KJCO Korean Journal of Clinical Oncology

Korean Journal of Clinical Oncology 2022;18:83-88
https://doi.org/10.14216/kjco.22011
pISSN 1738-8082 · eISSN 2288-4084

Case Report

Appendiceal mucocele masquerading as an epithelial borderline ovarian tumor: a case report and literature review

Young Joo Kim, Jong Hyuk Yun, Sung Hoon Hong, GeumJong Song, Jong Eun Lee, Myong Won Son, Sun Wook Han, Sung Yong Kim, Moon-Soo Lee

Department of Surgery, Soonchunhyang University Cheonan Hospital, Cheonan, Korea

Appendiceal mucocele is a rare mucin-producing neoplasm of appendiceal origin. Due to its location and imaging findings, appendiceal mucocele is easily confused with tumors of the right adnexa. We present a rare case of a patient initially misdiagnosed with an ovarian tumor intraoperatively diagnosed as an appendiceal mucocele and successfully treated. A 66-year-old postmenopausal woman was admitted to the gynecology department for an asymptomatic pelvic mass. Preoperative pelvic imaging showed an 8-cm cystic mass. Exploratory laparoscopy for the suspected epithelial borderline tumor from the right ovary revealed a cystic mass in the right pelvic area and normal uterus, fallopian tubes, and ovaries. Intraoperative consultation with the general surgery department confirmed the appendiceal origin. Laparoscopic appendectomy was performed. Histopathological examination confirmed a low-grade mucinous neoplasm of appendiceal origin. The patient was discharged on a postoperative day 5 without complications. The outpatient follow-up performed 1 month later showed no evidence of disease progression. Despite the use of advanced diagnostic tools, appendiceal mucocele may be confused for ovarian malignancies. Because the clinical features of appendiceal mucocele are nonspecific, clinicians and radiologists know the specific imaging findings. A multidisciplinary approach including general surgery, gynecology, and radiology is required for preoperative diagnosis and treatment.

Keywords: Appendiceal neoplasm, Case reports, Ovarian neoplasms, Mimic

INTRODUCTION

Clinicians encountering patients with pelvic masses should consider diseases originating from the urogenital or colorectal regions, including tumors originating from the appendix. Appendiceal mucocele is a rare mucin-producing neoplasm of appendiceal origin. Because no clinical symptoms distinguish it from other pelvic diseases, the diagnosis of appendiceal mucocele can be challenging for many clinicians. Due to its location and imaging findings, appendiceal mucocele is easily confused with tumors originating from the right adnexa, such as right ovarian benign masses or cysts, or even malignancy [1,2]. We report a case of a patient with an appendiceal mucocele initially misdiagnosed as an ovarian malignancy before surgery. This is a rare case of a mistaken ovarian tumor because all preoperative imaging tests, including magnetic resonance imaging (MRI), suggested a right borderline tumor of ovarian origin. Informed consent was obtained from patient involved in the study.

Received: Oct 7, 2022 **Revised:** Dec 2, 2022 **Accepted:** Dec 15, 2022
Correspondence to: Jong Hyuk Yun
Department of Surgery, Soonchunhyang University Cheonan Hospital, 31 Suncheonhyang 6-gil, Dongnam-gu, Cheonan 31151, Korea
Tel: +82-41-570-3638, **Fax:** +82-41-571-0129
E-mail: mdyoon87@schmc.ac.kr

CASE REPORT

A 66-year-old postmenopausal woman was admitted to the gynecology department for an asymptomatic pelvic mass incidentally discovered during a medical check-up. She had a medical history of hypertension and asymptomatic carotid artery stenosis. Her personal surgical and familial histories were unremarkable. Physical examination revealed a non-tender, palpable mass in the right lower quadrant. The results of the other abdominal examinations were unremarkable. Gynecological examination showed a normal uterus and cervix.

