OLGU SUNUMU / CASE REPORT

Gülhane Tıp Derg 2016;58: 190-193

doi: 10.5455/gulhane. 169123

A mucocele of appendix caused by endometriosis presenting clinically with acute appendicitis

Ali Kağan Coşkun(*), Rahman Şenocak(*), Elgün Samadov(*), Şahin Kaymak(*), Zafer KILBAŞ(*), Mustafa Öner Mentes(*), Yusuf Peker(*), Armağan GÜNAL(**)

ÖZET

Akut appendisit semptomlarıyla prezente olan ve endometriozis sonucu oluşan bir appendiks mukoseli

Endometriozise sekonder gelişen apendiks mukoseli oldukça nadir bir durumdur. Mukosel ve endometriozis gibi hastalıklarda hastaların çoğu asemptomatik olup akut appendisit benzeri semptomlarla prezente olurlar. Mukosel benign ya da malign durumlardan biriyle birliktelik gösterebilir. Appendiksin basit mukoseli postinflamatuvar, fekalitler ve endometriozis gibi çok çeşitli obstruktif lezyonlar nedeniyle oluşabilir. Mukoselin rupture olması peritoneal psodomiksomatozis gelişimine neden olabilir. Tedavi, basit appendektomiden daha agresif tedavilere kadar uzananan bir yelpazede değişkenlik gösterir. Bu vakada appendiks endometriozisi nedeniyle gelişen bir basit appendiks mukoseli sunulmuştur.

Anahtar Kelimeler: *Appendiks, Endometriozis, Mukosel*

SUMMARY

Mucocele of the appendix secondary to endometriosis is extremely rare situation. Most of the patients are asymptomatic in mucocele of appendix and appendiceal endometriosis, but they usually present acute appendicitis-like symptoms. Mucocele may be associated with either benign or malignant process. Simple mucocele of the appendix is caused by a variety of obstructive lesions such as postinflamation, fecaliths and endometriosis. The rupture of mucocele may lead to development of pseudomyxoma peritonei. Treatment is variable, extending from simple appendectomy to more aggressive therapies. In this case we present a simple mucocele of appendix secondary to endometriosis of the appendix.

Key words: *Appendix, Endometriosis, Mucocele*

*Gulhane School of Medicine, Department of Surgery.
**Gulhane School of Medicine, Department of Pathology.

Ayrı Basım İsteği: Rahman ŞENOCAK
Gulhane School of Medicine, Department of Surgery.
e-mail: rahmansenocak@yahoo.com

Makalenin Geliş Tarihi: Sep 15, 2014 • Kabul Tarihi: Apr 14, 2015 • Çevrim İçi Basım Tarihi: 10 Haziran 2016

Introduction

Mucocele of appendix (MA) is characterized with an obstructive dilatation of the appendix by intraluminal accumulation of mucoid material. Mucocele is seen 0.3% to 0.7% of all appendectomies (1). Clinical signs of mucocele in most of patients are non-specific, but acute or chronic pain in right iliac fossa is the most frequent symptom. If it is asymptomatic, up to 50% of mucoceles are found at the time of surgery. Majority of mucoceles of appendix are subdivided into four histologic subgroups: simple or retention cysts, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma (1, 2). The clinical course, the surgical treatment and also the prognosis of appendiceal mucoceles are related to their histologic subtype.

Endometriosis is the presence of ectopic endometrial tissues outside of the uterine cavity. It can give rise to chronic pelvic pain and infertility in females. Endometriosis involves in various parts of gastrointestinal system at a rate of 3-37%, mainly in the sigmoid colon and rectum (3). Appendiceal endometriosis is also a rare condition consisting of less than 1% pelvic endometriosis cases and symptomatology is variable (4). It is difficult to distinguish from the symptoms of acute appendicitis and definitive diagnosis cannot be made before surgery (3,4).

A MA and involvement of appendix by endometriosis are both relatively rare diseases (5). Since the appendicitis is commonly encountered in surgical area, the probability of general surgeons facing with a mucocele can also be highly possible. We would like to report a case with mucocele of the appendix secondary to endometriosis presenting as acute appendicitis.

