



Appendiceal endometriosis invading the sigmoid colon: a rare entity

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Abstract

Purpose We report an unusual case of endometriosis of the appendix with simultaneous invasion of the sigmoid colon.

Methods Clinical, radiological, surgical, and histological data of the patient were reviewed, as well as the current literature on gastrointestinal endometriosis.

Results A 41-year-old woman presented to the emergency department of our hospital with acute right lower quadrant pain, pronounced tenderness elevated white blood cell count, and increased C-reactive protein. Abdominal CT scan suggested a mucocele of the appendix. The patient was first treated with antibiotics, followed by en bloc resection of the appendix and of the sigmoid colon 2 months later. Histological examination revealed an endometriotic nodule of the appendix filling the appendiceal lumen and resulting in a mucocele which invaded the sigmoid colon wall.

Conclusions The diagnosis of gastrointestinal endometriosis can be challenging due to the variety of symptoms it can produce. Although extremely rare, a concomitant double gastrointestinal location of endometriosis may be possible and should be considered in women of reproductive age.

Keywords Endometriosis · Appendix · Sigmoid · Appendicitis

Introduction

Endometriosis is a common, benign, estrogen-dependent, chronic gynecological disorder characterized by the presence of uterine endometrial tissue outside its normal location. It affects 6 to 10% of women of reproductive age [1]. The presence of ectopic endometrial tissue and the resultant inflammatory reaction can cause devastating symptoms, including chronic pelvic pain, dysmenorrhea, dyspareunia, and infertility [2]. The pelvic cavity is the most common location for endometriotic implants, which

usually affect the retrocervical space, ovaries, vagina, rectosigmoid colon, bladder dome, and round ligaments [3]. Extrapelvic endometriosis is a rare entity, with the most common location being the gastrointestinal tract. The sigmoid colon is most commonly involved, followed by the rectum, ileum, appendix, and caecum [4].

Endometriosis of the appendix is a rare manifestation and is most often reported as an incidental finding [5]. Appendiceal endometriosis can imitate symptoms of acute or chronic appendicitis, and is also known to cause cyclic and chronic right lower quadrant pain, melena, lower intestinal hemorrhage, cecal intussusception, and intestinal perforation [6]. Endometriosis can also be a rare cause of mucocele, which is a distended mucous-filled appendix secondary to obstruction [5]. Even more rarely, endometriosis can have a simultaneous double gastrointestinal location manifested by atypical intestinal symptoms and signs.

We report herein an unusual case of a 41-year-old woman suffering from appendiceal endometriosis with concomitant sigmoid colon involvement, which was successfully treated by en bloc surgical resection of the involved segments.

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Case report

A 41-year-old woman, with no significant past medical history, presented to our hospital in March 2018 with sudden onset of abdominal pain in the right lower quadrant, accompanied by lower back pain and nausea. At physical examination, pronounced tenderness in the lower abdominal quadrants and a positive right psoas sign were found. Admission laboratory tests showed a white blood cell (WBC) count of 13,000/mm³ and a C-reactive protein (CRP) of 94 mg/l. Abdominal and pelvic CT scan revealed a short but enlarged appendix (27 mm × 19 mm) with an intraluminal mucocele-like mass and periappendiceal fat infiltration (Fig. 1). The patient was hospitalized and antibiotic therapy with amoxicillin/clavulanate was initiated for a total duration of 7 days. The patient was discharged at postoperative day 5, with a normal WBC count and a CRP at 33 mg/l. A colonoscopy performed 4 weeks later did not reveal any abnormal findings, except a millimeter-sized polyp that was resected. Histopathological examination of this small polyp revealed a hyperplastic polyp.

Surgery was performed in May 2018, after surgical meeting validation, anesthesiology evaluation, and patient's written informed consent. Initial planning consisted of simple laparoscopic appendectomy without rupture of the lesion in case of a normal appendiceal base, or, eventually, laparoscopic ileocecal resection in case of tumor invasion of the appendiceal base. Trocars were placed as for classic laparoscopic appendectomy (a 12-mm trocar at the umbilicus for the camera and two 5-mm trocars in suprapubic and left iliac fossa positions). The appendix found in the right iliac fossa showed no signs of inflammation but presented two round lesions at its distal end. Extreme care was taken throughout the procedure to avoid rupture of these lesions. Further exposition of the appendix revealed strong adhesion of the more distal appendiceal lesion to the terminal ileum (at 20 cm from the ileocecal valve) as well as to the sigmoid colon (Fig. 2). Careful dissection was initiated at the plane between the



