Appendiceal Endometriosis: A Rare Cause of Acute Appendicitis

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Abstract

Background: Acute appendicitis is the most common cause of acute abdomen. The etiology has not been defined clearly but potential causes are fecoliths, lymphoid hyperplasia and malignancies. However, an extremely uncommon cause of acute appendicitis is endometriosis which leads to dilemmas in the diagnostic process.

Case report: A 21-year-old Caucasian female patient presented in the Emergency department complaining of acute progressive abdominal pain in the right lower quadrant. The physical examination revealed signs consistent with acute appendicitis. The patient underwent diagnostic laparoscopy, which revealed early inflammation of the appendix and thus was subjected to appendicectomy. Histologic examination revealed endometriosis of the appendix.

Conclusion: The presented case emphasizes on how endometriosis can affect only the appendix without any involvement of the reproductive organs. A high index of clinical suspicion is required especially in women of reproductive age who present with periodic chronic pain associated with their menstrual cycle. Nonetheless, this can only be confirmed through diagnostic laparoscopy and histologic examination, which also provides the definite cure of the disease.

Keywords: appendicular endometriosis; acute abdomen; laparoscopy; inflammation

Introduction

Acute appendicitis is the most common cause of acute abdomen that requires emergency surgery [1, 2]. The etiology of this condition has not been completely understood yet, but it can mainly be attributed to either diffuse inflammation of the appendiceal wall due to bacterial invasion of its lymphoid tissue or obstruction of the appendiceal lumen by a fecolith, adhesive kink, swollen lymphoid tissue or mucus [1]. However, another more unusual cause of acute appendicitis is appendiceal endometriosis, which has a prevalence around 2.8% in women with endometriosis and 0.4% in the general population [3]. In this article we present a rare case of a histologically proven acute appendicitis in a patient with appendiceal endometriosis, who had no other abnormal findings suggestive of endometriosis in the laparoscopy.

Case Report

A 21-year-old Caucasian female patient presented in the Emergency Department complaining of acute onset right iliac fossa pain lasting for 24 hours. She denied any other symptoms but mentioned similar episodes of pain during the past few months. Her medical background was positive for hypothyroidism under treatment with oral levothyroxine sodium. On clinical examination the patient was apyrexial, hemodynamically stable and had a positive McBurney sign with rebound tenderness in the right iliac fossa. Her blood tests revealed a white blood cell value of 19.7*10^3/μL (normal range 4.9-10.8) with 83% neutrophils (normal range 40-75%) and a C-reactive protein value of 5.922 mg/dl (normal range 0.0-0.5). The patient was subjected to a computed tomography scan in order to confirm the suspected diagnosis of acute appendicitis and exclude any other intrabdominal pathology. However, the scan did not reveal any abnormal findings. Nonetheless, due to patient’s clinical condition and her blood results the decision to perform a diagnostic laparoscopy was made.

The procedure revealed an injected and inflamed appendix and, thus, an appendicectomy was performed. The uterus, ovaries and fallopian tubes were found macroscopically normal in appearance. The small bowel was also checked for Meckel’s diverticulum with a negative result. The specimen’s histology report revealed acute appendicitis with endometriosis foci in the appendix (Figures 1 and 2).
The patient recovered safely from the operation and was discharged on the first post-operative day.

**Discussion**

Endometriosis is a very common benign gynecologic disorder that affects up to 30% of women of reproductive age [4, 5]. Based on the localization of the ectopic endometrial tissue, endometriosis is divided into internal and external [4]. Internal endometriosis is defined as the presence of endometrial tissue in the muscle layer of the uterus, while external endometriosis is characterized by the presence of endometrial tissue outside the uterus [4]. Extra-uterine endometriosis is usually found in the genital organs and the pelvic peritoneum [4-6]. Nonetheless, ectopic endometrial tissue can be found in the gastrointestinal tract (3-37%), ureters, bladder, liver, diaphragm, lung, pleura, brain, cutaneous and scar tissue, pericardium, eye and other sites [4-6]. According to our review of the literature, there has been no report of endometrial tissue found in the spleen. Regarding gastrointestinal endometriosis, the most affected part is the rectosigmoid junction (72%), followed by the recto-vaginal septum (13%), small bowel (7%), cecum (4%), and appendix (3%) [4].

Appendiceal endometriosis was first described in 1860 by Von Rokitansky [4, 7, 8]. It is divided into primary and secondary. Primary appendiceal endometriosis is characterized by the presence of endometrial tissue in the appendix without clinicopathological evidence of any other endometriosal lesions anywhere in the body. Secondary appendiceal endometriosis is characterized by the presence of other foci of external or internal endometriosis [4]. Appendiceal endometriosis is usually seen in patients with concomitant ovarian endometriosis and is often associated with uterine leiomyomas and menstrual irregularities [4, 8, 9].

Patients with appendiceal endometriosis can be divided into four categories regarding clinical manifestation: 1) acute appendicitis, which
is the most common clinical presentation and usually appears during menstruation, 2) appendiceal invagination/intussusception, 3) atypical symptoms, such as abdominal colic, melena and nausea and 4) asymptomatic appendiceal endometriosis [4, 8, 9]. Appendiceal intussusception is a very rare condition, first reported by Mc Kidd in 1958, with an approximate incidence of 0.01% [4, 10]. Generally, it is associated with congenital abnormalities, mucocele, endometriosis and adenocarcinoma [10]. Appendiceal endometriosis leading to intussusception has been reported less than 30 times in literature during the past 50 years and in most cases, endometriosis was also present in other organs [11]. Clinical manifestation varies from asymptomatic or atypical, recurrent crampy lower abdominal pain to symptoms of bowel obstruction and acute appendicitis [4, 10, 12].

Differential diagnosis of appendiceal endometriosis includes inflammatory disorders, such as inflammatory bowel disease, chronic ischemic colitis, diverticular disease, infectious diseases such as bowel tuberculosis and benign or malignant neoplasms [4]. It is important to highlight that there is no pathognomonic imaging finding for endometriosis and most cases are confirmed during or after surgery [4, 12]. Definite establishment of the diagnosis is made by histopathology. Around half of the cases involve the tip and half involve the body of the appendix [4]. The mucosa of the appendix is never affected, while endometrial stroma, granular tissue and hemorrhagic lesions are identified in the muscular and seromuscular layers in two-thirds of cases and exclusively in the serosa layer in one-third of cases [7]. The treatment of endometriosis consists primarily of surgery and hormonal suppression therapy in cases of extraintestinal endometriosis [7, 13]. The extent of surgical resection is decided based on the patient’s age and the severity of the symptoms. Postoperatively, a gynecological assessment is necessary to determine the extent of endometriosis, while follow-up for appendiceal endometriosis is also mandatory [7].

Conclusion

Appendiceal endometriosis is an unusual diagnosis that is almost always established histopathologically. However, it should be included in the differential diagnosis of cases of female patients at childbearing age that present with clinical symptoms of acute appendicitis. Usually, these patients present with a history of menstrual disorders and periodic lower abdominal pain. Open or laparoscopic surgery provides definite diagnosis and treatment.

References


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