Case Report

A 41-year-old woman was admitted to the emergency unit with right lower abdominal pain. She was diagnosed as acute appendicitis according to her anamnesis, physical examination and laboratory tests. She was on third day of her menstruation cycle. She had no fever and gave no history of previous abdominal pain or gynecologic problem. She had a history of Multiple Sclerosis and received interferon medication and also she gave birth to three children. Physical examination revealed tenderness, muscular defense and positive rebound signs in the right lower abdominal quadrant. The sole pathological laboratory result was mild leukocytosis at a level of 12400 /uL. An uncompressible appendix with 6 cm in lenght and 1 cm in diameter with pericecal heterogeneity were found on abdominal ultrasonography (USG). After the patient was preoperatively consulted with the gynecology department, any abnormalities were declared. An appendicectomy was performed with

diagnosis of acute appendicitis via McBurney incision. During exploration appendix was seen as both grossly inflamed and extremely edematous and enlarged. Postoperative period was uneventful. During the histopathological evaluation of the specimen, mucocele of appendix due to endometriosis was diagnosed. There were no signs of endometriosis in other intra-abdominal locations of the operative field. Our patient's acute symptoms disappeared completely after appendicectomy. The patient was discharged on the 3th day postoperatively without any complications. Postoperatively, patient was consulted to gynecology department regarding of endometriosis, and close follow up without any medication was recommended.

Histopathological examinations were revealed that the diameter of the appendix was 1.2 cm at the tip, but at the body and distal excision area it was about 0.5 cm. The wall got thinned up to 0.1cm at dilated proximal tip. Microscopically, the mucosa was also atrophic with a thinned appendix wall at the tip, lumen contained large amounts of mucus-like material, but there was no epithelial hyperplasia and atypia, consistent with simple mucocele (figure 1). Extensive endometriosis foci were seen in the wall of the body of the appendix (figure 2). Immunohistochemically, endometrial stroma was positive with CD10 (inset-figure 2).

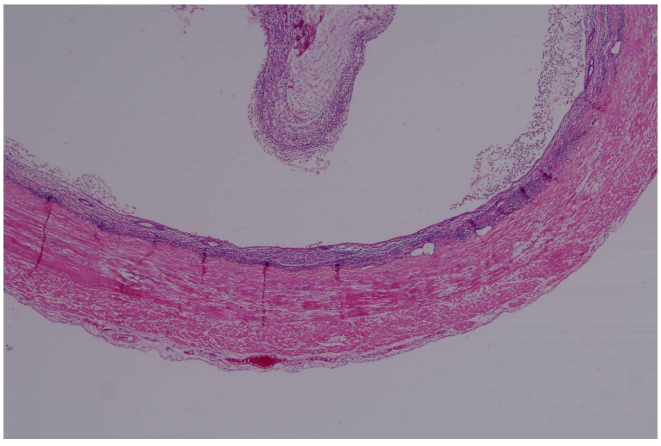

Figure 1: Simple mucocel area with atrophic mucosa and thinned wall at the tip of the appendix (HEx40).

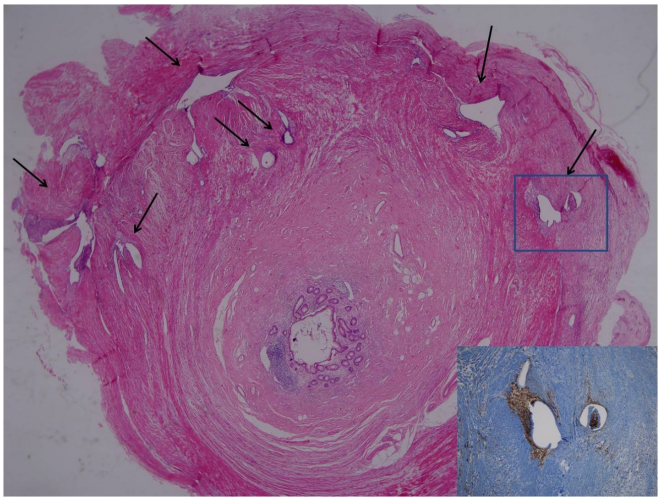

Figure 2: Extensive endometriosis foci **(arrows)** in the wall at the body of the appendix (HEx20). Endometrial stroma was positive with CD10, immunohistochemically, **(inset)** (IHC-CD10x100).

Discussion

A mucocele of the appendix is seen 0.3% of all appendectomies (6) and endometriosis of the appendix is also a rare condition with a rate of 0.8% of all appendectomies (7). Mucocele is a defined for an abnormal mucous accumulation distending the appendiceal lumen regardless of the underlying case. Early reports concluded that most of appendiceal mucoceles were secondary to obstruction, so called obstructive, retension or simple mucoceles. However, more recent studies have shown that secondary obstruction by fecaliths, postinflammatory scarring and endeometriosis is fewer than the other types (8). This type represents 20% of all mucoceles. Such a mucocele presents a plain epithelium, atrophy, and no proliferative changes (2,8,9,10).

A MA can be identified incidentally in radiological or endoscopic studies or at surgery performed for other reasons. Although some authors report that up to 50% of the cases are asymptomatic, acute or chronic pain in right iliac fossa is the main symptom. In our patient, the major symptoms were tenderness, muscular defense and positive rebound signs in right lower quadrant. Unusual manifestations include gastrointestinal bleeding associated with intussusception of mucocele, intestinal obstruction, sepsis, or genitourinary symptoms (11,12,13).