In the initial laboratory tests, the complete blood and white

blood cell differential counts were unremarkable. The chemistry, coagulation panels, and urine analysis were also unremarkable. The levels of tumor markers, including carcinoembryonic antigen (CEA; 3.95 ng/mL), carbohydrate antigen 19-9 (CA19-9; 9.41 U/mL), and cancer antigen 125 (CA125; 5.84 U/mL), were normal.

The transvaginal ultrasound showed a normal uterus, with a well-defined mass with heterogenous echogenicity on the right side, measuring 80×54 mm. The left adnexa was normal in ultrasound. Preoperative pelvic MRI showed an 8-cm cystic mass with high signal intensity in T2-weight imaging. The mass had internal septa with subtle enhancement. The formal reading by a radiologist was an epithelial borderline tumor from the right ovary. No enlarged lymph nodes or ascites were observed in the pelvic and abdominal areas. The left ovary and uterus were also unremarkable (Fig. 1).

Explorative laparoscopy was performed by the gynecologic department. The intraoperative findings included a huge cystic mass in the right pelvic area. The uterus and both fallopian tubes and ovaries were normal. The other specific findings were unremarkable. The general surgery department was called into the operating room to confirm that the mass originated from the appendix. Both the cystic portion and the base of the appendix were enlarged to similar diameters similar to that of the cecum. Serosal invasion, regional lymph node enlargement, and ascites were not observed. There were no suspicious findings of peritoneal seeding in peritoneum and liver. Based on these findings, laparoscopic appendectomy was performed using a linear stapler (Powered ECHELON FLEX stapler, Ethicon), without any damage to the adjacent organs, including the cecum and ovary.

Gross examination revealed an enlarged appendix measuring 9.5×5.0×5.0 cm. The distal portion was whitish in color had had a huge cystic feature, while the proximal portion had a serosal appearance and was enlarged. The lumen was filled with mucoid material and was not perforated (Fig. 2).

Histopathological examination confirmed a low-grade mucinous neoplasm (LAMN) originating from the appendix. The resection margin was clear. The patient was discharged on the fifth postoperative day without any complications. The outpatient follow-up performed approximately 1 month later showed no evidence of disease progression or any other symptoms. Colonoscopy was performed for postoperative follow-up 6 months later and there were no remarkable findings.

DISCUSSION

Appendiceal mucocele may result from the accumulation of mucin that causes obstruction and dilation of the appendix and cecum. This condition is observed in 0.2% to 0.7% of all appendix

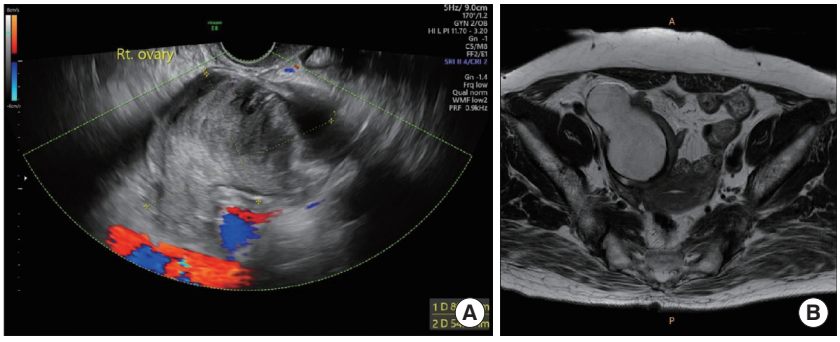

Fig. 1. Preoperative findings in imaging studies. (A) Right adnexal mass in transvaginal ultrasonography. (B) Right pelvic mass in T2-weighted imaging of pelvic magnetic resonance imaging.