Fig. 1 Preoperative contrast-enhanced abdominal CT scan showing an enlarged appendix with an intraluminal mass and periappendiceal fat infiltration (arrow)

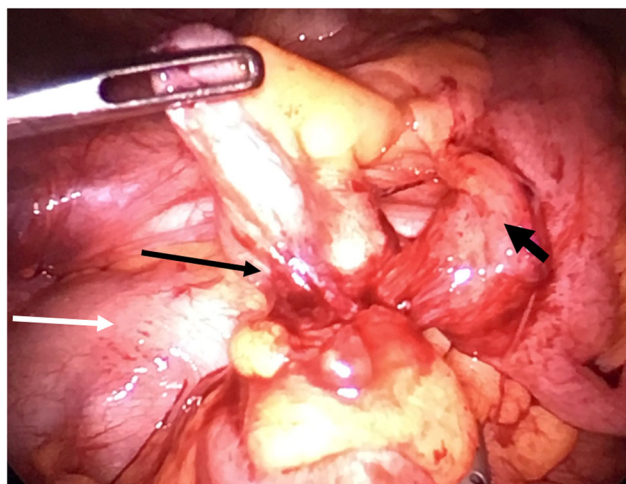


Fig. 2 Intraoperative view showing adhesion of the appendiceal mass (long black arrow) to the terminal ileum (short black arrow) and to the sigmoid colon (white arrow)

appendix and the terminal ileum, with successful dissociation of the two structures and without rupturing the appendiceal mass. Dissection was then carried out to the plane between the appendiceal mass and the sigmoid colon, with impossibility to dissociate the two structures due to tumor invasion of the sigmoid colon. In the context of mucocele suspicion, decision to convert to laparotomy was taken and a 10-cm lower midline incision was performed. En bloc appendectomy and segmental sigmoid colon resection were performed, with subsequent end-to-end anastomosis of the sigmoid colon. Nasogastric tube or abdominal drainage was not applied.

Operative time was 98 min and blood loss minimal (50 ml). Postoperative course was uneventful. Gastrointestinal transit resumed at postoperative day 2 and then oral intake was progressively introduced. The patient was discharged on postoperative day 4. Microscopic examination of the specimen revealed a voluminous endometriotic nodule of the appendix filling completely the appendiceal lumen and resulting in a mucocele, invading partially the sigmoid wall with absence of dysplasia or malignancy (Fig. 3). Colonoscopy performed 3 months after surgery was normal. The patient remains in good health 6 months after surgery.

Discussion

Endometriosis is defined as the presence of endometrial mucosa outside the uterine cavity. The most widely accepted theory is retrograde menstruation of endometrial tissue through the fallopian tube into the pelvis [7]. Moreover, in patients with endometriosis, immunological dysregulation may prevent normal clearance of ectopic endometrial cells facilitating their implantation [8]. Other theories implicated in endometriosis are the

