APPENDICEAL MUCOCELE DUE TO ENDOMETRIOSIS: A CASE REPORT AND LITERATURE REVIEW

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Rezumat. Mucocel apendicular cauzat de endometrioză: caz clinic şi revista literaturii

Mucocelul apendicular cauzat de endometrioză se întâlneşte extrem de rar, în literatura anglofonă fiind descrise aproximativ 10 cazuri. Autorii raportează un caz de mucocel apendicular cauzat de endometrioză şi prezintă revista literaturii relevante. Pacienta O., 23 de ani a fost spitalizată acuzând dureri în fosa iliacă dreaptă în decurs de 3 luni. Examenul ecografic a pus în evidenţă o formaţiune chistică bine delimitată cu conţinut lichid, cu sept interior, situată retrouterin. A fost stabilit diagnosticul preoperator de chist ovarian şi hidrosalpinx pe dreapta. Intraoperator a fost depistat apendicele vermicular dilatat şi a fost efectuată apendicectomia. În lumenul apedicelui dilatat a fost depistat conţinut mucinos.

Examenul histologic a confirmat un mucocel apendicular cauzat de endometrioză. Evoluţia postoperatorie a fost fără complicaţii. Autorii prezintă o revistă a literaturii pentru elucidarea patogenezei, manifestărilor clinice şi particularităţilor morfologice.

Cuvinte-cheie: mucocel, apendice vermicular, endometrioză

Summary. Mucocele of the appendix due to endometriosis is extremely rare, and there are only about 10 previously reported cases in the English literature

Herein authors report a case of mucocele of the appendix due to endometriosis and provide a review of the literature. A 23-year-old woman was admitted to the hospital because of right lower abdominal pain during last 3 months. Ultrasonography revealed a well defined cystic lesion with a septum with fluid content in the retrouterine position. Consequently, ovarian cyst and hydrosalpinx was suspected preoperatively. During surgery an enlarged appendix was found and appendectomy was performed. The dilated appendix contained mucus. Histopathological examination was consistent with a mucocele of the appendix due to endometriosis. The postoperative course was uneventful. We present a review of the literature to underline the pathogenesis, clinical and morphological features.

Key words: mucocele, appendix, endometriosis

Резюме. Мукоцеле червеобразного отростка в результате эндометриоза: клинический случай и обзор литературы

Мукоцеле червеобразного отростка развившийся в результате эндометриоза встречается крайне редко – в англоязычной литературе описано только около 10 случаев. Авторы описывают клинический случай мукоцеле аппендикса развившийся в результате эндометриоза и приводят обзор литературы. Пациентка О. в возрасте 23 лет поступила с жалобами на боли в правой подвздошной области в течение 3-х месяцев. Ультразвуковое исследование выявило кистозное жидкостное образование с внутренней перегородкой расположенное позади матки. Был установлен предоперационный диагноз: правосторонняя киста яичника и гидросалпинкс. Интраоперационно был выявлен увеличенный аппендикс и проведена аппендэктомия. Просвет червеобразного отростка содержал муцин. Гистологическое исследование выявило простой мукоцеле и эндометриоз червеобразного отростка. Послеоперационный период без осложнений. Авторы представляют обзор литературы с целью ознакомления с патогенезом, клиническими проявлениями и морфологическими особенностями.

Ключевые слова: мукоцеле, аппендикс, эндометриоз

Introduction

Mucocele of the appendix is an uncommon disease. It is observed in 0.2%-0.3% of appendectomies and 8%-10% of appendiceal tumors [1]. According to the modern classification [2, 3], mucocele of the appendix includes four histological groups: simple mucocele, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. Simple mucocele is caused by mucus distention secondary to an obstruction of the appendix due to fecaliths, postinflammatory scarring, or rarely, endometriosis. There are only about 10 previously reported cases of mucocele of the appendix due to endometriosis in the English literature [4-13]. We present a rare case of mucocele of the appendix due to endometriosis and a review of the literature.

