. Therefore, the patient underwent emergency surgery with resection of a 17 cm portion of the terminal ileum, which revealed a 1 cm long stricture area and 6 cm of the mucosa affected by ulcers (Fig. 1B). The microscope examination showed endometrial glands and stroma.

Case 2

A 39-year-old woman suffered from constipation, nausea, vomiting and abdominal pain for 4 days. The physical examination revealed pain and a mass in the right iliac fossa, and noise metal air speedboats. There were no palpable hernias or a clinical suspicion of appendicitis or involvement of gynecological organs. A plain X-ray of the abdomen showed marked dilatation of the pre-colonic intestinal area. She underwent urgent surgery due to the suspicion of a malignant mass in the abdomen causing subacute intestinal subocclusion. The intraoperative findings were a segment of the terminal ileum with wall thickening, which caused a bowel obstruction and had a whitish area located in the intestinal wall without involvement of the mucosa. The histological finding was a focus of endometrial glands and stroma (Fig. 3).

Case 3

A 77-year-old woman was admitted to the hospital due to symptoms of constipation, vomiting, abdominal pain and a
fever over the previous couple of days. She had no history of gynecological surgeries or hormonal treatment. The physical examination showed a grassed area in the hypogastrium with slight periumbilical fluctuation. CT of the abdomen showed a small bowel perforation in the ileum complicated with an intrauterine abscess, and findings suggesting fistulas connecting the uterus with the intestine (Fig. 4). The initial clinical and image suspicion was an intestinal perforation secondary to an intestinal tumor or an inflammatory bowel disease due to aging. Other possibilities included a complication of infectious ileitis or acute appendicitis. There were no palpable hernias. The CT image did not suggest a primary process of the uterine cavity. The uterine involvement appeared to be secondary to the intestinal damage. During the surgery, we observed a fistula connecting the uterine cavity with the terminal ileum and the cecum. The presence of a fistula connecting the uterus with the bowel could be explained from an initial focus of intestinal endometriosis, which results in a spontaneous perforation and causes the appearance of a communication between the intestinal region and uterine cavity (a fistula), which could also explain the development of an abscess inside the uterus. The surgical specimen was a 2 cm long bowel segment. The pathological study confirmed the presence of endometrial glands in the subserosa near the inflammatory fistula (Fig. 5).

**Case 4**

A 39-year-old woman went to the emergency room due to a 48-hour history of abdominal hypogastric pain and a low-grade fever not perceived by the patient. She entered a diagnostic study, with the clinical suspicion of an inflammatory bowel disease, inflammatory pelvic disease or complication due to appendicitis or cystitis. During admission, the patient presented with clinical deterioration with recurrence of abdominal pain and the low-grade fever. An abdominal CT scan showed multiple pelvic and iliac collections of uncertain origin, some of which appeared to be complicated with ectopic gas (Fig. 6). Ileocolic resection was performed. The specimen was composed of a 4 cm long segment of the small intestine and 19 cm long segment of the large intestine with a perforated area in the small bowel. There was a white indurated area within the bowel wall inside the most proximal segment of the colon. Endometriosis was found in both the small and large bowels and in the appendicular wall. All of the foci of endometriosis were found in the mucosa and the muscularis layer.

**Discussion**

The symptoms of gastrointestinal endometriosis are non-specific, with a broad spectrum of differential diagnoses. The most common presentation is cramp lower abdominal pain (76.5%), with a cyclic pattern only in 41.2% of all cases. Other symptoms include constipation or diarrhea (25-40%), a palpable mass (41.2%), melena, rectal bleeding, tenesmus, abdominal distension and meteorism (2, 5, 8). Much less frequently, gastrointestinal endometriosis can also debut as an acute abdomen, namely, perforation, intussus-
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Figure 6. abdomino-pelvic CT showing an area of thickening of the intestinal wall (*), which has punctured, and observing multiple extra-luminal air collections (marked with arrows).

ceptions and obstruction; however, these forms of debut are exceptional. An intestinal obstruction accounts for 7-23% of gastrointestinal endometriosis (9), and the majority of cases involve the rectosigmoid area. Perforation of the bowel wall is a rare outcome of endometriosis. A literature search revealed perforations in 7 cases of jejunum, 3 of ileum, 9 of large bowel and 3 of appendix endometriosis (2, 10).