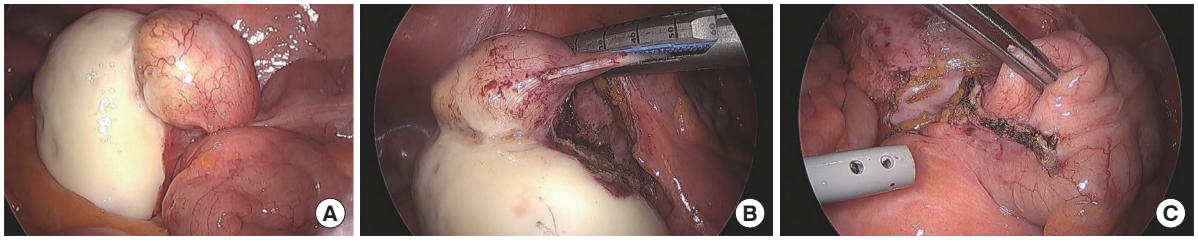

Fig. 2. Intraoperative finding during a diagnostic laparoscopy. (A) A huge cystic mass originating from the appendix is observed. The right adnexa and uterus are normal. (B) The appendix base was intact, laparoscopic appendectomy using linear stapler. (C) Laparoscopic appendectomy was performed.

specimens. It can be asymptomatic or present a variety of symptoms. Appendiceal mucocele may be discovered incidentally in radiologic or endoscopic tests or during surgery for other reasons [3,4]. Appendiceal masses usually show similar symptoms to appendicitis or adnexal masses, including pain in the right lower quadrant or pelvic area and palpable masses or enlarged intra-abdominal structures in imaging studies such as computed tomography (CT) or ultrasound.

Previous reports have proposed ultrasound as a preliminary diagnostic tool. A threshold appendix diameter of ≥ 15 mm for the diagnosis of appendiceal mucocele has an 83% sensitivity and 92% specificity. CT is the method of choice for better characterization of appendiceal mucocele [5]. Other reports have suggested that if appendiceal mucocele is suspected, and there are no abnormalities in the right ovary in imaging studies, it may be helpful to identify the "layered structure (onion skin sign)" by transabdominal ultrasound. Previous studies reported mucinous tumors in the appendix and ovary of 16 of 89 female patients. Moreover, the onion skin sign was present in patients with appendiceal mucocele but not in those with ovarian mucinous tumors. Thus, even when normal ovaries are not detected, clinicians observing the onion skin sign in the lower abdominal area by ultrasound should strongly consider appendiceal mucocele [5,6]. Moreover, if appendicitis is suspected and the diameter of the appendix on the CT scan is ≥ 1.3 cm, appendiceal mucocele should also be considered [5].

A literature review of the PubMed/MEDLINE, Google Scholar, and Web of Science databases from 2000 to 2022 as searches of Science Citation Index (SCI) and Scopus journals revealed similar cases of appendiceal mucocele mimicking adnexal lesions, as summarized in Table 1 [2,4,7-14]. A total of 18 cases of tumors originating from the appendix were preoperatively diagnosed as tumors originating from the adnexa. Most patients showed abdominal pain or palpable masses; however, five cases were asymptomatic and incidentally diagnosed. For preoperative diagnosis, ultrasound was performed in 15 cases (11 cases by transvaginal, 4 cases by transabdominal), CT was performed in seven cases, and MRI was performed in six cases.

In the present case, imaging studies, including ultrasound and MRI, were performed. However, the results indicated a right ovarian borderline tumor and the appendix appeared unremarkable. In the operative findings, the appendix looked like a tubular structure owing to the enlarged appendiceal orifice. Therefore, the radiologic characteristics differentiating the appendix from the cecum by MRI might have disappeared. Laparoscopic appendectomy was performed in six cases, conventional open appendectomy was performed in nine cases, and right hemicolectomy was performed in three cases. The cases that underwent right hemicolectomy were diagnosed with LAMN based on the findings of the pathologic reports.