Case description

A 23-year-old female patient was referred to our department after a cystic mass was found during an ultrasonography examination which was interpreted as a right sided ovarian cyst. Patient's past medical history was unremarkable, except lower abdominal pain during last 3 months. At admission she presented hypogastric pain, mainly on the right side, physical examination revealed tenderness in this area. Gynecological exam revealed a hard mass in the right anexa. All the routine tests were within normal range. USG revealed a well defined cystic lesion with a septum with fluid content 50x40 mm with thick walls in the retrouterine position (**Fig. 1**). A preoperative diagnosis of right sided ovarian cyst and hydrosalpinx was established and the patient was scheduled for surgery. A vermiform appendix 62x26 mm, with narrow base and significantly enlarged distal half was found on laparotomy. The wall of the appendix was intact, without signs of content leakage. Peritoneal cavity was inspected thoroughly and no signs of mucin peritoneal dissemination were found. Features of endometriosis were not observed in the pelvis, the uterus, or the rest of the abdominal cavity. Appendectomy was performed. After surgery gross examination of the resected specimen showed a distended distal part of the appendix containing mucin with a significantly thickened wall (**Fig. 2**). Histological examination revealed a simple mucocele with endometrial glands within the muscle and serosal layer in the appendiceal wall (**Fig. 3, 4**). The postoperative course was uneventful. At 6 months follow up no signs of peritoneal involvement were found.

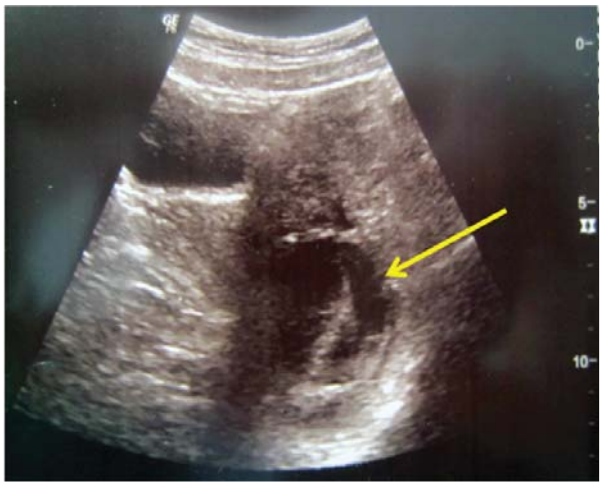

Fig. 1. *Ultrasonography showing a well defined cystic lesion with a septum and fluid content (arrow)*

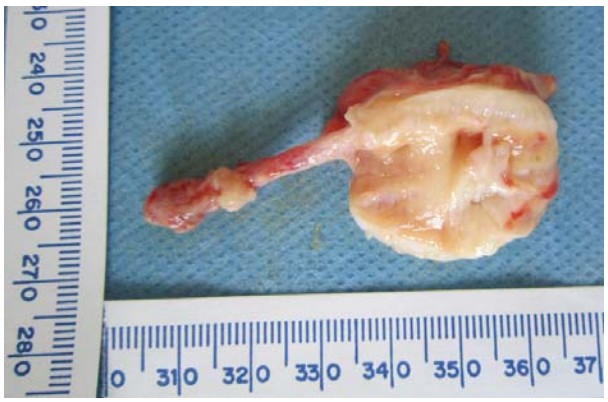

Fig. 2. *Resected specimen showing the enlarged distal part of the appendix with significantly thickened walls and mucin in lumen*

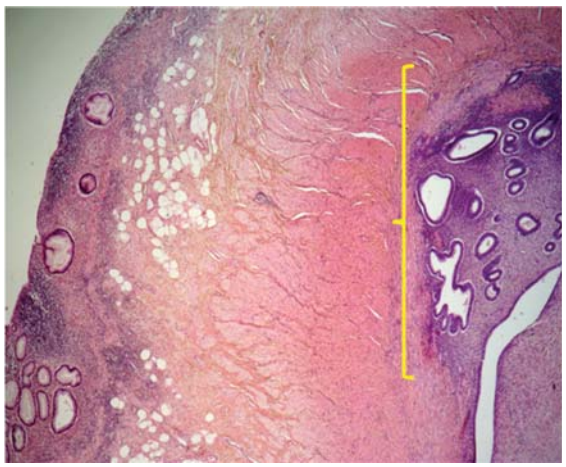

Fig. 3. *Histopathological examination revealed endometriosis with gland and stroma within the appendix seromuscular wall lined with endometrial epithelium (accolade). HE x25*

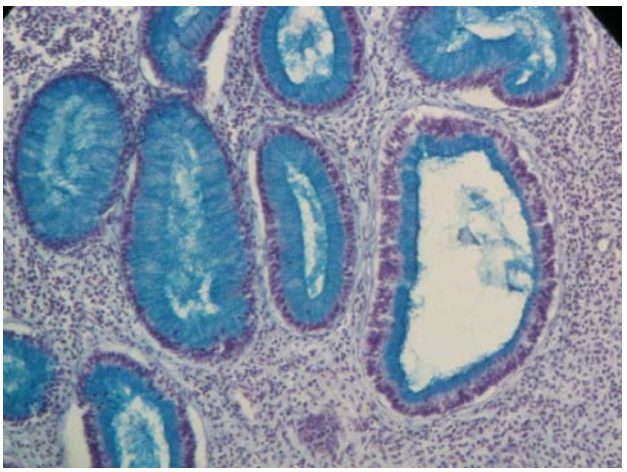

Fig. 4. *Histopathology specimen showing positive coloration for mucin within the epithelial glands. Alcian Blue x100*