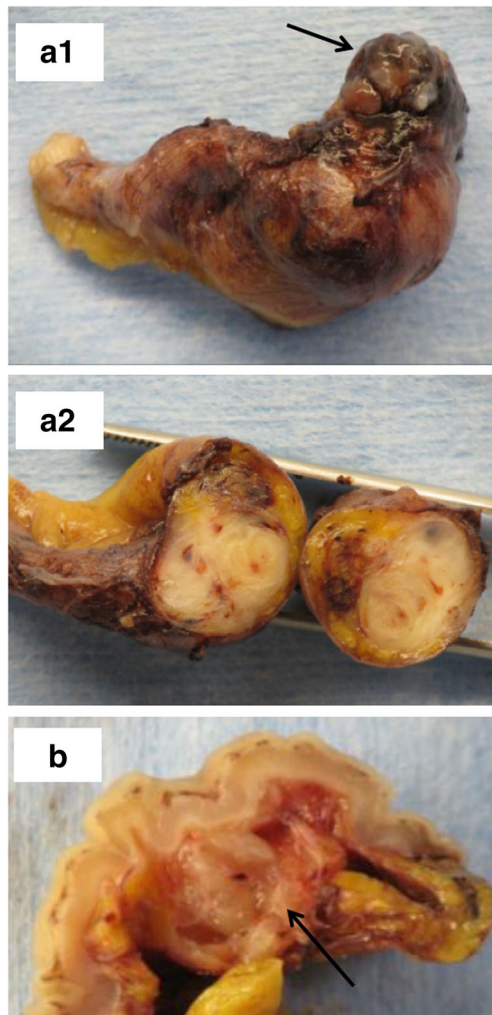


Fig. 3 Macroscopic examination of the resected surgical specimen: **a** Appendix specimen, 4.5 cm long, with (A1) pseudotumoral enlargement of the head and evidence of wall perforation with mucous leak (arrow); (A2) on cut section, obstruction of the appendiceal lumen by white fascicular endometriotic nodule of 1.3 cm with bluish microcysts. **b** Transversal section of sigmoid resection specimen (9.5 × 7 cm) showing the invaded part of the sigmoid colon wall (arrow)

coelomic metaplasia theory according to which normal peritoneal tissue mutates into ectopic endometrial tissue [9], bone marrow-derived stem/progenitor cells that differentiate in endometriotic tissue [10], or embryonic Mullerian rest according to which cell residual from Mullerian duct develop into endometriotic tissue under the estrogen stimulation at puberty [9].

Although endometriosis is a quite common pathology among women of reproductive age, gastrointestinal tract involvement occurs in only 12–15% of cases [11]. The most common presentation of gastrointestinal endometriosis is a cramp-like lower abdominal pain that follows the menstrual cycle. Other symptoms are constipation or diarrhea, melena, rectal bleeding, palpable mass, meteorism, and abdominal distention [12]. In some cases, as in the one reported here,

gastrointestinal endometriosis can present with sudden onset abdominal pain, mimicking other conditions as, for example, acute appendicitis or intestinal obstruction. This mimicking of multiple conditions in addition to the lack of specific radiologic findings for gastrointestinal endometriosis makes accurate preoperative diagnosis difficult [4].

In the hereby presented case, preoperative examinations never suspected the presence of gastrointestinal endometriosis. Colonoscopy showed no abnormalities, since, in contrast to the appendix, the endometrioma invasion of the sigmoid colon was not transmural. Abdominal and pelvic CT scan was in favor of a mucocele. Even if magnetic resonance imaging (MRI) is the most accurate imaging modality for gastrointestinal endometriosis detection, the gold standard remains surgical resection of the involved intestinal segment and histopathological examination [4]. In our case, appendiceal endometriosis invading the sigmoid colon was an incidental finding after histopathological analysis of the resected specimen.

Surgical resection of the involved intestinal segments affected by endometriosis is the treatment of choice in presence of symptoms, such as intestinal obstruction, frequent bleeding episodes, severe pain, or malignancy suspicion [4]. After surgery, symptomatic lesion recurrences are frequent (40–50% at 5-year follow-up), especially when no postoperative adjuvant treatment is applied [13]. In the present case, the therapeutic attitude used was laparotomy with en bloc resection of the appendix and the involved part of the sigmoid colon which were impossible to dissociate during surgery. Even if laparoscopic resection of the appendix was initially intended, the suspicion of a malignant appendiceal mucocele, with fear of rupture in the peritoneal cavity and development of a pseudomyxoma peritonei, led us to convert to laparotomy. Appendiceal mucocele is a rare finding, occurring in 0.3% of appendectomy specimens [14]. It is even more rarely due to endometriosis, with only few cases reported in the literature [5]. One devastating complication of malignant mucocele is pseudomyxoma peritonei, defined as mucinous ascites of the abdomen due to dissemination of mucinous neoplastic cells throughout the peritoneal cavity. In 94% of cases of pseudomyxoma peritonei, the source of these cells is a perforated appendiceal tumor [15]. Therefore, the decision to convert to laparotomy in the present case was judicious and should not be considered a failure.

Conclusion

Endometriosis of the gastrointestinal tract is a rare finding, but remains a possible diagnosis in the context of gastrointestinal symptoms in women of reproductive age. Diagnosis of this condition is challenging, as it can mimic different conditions and has few specific clinical and radiologic findings. Surgery is currently the treatment of choice for symptomatic intestinal

endometriosis. Although extremely unusual, simultaneous double gastrointestinal location is possible and must be considered. We reported here a very rare case of transmural appendiceal endometriosis invading the sigmoid wall, which was clinically expressed with acute abdominal symptoms and was successfully treated by en bloc resection of the involved intestinal segments.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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