The surgical method was determined according to the lesion characteristics and size as identified in preoperative imaging. In operative findings, if a perforated appendix, positive pelvic fluid cytology, or enlarged periappendiceal lymph node are observed surgeons should perform a right hemicolectomy. Otherwise, patients require only appendectomy or cecectomy [3]. In three cases that underwent a right hemicolectomy and in which LAMN was diagnosed the patients were overtreated in emergency operations. However, in our case, the gynecology department quickly detected the normal findings of the female genital organs and called the general surgery department to the operating room to avoid inappropriate procedures. Thus, appropriate minimal invasive surgery was performed in this elderly patient. However, a multidisciplinary team approach may have preoperatively determined a proper management plan.

Measurement of tumor markers was selectively performed according to the nature and size of the preoperative lesion and patient history. In most cases, the tumor marker levels were in the normal range; however, two cases showed elevated levels. The patients in those cases were diagnosed with malignancy and had a history of surgery with malignant ovarian teratoma.

There remains controversy regarding the usefulness of tumor markers. Some studies have been reported the diagnostic and prognostic value of tumor markers such as CEA and CA19-9 in appendiceal mucocele. However, other studies reported that they did not significantly improve diagnosis and prognosis. In the present case, the CEA and CA19-9 and CA125 levels were unremarkable [10].

Appendiceal mucocele can cause terrible complications such as peritoneal seeding or pseudomyxoma peritonei if appendiceal mucocele ruptures in the intra-abdominal space during surgery, remains in the surgical margin, or mucinous adenocarcinoma is confirmed. Histologic type is an important factor for planning treatment options including the range of surgical resection. Unfortunately, there remains controversy regarding histological classifications and terminology, which are not yet established for appendiceal mucinous neoplasm in various academic societies.

The most recently announced classification was released in 2010 by the World Health Organization (WHO), which included three categories of appendiceal mucocele: mucinous adenoma, low-grade appendiceal mucinous neoplasm, and mucinous adenocarcinoma. In 2016, the Modified Delphi Consensus was released by the Peritoneal Surface Oncology Group International (PSOGI). In addition to the three categories included in the WHO classification, the histological characteristics were further subdivided for a

Table 1. Reviews of 18 cases of appendiceal mucocele mimicking adnexal disease

Author (year)	Symptoms	Imaging modality	Preoperative diagnosis	Tumor marker	Surgery	Pathology
Borges et al. (2021) [4]	-	TVUSG	Right ovarian cyst	CA125: normal	Laparoscopic appendectomy	Mucinous cystadenoma
	-	TVUSG, MRI	Left ovarian tumor	CA125, CA15-3, AFP: normal	Open appendectomy	LAMN
	RLQ pain	CT	Ruptured ovarian cyst	NA	Laparoscopic appendectomy	Mucinous cystadenoma
	No symptom	TVUSG, MRI	Right ovarian mass	CA125, CEA, CA19-9, CA15-3: normal	Open appendectomy	Mucinous cystadenoma
	RLQ pain	TVUSG	Right ovarian cyst	NA	Laparoscopic appendectomy	Mucinous cystadenoma
	RLQ pain	MRI, TAUSG, CFS	Right ovarian cyst	CEA, CA19-9: normal	Right hemicolectomy	LAMN
	Lower abdominal pain, fever	MRI, TVUSG	Right ovarian cyst	CA125, CA19-9, CEA: normal	Open appendectomy	Retention cyst
	Pelvic pain	TVUSG, MRI	Right adnexal mass	CA 125: normal	Open appendectomy	LAMN
Onyishi et al. (2022) [12]	RLQ mass	TAUSG, CT	Right ovarian mass	NA	Right hemicolectomy	LAMN
Tanaka et al. (2011) [13]	-	NA	Right ovarian teratoma	NA	Open appendectomy	LAMN
Lee et al. (2021) [11]	Abdominal pain	TVUSG, CT	Right ovarian mass	CA125, CA19-9: normal	Laparoscopic appendectomy	Mucinous cystadenoma
Cubro et al. (2019) [8]	RLQ pain, pelvic pain, vomit, fever	TVUSG	Right Tubo-ovarian abscess	NA	Open appendectomy	LAMN
	RLQ mass	TVUSG	Right ovarian mass	NA	Laparoscopic appendectomy	Mucinous cystadenoma
Dellaportas et al. (2014) [9]	RLQ mass	TAUSG, CT	Right ovarian cyst	NA	Laparoscopic appendetomy	Mucinous cystadenoma
Kumar et al. (2020) [10]	Abdominal distention	TAUSG, CT	Both ovarian tumor	CA125: 149.5 U/mL, CA19-9: 4.89 U/mL, CEA: 100 ng/mL, AFP 1.70 ng/mL	Open appendectomy	LAMN
Toffaha et al. (2020) [14]	Vaginal bleeding, abdominal pain, vomit	TVUSG, MRI	Right adnexal mass	CA19-9, CA125, CA15-3: normal	Open appendectomy	LAMN
Perivoliotis et al. (2021) [2]	No symptom	TVUSG, CT	Right ovarian cancer	NA	Right hemicolectomy	LAMN
Ayadi et al. (2022) [7]	RLQ pain	CT	Right ovarian cancer	CEA 138.8 ng/mL, CA125 306.60 U/mL, CA19-9 527.85 U/mL	Open appendectomy	Appendiceal mucinous cystadenocarcinoma

