

Case Report

Appendiceal and Terminal Ileal Endometriosis Presenting as Acute Intestinal Obstruction: A Case Report

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Abstract: Endometriosis usually occurs in menstruating women up to 15%. Most common gastrointestinal involvement of endometriosis is found in the sigmoid colon, rectum and terminal ileum in 3%–37% of women. Terminal ileum is rarely involved in endometriosis. Patients with appendicular endometriosis constitute <1% cases of all pelvic endometriosis cases. Surgery is the choice of treatment for intestinal endometriosis in most cases.

Keywords: Endometriosis, Appendix, Ileum.

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INTRODUCTION

Endometriosis is characterized by the presence of functional endometrial tissue consisting of glands and stroma outside the uterus. Endometriosis usually occurs in menstruating women up to 15% [1]. Endometriosis can be divided into intra- and extra-peritoneal sites. In decreasing order of frequency, the intra-peritoneal locations are ovaries (30%), uterosacral and large ligaments (18%-24%), fallopian tubes (20%), pelvic peritoneum, pouch of Douglas, and gastrointestinal (GI) tract. Extra-peritoneal locations include cervix (0.5%), vagina and rectovaginal septum, round ligament and inguinal hernia sac (0.3%-0.6%), navel (1%), abdominal scars after gynaecological surgery (1.5%) and caesarian section (0.5%). Endometriosis rarely affects extra-abdominal organs such as the lungs, urinary system, skin and the central nervous system [2].

Most common gastrointestinal involvement of endometriosis is found in the sigmoid colon, rectum and terminal ileum in 3%–37% of women [1]. Proliferation and infiltration of the intestinal wall with endometrial implants may cause fibrotic reaction with formation of strictures and adhesions, probably from the effect of cyclical hormonal influences of menstruation. Eventually, this may lead to bowel obstruction and recurrent abdominal pain. Patients with appendicular endometriosis (AE) constitute <1% cases of all pelvic endometriosis cases. AE presents with pain in right iliac

fossa and symptomatically mimics appendicitis and definitive diagnosis is possible only after histopathological examination of excised appendix [3]. Endometriosis of the small bowel should be suspected in young, nulliparous patients with abdominal pain, in conjunction with signs of obstruction [4].

CASE REPORT

The patient is a 32 year old married woman who presented with complaints of abdominal pain, nausea and vomiting for 2 days duration. The pain was colicky, localized in the right iliac fossa. She had a history of mild occasional abdominal pain and amenorrhoea for past 3 months. X-ray and CECT abdomen was taken which showed multiple air-fluids levels, dilated segments of small bowel and absence of gas in the rectum. Intraoperative findings were stricture at I-C junction, dilated small bowel, collapsed large bowel, fibrotic strictures over distal ileum distorting distal ileum. Exploratory laparotomy and right hemicolectomy was done for the patient. The histopathology report was appendiceal and terminal ileal endometriosis. Immunohistochemistry was done which showed ER - positive and CDX2 - negative endometrial glands.

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Figure 1: Intraoperative specimen showing multiple strictures



Figure 2: Gross specimen

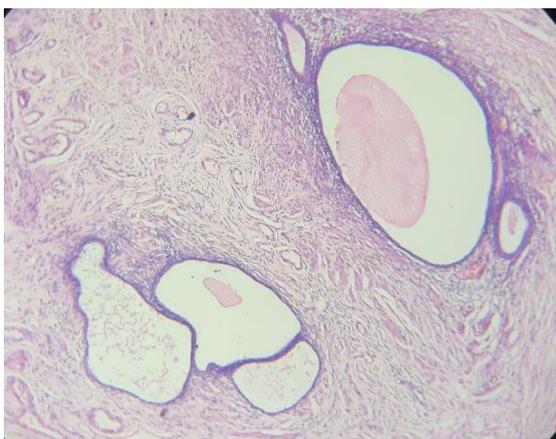


Figure 3: Ileal endometriosis



Figure 4: Appendiceal endometriosis



Figure 5: Endometrial glands showing ER positivity

DISCUSSION

The presence of functional endometrial tissue consisting of glands and stroma outside the uterus is called endometriosis. Sampson's retrograde menstruation theory is the most widely accepted theory of endometriosis. Endometrial tissue refluxes through the fallopian tubes, implanting on the serosal surface of abdominal and pelvic organs which commonly occurs during menstruation. However, other theories and factors, immunological, genetic and familial, could be involved in the pathogenesis of this disease. Endometriosis presents usually with pelvic pain, infertility and dyspareunia, but it may often be non-specific [1].