USG, computerized tomography (CT) scan and colonoscopic examinations can help surgeon to diagnose preoperatively mucocele. USG is the first line modality for patients with acute abdominal pain. Appendix with a diameter 15 mm or more has been determined as the treshold for mucocele diagnosis with a sensivity of 83% and a specificity of %92 (15). Sausage-shaped cyctic structure in the appendical region and onion skin-like circles produced by multiple echogenic layers along the dilated appendix may be patognomic for mucocele (16). Souei-Mhiri et al. reported that USG was useful to determine appendiceal abnormalities but did not make an accurate diagnosis (17). USG was diagnostic in 58% of the cases and CT in 89% (11). Therefore, diagnosis of a mucocele is often confirmed by a CT of abdomen. In our case, USG demonstrated uncompressible appendix with tubular distension similar to those with acute appendicitis and no sign of mucocele. For this reason, we did not need to perform other workup such as Alvarado scale and CT for further verification

Abnormal mucoid material accumulation in the apendiceal lumen can be acellular or contain epithelial cells with or without different grades of atypia. Microscopically, retention cysts are lined by flat epithelium, dystrophic mineralization, fibrosis and mucus in the lumen of the cyst. Mucosal hyperplasia is characterized by additional hyperplastic epithelium, a mucinous cystadenoma by cellular atypia, glandular and papillary proliferation [1]. Examination of the appendix during surgery can not tell whether the tumor is benign or malign (18). An intact mucocele is considered to carry no future risk for the patient, but if perforation occurs and epithelial cells escape into the peritoneal cavity, patients may have a different clinical course in which mucinous tumours develop (19). It is important to keep a mucocele intact and to handle tissues carefully and to avoid dissemination of mucoid material into peritoneum. The role of the pathologist is critical and must include study of the visceral peritoneum, looking for a perforation inadvertently caused by the surgeon In our case, histologically this mucocele is characterized by an atrophic mucosa and goblet cell lining

and absence of epithelial atypia. The mucocele appears to be purely obstructive and related to endometriosis. The mucosa of appendix is not affected. Gross macroscopy of appendix did not give us any clue about endometriosis. Pathological examination showed no perforation with intact appendix.

In the management of mucoceles, surgery could be a gold standart treatment modality, because apparently benign lesions can progress to mucinous cystadenocarcinoma, and the rupture of mucocele may determine the development of pseudomyxoma peritonei (11). A simple pathway for treatment of MA has been presented by Dhage-Ivatury and Sugarbaker (19). Simple appendectomy is choice of treatment for benign non perforated mucoceles that have negative cytology and margins.

In spite of an immediate good outcome of operation for mucocele, follow-up is recommended, because there are cases of recurrences as pseudomyxoma peritonei and instances of metachronic colonic neoplasms. Follow-up is recommended in all cases, even those with benign histology (simple mucocele, mucosal hyperplasia, and mucinous cystadenoma), because there are cases reported of development of pseudomyxoma peritonei with these histological types (23).

Endometriotic implants of gastrointestinal tract are estimated to occur in 12-37% of patients with endometriosis. The frequency of endometriosis on appendectomy has been reported to be 0.05% to 0.3% (14).

Involvement of appendix may present as appendicitis, mucocele of appendix or appendicular mass, perforation and cecal intusseption (8,13,20). The most commonly clinic presentation is appendicitis mostly occurring during menstruation. The implants are usually serosal but can eventually erode through the subserosal layers and cause marked thickening and fibrosis of the muscularis propria (21). Whatever the symptomatology the patient has, the treatment for appendicecal endometriosis should be appendectomy. CD10 is a sensitive immunohistochemical marker of endometrial stromal cells at ectopic sites. In cases of suspected endometriosis where the reporting pathologist is unsure whether or not endometrial-type stroma is present, CD10 staining is of value in establishing a definitive diagnosis of endometriosis (22).

The present case shows us that involvement of appendix by endometriosis and subsequently developing mucocele of appendix are relatively rare disease, USG can not be sensitive enough to definitely diagnose the mucocele, lesions smaller than 1.5 cm can also harbour risk of mucocele, and appendicitis like symptoms during menstruation can be related to endometriosis, for these reasons it is hard to diagnose preoperatively mucocele and endometriosis. The critical point is the quick pathologic diagnosis for the surgical decision as only appendectomy or extended surgery should be required.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the authorship and/or publication of this article.

