Diverticular disease and endometriosis of the vermiform appendix: A unique combination

Apendiks vermiformiste divertiküler hastalıık ve endometriozis: Özgün bir kombinasyon

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Abstract

Endometriosis is characterized by the presence of endometrial glands and stroma outside the uterine cavity. Appendiceal endometriosis, which is very rare, may have nonspecific presentation such as chronic pelvic pain, but sometimes it can masquerade clinically as acute appendicitis. Also appendiceal diverticulum may be congenital or acquired and is infrequently encountered lesion. They are often asymptomatic but when diverticulitis or perforation develops, it may clinically mimic acute abdomen of acute suppurative appendicitis. Herein we report a case with a unique combination of appendiceal diverticulosis and endometriosis in a 39-year-old woman that was diagnosed preoperatively as acute appendicitis.

Keywords: Appendix, endometriosis, diverticulum.

Öz

Endometriozis, endometriyal bez ve stromanın uterin kavite dışında görüldüğüyle karakterlidir. Oldukça nadir görülen apendisyel endometriozis, kronik pelvik ağrı gibi nonspesifik bulgularla seyredebileceği gibi bazen klinik olarak akut apandisiti de taklit edebilir. Apendisyel divertikül konjenital veya edinsel olabilir ve görüleme sıklığı nadirdir. Genellikle asemptomatiktir; ancak divertikülit veya perforasyon geliştiğinde, akut süpüratif apandisitin akut batın tablosunu taklit edebilir. Bu çalışmada preoperatif olarak akut apandisit tanısı konulan 39 yaşındaki kadın hastada apendisyel divertikülözis ve endometriozis birlikteilinin özgün olması nedeni ile olgumuz sunulmuştur.

Anahtar Sözcükler: Apendiks, endometriozis, divertikül.

Introduction

Endometriosis is characterized by the presence of endometrial glands and stroma outside the uterine cavity. It usually occurs in pelvis, involving the ovaries, uterine ligaments, rectovaginal septum and sometimes laparotomy scars, vagina and vulva (1). The bowel is affected in 3-37% of extrapelvic cases while the appendix is affected around 0.8-1% (1, 2). Appendiceal endometriosis more typically has nonspecific presentation such as chronic pelvic pain, but sometimes it can masquerade clinically as acute appendicitis.

Appendiceal diverticulum may be congenital or acquired and is infrequently encountered with reported incidence rates of 0.014% to 2.2% (3-5). They are often asymptomatic but when diverticulitis or perforation develops, it may clinically mimic acute abdomen of acute suppurative appendicitis. It is characterized by mucosal herniations through a muscular defect on the mesenteric border of the appendix. Acquired diverticula which are usually small with a diameter of 3 to 5 mm have no muscularis propria layer.
Herein we report a case with a unique combination of appendiceal diverticulosis and endometriosis in the same patient that was diagnosed preoperatively as acute appendicitis.

Case Report

A 39-year-old woman who has no significant medical history was admitted to the emergency service with complaint of nausea and lower quadrant dull pain for two days duration which was not related with her menstrual cycles. Abdominal examination showed tenderness in the right lower quadrant with no rigidity or any palpable mass. There was a slight increase in the white blood cell count. Abdominal ultrasound and computed tomography scan showed moderate appendiceal swelling favoring the diagnosis of acute appendicitis. As leukocytosis and increase in C-reactive protein developed in the patient laparoscopic appendectomy was performed with a clinical diagnosis of acute appendicitis after the informed consent form was taken from patient. Macroscopic examination revealed an appendix of 5 cm of length and 2 cm diameter. On serial cuts, there were second lumen formations measuring 2-3 mm each adjacent to the actual lumen and also multiple hemorrhagic areas in the appendiceal wall. No tumoral mass was noted. Histopathologic examination showed second lumens formed by the herniation of the mucosa through the muscularis propria with minimal inspissated intraluminal secretion and diverticulitis (Figure-1). There was no diverticular hemorrhage but severe acute peritonitis. Interestingly there were multiple foci of endometrial gland and stroma involving the appendiceal wall with many hemosiderin laden macrophages and acute serosal inflammation (Figure-1, inset a-b).

Figure-1. Actual lumen of the appendix and adjacent diverticulum, X20, inset a-b: endometrial glands and stroma within the appendiceal wall (a X40, b X100).

Discussion

Endometriosis is the presence of endometrial tissue outside its normal location. 5-15% of women in the reproductive period have this condition which is usually associated with dysmenorrhea, chronic pelvic pain and infertility (1, 6). It is called genital endometriosis when located in the uterine muscle, cervix, vagina, ovary and fallopian tubes, and called extra genital endometriosis if located outside the genital organs in almost every organ and tissue in the body except the spleen (1, 6). Endometriosis of the gastrointestinal tract involves rectosigmoid region 72%, rectovaginal septum 13%, small bowel 7%, caecum 4%, appendix 4% and other intestinal sites 0.5% (2). Patients with appendiceal endometriosis may be asymptomatic but sometimes atypical symptoms such as right lower abdominal quadrant pain, nausea, melena may present or symptoms of acute appendicitis may develop. There are three theories to explain the origin of this pathology; the first one is implantation with retrograde menstruation from the uterus to the abdominal cavity through the fallopian tubes, the second one is the metaplasia of the coelomic cells and the third theory is systemic metastasis of embolization of endometrial tissue through vascular circulation (1, 6).

Diverticula characterized by the herniation of mucosal surface through the muscularis propria, result from the increased intraluminal bowel pressure. Diverticula of the vermiform appendix either congenital or acquired are rare lesions. While approximately 50 cases are reported for the congenital form, acquired type is more common with a reported incidence ranging from 0.3%-2.2% (4). They are often asymptomatic but chronic intermittent right lower quadrant abdominal pain may develop. When diverticulitis or perforation develops clinical features mimic the acute abdomen of suppurative appendicitis (7). Many mechanisms have been proposed for the pathogenesis of acquired appendiceal diverticulum; including luminal obstruction and/or increased intraluminal pressure and uncoordinated muscular contractions that cause herniation of mucosa through the appendiceal wall (3, 4). A relatively high risk of developing primary appendiceal neoplasms such as neuroendocrine tumors, adenomas and adenocarcinomas in the
patients with appendiceal diverticulosis is reported in the literature (3, 5).

Preoperative radiologic and clinical diagnoses of these two entities are not usually possible and the precise diagnosis is only made by histopathologic examination. Appendectomy is the treatment of choice for both of the entities and our patient was treated by laparoscopic appendectomy with favorable outcome.

The present case with a unique combination of two different entities; appendiceal diverticulosis and appendiceal endometriosis is highly interesting as no known case with this combination is reported in the literature before.

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