Discussion

When the endometrial tissue is found outside the normal place in the uterus, it is named "endometriosis". This condition is seen in 10% of women within their menstrual ages [14, 15]. It is called adenomyosis or internal endometriosis when the endometrial tissue is found within the uteral muscles. External endometriosis is commonly found in genital organs and pelvic peritoneum, though it may be seen in the gastrointestinal system, omentum majus, mesenterium, liver, operation scars and rarely in kidneys, lungs, central nervous system, skin and extremities [16, 17].

Several theories explain the pathogenesis of extrauterine endometriosis [18]. First is the implantati-

on or retrograde menstruation theory that endometrial tissue from the uterus is transported in a retrograde fashion through the fallopian tubes [19]. Second are the direct transplantation and dissemination theories, which may explain extrapelvic endometriosis [20]. Third is the coelomic metaplasia theory that the peritoneal cavity contains progenitor cells capable of differentiating into endometrial tissue [21]. 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 [18].

Endometriosis is seen at a rate of 3–37% in various parts of the gastrointestinal system, from the small intestine to the anal canal and 76% of these occur in the sigmoid colon and rectum [15]. It presents mostly as asymptomatic or with atypical symptoms. Patients may present with symptoms, which are seen in malignant or inflammatory intestinal diseases. Surgeons frequently face endometriosis during a laparotomy performed due to acute abdomen, ileus signs or other reasons. A definitive diagnosis can be reached only through histopathological examination [14, 22]. Appendiceal endometriosis is a rarely seen condition comprising less than 1% of pelvic endometriosis cases [23]. As with other gastrointestinal endometriosis cases, appendiceal endometriosis is generally asymptomatic, too. When it presents with symptoms, they are difficult to differentiate from those of acute appendicitis and definitive diagnosis cannot be made prior to operation [24].

Endometriosis of the appendix is a rare lesion, and is observed in 0.054%-0.8% of all appendectomies performed [25-27], and it was first described in 1860 [28]. Finally, mucocele of the appendix due to endometriosis is extremely rare. A simple mucocele is characterized by degenerative epithelial changes due to obstruction and distention of the appendix. This type represents 20%-30% of cases. A simple mucocele is caused by mucus distention secondary to an obstruction of the appendix due to fecaliths, post-inflammatory scarring, or rarely, endometriosis [13].

Hapke et al. [5] noted that the progression of mucocele of the appendix due to endometriosis consists of the following steps. Endometriosis results in smooth muscle hypertrophy of the appendix, including the muscularis mucosa, with obstruction of some of the gland crypts. These obstructions lead to local increased mucin production from multiple small cysts. Ultimately, several of these small cysts coalesce, resulting in a single layer cyst that can be dissected through the submucosa proximally.

Appendiceal endometriosis patients can be categorized into four groups in terms of symptomatology as follows: patients presenting with acute appendicitis, patients with appendix invagination, patients manifesting atypical symptoms such as abdominal colic, nausea and melena, and asymptomatic patients [29, 30]. The most commonly seen group comprises patients who present with appendicitis to the clinic and the condition mostly occurs during menstruation. Endometriosis is mostly seen in the distal part of the appendix. The underlying causes of appendicitis are endometrium hemorrhagia within the seromuscular layer of appendix and the following edema and inflammation [31].

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 [32]. 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 [33]. Abdomen ultrasonography can be considered for patients who cannot receive APCT.

When mucocele of the appendix is diagnosed preoperatively, open surgery is favored over laparoscopy to prevent rupture of the mucocele, which may induce PMP. If mucocele is detected during a laparoscopic procedure, the patient must undergo conversion to open surgery.

Appendiceal endometriosis is diagnosed pathologically. Glandular tissue, endometrial stroma, and hemorrhage are typically assessed in patients who present with endometriosis [24]. Approximately half of the cases of endometriosis of the appendix involve the body and half involve the tip of the appendix. Muscular and seromuscular involvement occurs in two-thirds of patients, and the serosal surface is involved in one-third of patients. The mucosa is not involved and the submucosa is involved in one-third of patients.

In conclusion, appendiceal endometriosis is rare, mucocele of the appendix due to endometriosis being exceptional, 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.

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