RLQ, right lower quadrant; TVUSG, transvaginal ultrasonography; MRI, magnetic resonance imaging; CT, computed tomography; TAUSG, transabdominal ultrasonography; NA, not available; CFS, colonofiberscopy; CA125, cancer antigen 125; CA19-9, carbohydrate antigen 19-9; CA15-3; cancer antigen 15-3; AFP, alpha fetoprotein; CEA, carcinoembryonic antigen; LAMN, low-grade appendiceal mucinous neoplasm.

total of eight categories [15].

The literature review revealed seven cases of appendiceal mucinous cystadenoma, nine cases of LAMN, one case of appendiceal mucinous adenocarcinoma, and one case of retention cyst. The diagnosis of LAMN in the present case was pathologically confirmed.

While there is no standard surgical treatment protocol and margin positivity, many surgeons have performed and recommended surgical treatment depending on the histological classification. Patients with LAMN usually require appendectomy alone, while patients with mucinous adenocarcinoma require right hemicolectomy. If peritoneal seeding or pseudomyxoma peritonei are suspected, surgeons should consider debulking surgery or hyperthermic intraperitoneal chemotherapy [5].

Appendiceal mucocele is a rare disease that is easily confused with other pelvic diseases. Therefore, physical examinations, proper imaging studies, and laboratory tests including tumor markers are helpful to determine the management plan.

However, in this case, even with sufficient imaging studies, examinations, and imaging readings by a well-trained radiologist, the preoperative diagnosis was not accurate. Surgeons encountering masses in the pelvic area should be aware of the various possibilities, including appendiceal mucocele.

Therefore, it is important to determine the origins of lesions in the pelvic cavity. A proper preoperative clinical work-up and management plan should be performed, and a multidisciplinary team approach involving gynecologists, surgeons, radiologists, and pathologists is needed to provide better detection and treatment for rare conditions such as our case. This manuscript introduces the interesting case and imply on the importance of caution for the possibility of unexpected pathology. Because of the risk of spillage of mucin from appendiceal mucocele during surgery, gentle touch and complete excision of appendix including tumor is needed.

CONFLICT OF INTEREST

Jong Hyuk Yun is an editorial board member of the journal but was not involved in the peer reviewer selection, evaluation, or decision process of this article. No other potential conflicts of interest relevant to this article were reported.

FUNDING

This work was supported by a Soonchunhyang University Research Fund.