Our four cases, during a seven-year period, represent an unusual acute clinical presentation of an unusual disease: gastrointestinal endometriosis. In our report, two cases presented with an acute small bowel obstruction, and two with a small bowel perforation. None of the cases had a previous diagnosis of endometriosis.

Bowel obstruction is a result of strictures, which are caused by adhesions with nearby structures or fibrosis within the intestinal wall. If deep wall involvement is present, fibrosis may produce kinking of the mucosa, which later results in bleeding (4). Likewise, superficial implants of the serosa suffer cyclical hemorrhaging, giving rise to chronic inflammation that produces fibrosis and causes injury of the inner layers due to the inflammation process itself or to an accompanying edema (2, 5). Endometriosis has a predilection to obstruct the terminal ileum. We identify 12 case reports of acute small bowel obstruction in the literature (1, 3, 6, 7, 9, 11-14). Our report describes two additional cases.

The pathophysiology of bowel perforation secondary to intestinal endometriosis is not clear. However, the presence of foci of intestinal endometriosis may cause a weakness in the affected region of the intestinal wall, facilitating its drilling secondary to peristalsis or traction phenomena of the intestinal wall (15).

Endometriosis affects different layers of the intestine wall, most commonly the subserosa, muscular layer and submucosa. The mucosa is only affected in approximately 10% of previously described cases (2, 5, 6, 13), showing different features such as architectural distortion, chronic inflammatory infiltrate, ulcers, structures and fistulas, which can be mistaken for Crohn’s disease. In the literature search, we found out that a preoperative diagnosis of Crohn’s disease was made in 14 case reports (3, 6, 7, 10, 14, 16), and we similarly initially suspected Crohn’s disease in our first case. In some previously reported cases, there was a coexistence of both diseases (16); although the presence of a source of intestinal endometriosis presumably causes a weakness in the affected region of the intestinal wall, in other cases the ischemic and inflammatory events were secondary to endometriosis (2, 7, 14). This is why the exclusion of the coexistence of the two entities is difficult, if not impossible, in some cases.

The second most frequent preoperative diagnosis is of a tumoral mass that is caused by an endometriotic focus surrounded by fibrosis, as in our second patient. Fibrosis is a quite common finding associated with endometriosis. Among the previous reported cases in the literature, we found a fibrous mass infiltrating the small bowel and causing an extrinsic obstruction, as well as a fibrous mass in the rectum (11), a mass arising from the appendix (1, 14), a hard mass with no clear origin site (12) and an inflammatory plastron involving the uterus, terminal ileum and sigma (3). Potential clinical diagnoses include carcinoid, adenocarcinoma, lymphoma, sarcoma, gastrointestinal stromal tumor or tumors in the vicinity that can comprise the bowel lumen. The microscope examination can be confusing as well because the endometrial gland epithelium can have reactive features, giving a false impression of dysplasia.

With respect to case 3, if intestinal endometriosis is a rare entity, it is much more uncommon during postmenopausal age. The focus of intestinal endometriosis may develop in premenopausal age pathophysiological mechanisms such as the retrograde menstrual flow or vascular dissemination. Another possible mechanism is spontaneous metaplastic changes in mesothelial cells of the intestine, leading to the development of a focus of endometriosis, which can occur both in postmenopausal or premenopausal age. Once the disease is established, its persistence and development requires stimulation by estrogens. In case 3, who was not receiving hormone treatment or had any source of endogenous
estrogen production, the source of estrogen production may have been the endometriosis focus due to the local production of estradiol, with autocrine and paracrine effects, allowing the progression of the disease or the peripheral conversion of androgens into estrogens in the adipose tissue (13, 17, 18). Posterior intestinal perforation may have occurred due to weakness in the intestinal wall caused by endometriosis, as explained above in this document.

Our goal is to stress the importance of taking gastrointestinal endometriosis into consideration in the differential diagnosis of gastrointestinal disorders in menstruating women. The present report also reveals how nonspecific and diverse the symptomatology of bowel endometriosis is and how it can mimic other pathologies, such as Crohn’s disease.

In conclusion, we reported four cases of intestinal endometriosis which presented as acute abdomen, two as an acute small bowel obstruction and two as an acute small bowel perforation. All of these cases required urgent surgical treatment and the final diagnosis was made through the histological studies. We believe that considering the diagnosis of intestinal endometriosis in women in fertile age who develop nonspecific gastrointestinal symptoms will help us to prevent urgent surgeries and offer our patients the best possible outcome.

The authors state that they have no Conflict of Interest (COI).

References


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