

Endometriosis of the appendix

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Purpose: Endometriosis is defined as functioning endometrial tissue outside of the uterus. Endometriosis of the appendix is uncommon. Its clinical presentation varies from asymptomatic to acute abdominal pain. The aims of this study were to describe our experience of managing patients with appendiceal endometriosis and to review the clinical characteristics of this medical condition.

Methods: Five cases of appendiceal endometriosis diagnosed between January 2007 and December 2012 were retrospectively reviewed. Patients treated for intra-abdominal endometriosis were excluded.

Results: The mean age at diagnosis was 33.8 years old. One patient was in the second trimester of pregnancy. Two patients were asymptomatic. Three patients had clinical symptoms including two with acute pain in the right lower quadrant and one with abdominal discomfort. Four patients showed appendiceal abnormalities in imaging studies including two cases of suspected mucocoele and two cases of appendiceal infection. There were no suspicions of endometriosis of the appendix based on preoperative imaging studies. Three patients underwent appendectomy due to clinical symptoms, and two underwent incidental appendectomy combined with another operation. None of the patients received adjuvant therapy.

Conclusion: Appendiceal endometriosis should be included in the differential diagnosis for acute abdominal pain, especially when women of childbearing age present with clinical symptoms of acute appendicitis.

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Key Words: Appendix, Appendicitis, Endometriosis, Abdominal pain, Acute abdomen

INTRODUCTION

Endometriosis is defined as functioning endometrial tissue in a location outside of the uterus. It is a highly prevalent disease that affects up to 10% of menstruating women, approximately 70% of women with chronic pelvic pain, and up to 50% of women with infertility [1].

Clinical presentation of endometriosis varies, and some affected women may be asymptomatic. Symptomatic women have mild to severe pain that may or may not be cyclic depending on the site involved and severity of the disease. Endometriosis most often causes chronic pelvic pain; however, women may suffer acute abdominal or pelvic pain severe enough to cause them to seek emergency medical care [1].

Appendiceal endometriosis was first described in 1860 [2].

Endometriosis of the appendix is uncommon, and presentation as acute abdominal pain is rare. Endometriosis of the appendix may be asymptomatic or present as acute appendicitis, lower gastrointestinal bleeding, intestinal perforation, or intestinal obstruction from intussusception [3-5].

The aims of this study were to describe our experience of managing patients with appendiceal endometriosis and to review the clinical characteristics of this medical condition.

METHODS

Between January 2007 and December 2012 in the Department of Surgery, Gangnam Severance Hospital, Yonsei University College of Medicine, 2,173 patients underwent appendectomy. Of these, five patients (0.2%) were diagnosed with appendiceal

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endometriosis. Their medical records were retrospectively reviewed. Patients treated for intra-abdominal endometriosis were excluded.

Descriptive variables including patient demographics, preoperative imaging findings, symptoms and signs, and final pathologic reports were reviewed.

Approval to conduct a retrospective review of the images and medical records of patients was obtained by the Institutional Review Board of Gangnam Severance Hospital, Yonsei University College of Medicine, IRB No. 3-2012-0225. The Institutional Review Board approved this retrospective observational study and did not require patient approval or informed consent for the review of their records.

RESULTS

Patient demographics and clinical characteristics are listed in Table 1. The mean age at diagnosis was 33.8 years old with ages ranging from 26 to 60 years old. One patient had an intrauterine pregnancy of 18 weeks.

Two patients were asymptomatic. Three patients had clinical symptoms including two with acute right lower quadrant (RLQ) pain and one with abdominal discomfort. The two asymptomatic patients underwent incidental appendectomies, one during a laparoscopic right hemicolectomy and the other during a left salpingo-oophorectomy.

Preoperative imaging studies included abdomen-pelvic computed tomography in four patients and ultrasonography in one patient who was pregnant. Imaging studies showed appendiceal abnormalities in four patients. Two abnormalities were suspected to be mucocele, and two were thought to be due to an infected appendix. There were no suspicions of endometriosis of the appendix in the preoperative imaging studies.

Direct tenderness in the RLQ of the abdomen was noticed in three patients with clinical symptoms but leukocytosis was not specific. The leukocyte count for each of the three patients was 8,950/ μ L (65.4%), 9,150/ μ L (91.2%), and 11,480/ μ L (78.1%) (normal, 4,000–10,800/ μ L).

In one patient who received an incidental appendectomy

during a left salpingo-oophorectomy, gynecologic surgery was performed for pyosalpinx; the incidental appendectomy was performed because of appendiceal wall thickening and inflammation.

None of the patients received adjuvant therapies including hormonal therapy.