Gastrointestinal endometriosis (GE) accounts for 3–37% of all endometriosis cases, whereas appendiceal endometriosis (AE) is present in only ~3% of all GE cases and constitutes <1% of all the endometriosis cases. AE frequently involves tip and body of the appendix. The layers of appendix most commonly affected are muscular and seromuscular (~2/3rd cases), followed by the serosa (~1/3rd cases). Appendiceal endometriosis is classified into primary and secondary types. Lesion with histopathological evidence of endometriosis within the appendix with no clinico-pathological evidence of extra-appendicular endometriosis is described as primary form while secondary form is associated with features of external endometriosis. Many patients diagnosed with appendiceal endometriosis also suffer from menstrual irregularities and uterine leiomyomas. However, primary appendiceal endometriosis cases have also been reported without involvement of any other gynecological organ [3].

Based on clinical symptoms, appendiceal endometriosis may be sub classified into four groups:

1. Patient presenting clinically as acute appendicitis
2. Patient with appendix invagination
3. Patients presenting with non-specific symptoms like colicky abdominal pain, nausea/vomiting and malena

4. Cases who are asymptomatic [7].

Finding of an inverted or bulbous appendiceal orifice on colonoscopy suggests a more likely diagnosis of appendiceal endometriosis. Complications that can occur due to endometriosis of the appendix include hemorrhage, intussusception and bowel perforation [6]. Appendiceal wall affected by endometriosis show secondary changes such as chronic inflammation, fibrosis, hypertrophy/hyperplasia of muscularis propria [8].

The rectosigmoid is the most common site for intestinal endometriosis, accounting for 70% of all cases, while small bowel involvement, usually confined to the distal ileum, is less frequent (1%-7%). GI endometriosis patients present with relapsing bouts of abdominal pain, abdominal distention, tenesmus, constipation and diarrhoea. Rectal bleeding and pain during defecation may also occur. Endometriosis infiltrating the muscularis propria may lead to localized fibrosis in the bowel wall, strictures and small or large bowel obstruction [9].

The histologic hallmark of endometriosis includes presence of endometrial glands, stroma, fibrosis, and hemosiderin-laden macrophages. Layers involved by endometriosis of appendix wall include serosa and seromuscular. In the present case, the lesion was localized to tip of appendix and involved serosa and muscularis propria. Histology may not always show definite evidence of endometriosis. Endometriosis responds and shrink to hormonal therapy [10].

Endoscopic biopsies usually yield insufficient tissue for a definitive pathologic diagnosis as endometriosis involves the deep layers of the bowel wall. Endometriosis can induce mucosal changes without any specific pattern, which mimic findings of other diseases such as inflammatory bowel disease, ischemic colitis or neoplasm [12].

Radiologically, lesions of endometriosis are either of constricting and polypoid type or both. On barium studies, radiographic findings caused by implants in the ileum are similar to those in the colon. Rectosigmoid or cecal endometriosis on double contrast barium enema studies is seen as an extrinsic mass with spiculation and tethering of folds [13]. CT is not the primary imaging modality for evaluation of bowel endometriosis, although it can occasionally demonstrate a stenosing rectosigmoid mass. Multislice CT (MSCT) has a great potential for detecting alterations in the intestinal wall, especially if it is combined with enteroclysis [11].

Magnetic resonance imaging (MRI) has a high sensitivity (77%-93%) in the diagnosis of bowel endometriosis. The depth of rectal wall infiltration by endometriosis is poorly defined by MRI. A combination

of MRI and rectal endoscopic ultrasonography (EUS) has recently been proposed. When retroperitoneal infiltration is present, it is mandatory to know if the bowel wall is involved in order to identify patients requiring bowel resection. Both rectal EUS sensitivity and negative predictive value range from 92% to 100%. Surgery is the choice of treatment for intestinal endometriosis in most cases. For the accidental finding without symptoms of obstruction, hormone therapy with danazol or gonadotrophin-releasing hormone (GnRH) analogs may be considered. Surgical treatment should be indicated for women with pain, bleeding, changes in bowel habits and intestinal obstruction. In the small bowel, the treatment of endometriosis is surgical resection of the involved bowel, while medical therapy is only a temporary treatment [5, 14].

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