ORCID

Young Joo Kim https://orcid.org/0000-0001-8374-5486
Jong Hyuk Yun https://orcid.org/0000-0002-8746-4945
Sung Hoon Hong https://orcid.org/0000-0002-3279-3581
Geum Jong Song https://orcid.org/0000-0002-1067-8099
Jong Eun Lee https://orcid.org/0000-0003-2938-4960
Myong Won Son https://orcid.org/0000-0001-5255-9234
Sun Wook Han https://orcid.org/0000-0002-0671-1212
Sung Yong Kim https://orcid.org/0000-0002-2070-0915
Moon-Soo Lee https://orcid.org/0000-0002-3955-1331

REFERENCES

1. Panarelli NC, Yantiss RK. Mucinous neoplasms of the appendix and peritoneum. Arch Pathol Lab Med 2011;135:1261-8.
2. Perivoliotis K, Christodoulidis G, Samara AA, Sgantzou IK, Floros T, Volakakis G, et al. Low-grade appendiceal mucinous neoplasm (LAMN) primarily diagnosed as an ovarian mucinous tumor. Case Rep Surg 2021;2021:5523736.
3. Dhage-Ivatury S, Sugarbaker PH. Update on the surgical approach to mucocele of the appendix. J Am Coll Surg 2006;202:680-4.
4. Borges AL, Reis-de-Carvalho C, Chorao M, Pereira H, Djokovic D. Low-grade mucinous appendiceal neoplasm mimicking an ovarian lesion: a case report and review of literature. World J Clin Cases 2021;9:2334-43.
5. Sharma P, Soin P, Chugh M, Goyal P. Dilated appendix: is there more to it? Case report and brief review of literature with radiologic-pathological correlation. J Clin Imaging Sci 2019;9:9.
6. Kameda T, Kawai F, Taniguchi N, Omoto K, Kobori Y, Arakawa K. Evaluation of whether the ultrasonographic onion skin sign is specific for the diagnosis of an appendiceal mucocele. J Med Ultrason (2001) 2014;41:439-43.
7. Ayadi C, Naggar A, Andour H, Chraa FZ, Jerguigue H, Latib R, et al. Appendiceal mucocele with pseudomyxoma peritonei mimicking ovarian tumor with peritoneal carcinomatosis. Radiol Case Rep 2022;17:3000-4.
8. Cubro H, Cengic V, Burina N, Kravic Z, Beciragic E, Vranic S. Mucocele of the appendix presenting as an exacerbated chronic tubo-ovarian abscess: a case report and comprehensive review of the literature. Medicine (Baltimore) 2019;98:e17149.
9. Dellaportas D, Vlahos N, Polymeneas G, Gkiokas G, Dastamani C, Carvounis E, et al. Collision tumor of the appendix: mucinous cystadenoma and carcinoid: a case report. Chirurgia (Bucur) 2014;109:843-5.
10. Kumar T, Khanna P, Nigam JS, Nishit, Singh A, Agrawal N. Mucinous appendiceal tumor presenting as bilateral adnexal mass. J

Gastrointest Cancer 2021;52:347-50.
11. Lee SY, Cho DH. Giant appendiceal mucocele mimicking an ovarian tumour. BMJ Case Rep 2021;14:e243753.
12. Onyishi NT, Edeh AJ, Dim NR. Sclero-hyalinized low-grade appendiceal mucinous neoplasm clinically mimicking an ovarian mass. Case Rep Pathol 2022;2022:9404615.
13. Tanaka H, Kobayashi T, Yoshida K, Asakura T, Taniguchi H, Mikami Y. Low-grade appendiceal mucinous neoplasm with disseminated peritoneal adenomucinosis involving the uterus, mimicking primary mucinous endometrial adenocarcinoma: a case report. J Obstet Gynaecol Res 2011;37:1726-30.
14. Toffaha A, El Ansari W, Aleter A. What you see might not be what you get: discrepancies between intraoperative findings and preoperative diagnosis of ovarian tumors: appendicular mucocele presenting as an adnexal mass: case report and review of literature. Int J Surg Case Rep 2020;75:543-9.
15. Gundogar O, Kimiloglu E, Komut N, Cin M, Bektas S, Gonullu D, et al. Evaluation of appendiceal mucinous neoplasms with a new classification system and literature review. Turk J Gastroenterol 2018;29:533-42.