DISCUSSION

Endometriosis is the presence of endometrial glands and stroma outside of the uterine cavity and musculature. Several theories explain the pathogenesis of extrauterine endometriosis [6]. First is the implantation or retrograde menstruation theory that endometrial tissue from the uterus is transported in a retrograde fashion through the fallopian tubes [7]. Second are the direct transplantation and dissemination theories, which may explain extrapelvic endometriosis [8]. Third is the coelomic metaplasia theory that the peritoneal cavity contains progenitor cells capable of differentiating into endometrial tissue [9]. Fourth is the induction theory that sloughed endometrium produces substances that cause endometriosis. Fifth is the embryonic rest theory that a specific stimulus to a Müllerian origin cell nest produces endometriosis. Sixth, the most recently developed theory, is the cellular immunity theory, which suggests that alterations in cell-mediated and humoral immunity allow ectopic endometrial cells to proliferate [6].

Endometriosis is usually asymptomatic; symptoms are often related to the location of the lesions. For that reason, endometriosis of the gastrointestinal tract may cause a wide spectrum of symptoms. Endometriosis should be considered by surgeons for cases of acute abdominal pain manifesting with complex symptomatology.

Appendiceal endometriosis is a rare entity and was first described in 1860 [2]. Patients with appendiceal endometriosis can be categorized into four groups: (1) patients who present with acute appendicitis, (2) patients who present with appendiceal invagination, (3) patients manifesting atypical symptoms such as abdominal colic, nausea, and melena, and (4) patients who are asymptomatic [3]. Pain in the right lower abdominal quadrant is one of the most common symptoms of appendiceal

Table 1. Clinicopathologic characteristics of the patients

Gender/age	Symptom	Imaging modalities	WBC count (normal, 4,000–10,800/ μ L)	Appendectomy	Combined op.
F/60	Ileus	Mucocele	8,950/ μ L	Laparoscopic	-
F/27	None	Infected mucocele	17,420/ μ L	Laparoscopic	Right hemicolectomy
F/26	None	PID	9,430/ μ L	Laparoscopic	Left salpingo-oophorectomy
F/26	RLQ pain	Inflamed appendix	9,150/ μ L	Laparoscopic	-
F/30	RLQ pain	No specific findings	11,480/ μ L	Open	-

RLQ, right lower quadrant; PID, pelvic inflammatory disease.

endometriosis. One-third of patients with abdominal pain present with all of the symptoms of appendicitis. In the present series, two patients belonged to the first group; one was representative of the third group; two represented the fourth group.

Although laboratory results were not specific, leukocytosis along with a subfebrile temperature was usually present. While the overall profile of our second case cannot be related completely to appendicitis, a similar condition has been reported [3].

Hormonal changes during pregnancy are proposed to cause endometriotic foci to enlarge during the first two trimesters, placing pregnant patients at greater risk of perforation [10]. One of our patients was in the second trimester of pregnancy.

Abdomen-pelvic computed tomography (APCT) is usually the imaging modality of choice for evaluating acute RLQ pain. If endometrial involvement is isolated to the appendix, the imaging findings of endometriosis with secondary appendicitis are usually indistinguishable from acute appendicitis [11].

APCT findings that demonstrate acute appendicitis combined with additional findings suggesting abdominal or pelvic endometriosis may indicate the etiology. However, the CT findings for endometriosis are almost always nonspecific and are usually not evident in imaging studies [1]. Abdomen ultrasonography can be considered for patients who cannot receive APCT.

In the present series, although four patients showed appendiceal abnormalities on preoperative APCT and abdominal ultrasonography, there were no suspicions of endometriosis of the appendix in preoperative imaging studies.

Appendiceal endometriosis can be diagnosed histopathologically. The mucosa of the appendix is never affected. How-

ever, glandular tissue, endometrial stroma, and hemorrhagia are observed in the muscular and seromuscular layers in two-thirds of cases and solely in the serosa layer in one-third of cases [3].

The treatment consists primarily of surgery and hormone therapy and tends to be determined by the age of the patient and the severity of symptoms [3]. Thus, the extent of resection should be appropriate. Intraoperative investigations usually result in an accurate diagnosis of endometriosis with minimal resection. A gynecological assessment should be performed to determine the extent of endometriosis. Postoperative follow-up is mandatory for appendiceal endometriosis [12].

Laparoscopic appendectomy is now commonly performed for appendicitis. Laparoscopic surgery is useful for women with chronic abdominal pain caused by endometriosis, ovarian cysts, adhesions, and hernias. Laparoscopy enables exploration of the total peritoneal cavity and selection of the appropriate method for a definitive diagnosis. Medical treatments such as hormonal therapy for endometriosis are secondary. Hormonal therapy is sometimes effective for relieving chronic pain. Appendiceal endometriosis appears to be an incidental finding, and one that is not clinically important [13].

In conclusion, appendiceal endometriosis is rare, and its preoperative diagnosis is difficult. However, it should be included in the differential diagnosis of acute abdominal pain, especially when women of childbearing age present with clinical symptoms of acute appendicitis.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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