Author Contributions:

Concept – AK COŞKUN

Design – Ş KAYMAK, Z KILBAŞ

Data Collection&/or Processing – A GÜNAL, AK COŞKUN

Analysis&/or Interpretation – R ŞENOCAK

Literature Search –E.SAMADOV, R ŞENOCAK

Writing – R ŞENOCAK

Critical Reviews – Ö MENTEŞ, Y PEKER

References

1. Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix: a re-evaluation of appendiceal mucocele. Cancer 1973;32:1525-41.
2. Landen S, Bertrand C, Maddern GJ, Herman D, Pourbaix A, de Neve A, et al. Appendiceal mucoceles and pseudomyxoma peritonei. Surg Gynecol Obstet 1992;175:401–404
3. Agarwal N, Subramanian A. Endometriosis – Morpholojy, Clinical Presentations and Molecular Pathology. J lab Pyhsicians 2010;2:1-9.
4. Mann WJ, Fromowitz F, Saychek T, Madariaga JR, Chalas E.Endometriosis associated with appendiceal intusseption. J Reprod Med 1984;29:625-629
5. Hapke MR, Bigelow B. Mucocele of the appendix secondary to obstruction by endometriosis. Hum Pathol 1977;8:585-589
6. García Lozano A, Vázquez Tarrago A, Castro García C, Richart Aznar J, Gómez Abril S, Martínez Abad M. Mucocele of the appendix: Presentation of 31 cases. Cir Esp 2010; 87: 108-112 7.Berker B, Lashay N, Davarpanah R, Marziali M, Nezhat CH, Nezhat C. Laparoscopic appendectomy in patients with endometriosis. J Minim Invasive Gynecol 2005; 12: 206-209
8. Driman DK, Melega DE, Vilos GA, Plewes EA. . Mucocele of Appendix Secondary to Endometriosis. Am J Clin Pathol 2000;113:860-864
9. Carr NJ, McCarthy WF, Sobin LH. Epithelial noncarsinoid tumors and tumor like lesions of the appendix:a clinicopathologic study of 184 patients with multivariate analysis of prognostic factors. Cancer. 1995;75:757-768
10. Isaacs KL, Warshauer DM. Mucocele of the appendix: computed tomographic, endoscopic and pathologic correlation. Am J Gastroenterol 1992;87:787–789.
11. Ruiz-Tovar J, Teruel DG, Castiñeiras VM, Dehesa AS, Quindós PL, Molina EM. Mucocele of the Appendix. World J Surg 2007;31: 542–548
12. Serrano Sa´nchez PA, Pe´rez-Bedmar JA, Larran˜aga Barrera E. Mucocele apendicular. Revisio´n de la literatura y aportacio ´n de 8 casos. Rev Esp Enferm Dig 1989;76:35–41
13. Kimura H, Konishi K, Yabushita K, Maeda K, Tsuji M, Miwa A.. Intussusception of a mucocele of the appendix secondary to an obstruction by endometriosis:report of a case. Surg Today 1999;29:629-632
14. Akagi T, Yamamoto S, Kobayashi Y, Fujita S, Akasu

T, Moriya Y, Kato T A case of endometriosis of the appendix with adhesion to right ovarian cyst presenting as intussusception of a mucocele of the appendix. Surg Laparosc Endosc Percutan Tech. 2008:18:622-625

15. Lien WC, Huang SP, Chi CL, Liu KL, Lin MT, Lai TI et al. Appendiceal outer diameter as an indicator for differentiating appendicecal mucocele from appendicitis. Am J Emerg Med 2006;24:801-805
16. Miraliakbari R, Chapman WH. Laparoscopic treatment of an appendiceal mucocele. J Laparoendosc Adv Surg Tech A. 1999;9:159-163
17. Soueï-Mhiri M, Tlili-Graies K, Ben Cherifa L, Derbel F, Hmissa S, Dahmen Y et al. Mucocele of the appendix. Retrospective study of 10 cases. J Radiol 2001;82:463–468.
18. Misdraji J, Yantiss RK, Graeme-Cook FM, Balis UJ, Young RH: Appendiceal mucinous neoplasms: a clinicopathologic analysis of 107 cases. Am J Surg Pathol 2003;27:1089–1103
19. Dhage-Ivatury S, Sugarbaker PH. Update on the surgical Approach to mucocele of the appendix. J Am Coll Surg 2006;202;680-684
20. Nopajaroonsri C, Mreyoud N. Retention mucocele of appendix due to endometriosis. South Med J. 1994;87:833-5
21. Uncu, H, Dilara T. Appendiceal endometriosis:two case reports. Arch Gynecol Obstet 2008;278:273-275
22. Sumathi VP, McCluggage WG. CD10 is useful in demonstrating endometrial stroma at ectopic sites and in confirming a diagnosis of endometriosis. J Clin Pathol 2002;55:391-392
23. Smith JW, Kemeny N, Caldwell C, Banner P, Sigurdson E, Huvos A. Pseudomyxoma peritonei of appendiceal origin. The Memorial-Sloan Kettering Cancer Center experience. Cancer 1992;70(2):396–401.