CASE REPORT

Villous Mucinous Cystadenoma of the Appendix in a Postmenopausal Woman

K. Nouri, M. Demmel, J. Ott, R. Promberger, J.C. Huber, K. Mayerhofer

ABSTRACT

Objective: To present the case of a postmenopausal woman, who was suspected of having an ovarian cyst. Instead, a cystadenoma of the appendix was discovered during laparoscopy.

Methods: A 64-year-old postmenopausal nulliparous woman was admitted to our hospital because of a cystic lesion, which had been detected in the course of a routine gynecological examination. The patient underwent vaginal ultrasound, magnetic resonance tomography, and laparoscopy.

Results: During vaginal ultrasound, a dumbbell-shaped anechogenic cystic structure 70 x 32 x 22 mm in diameter was found in the region of the right adnexa. Magnetic resonance tomography revealed no additional information. During diagnostic laparoscopy, the cystic lesion was found to be a distended appendix. A laparoscopic appendectomy was performed. Subsequent histological analysis revealed a villous mucinous cystadenoma of the appendix with low-grade intraepithelial neoplasia.

Conclusion: Gynecologists should routinely consider this disease in the differential diagnosis of right lower dumbbell abdominal cysts. Eleven percent to 20% of mucoceles are caused by mucinous cystadenocarcinomas, which carry the risk of peritoneal tumor implantation caused by rupture or laparoscopic resection. Therefore, it should be mandatory that a general surgeon be involved in the laparoscopic procedure and the conversion to laparotomy for resection of the structure.

Key Words: Mucocele, Cystadenoma, Appendix, Laparoscopy, Ultrasound, Ovarian cyst.

INTRODUCTION

Rarely, mucocele of the appendix is caused by mucinous cystadenoma and is thus regarded as a "diagnostic dilemma."[1] Gynecologists perform vaginal ultrasound examinations as a standard procedure to assess pelvic tumors originating from the ovaries, the tubes, and the uterus. Masses that are detected in the pelvis, but are not related to these structures, might consequently be misjudged. Therefore, a definite diagnosis may not be made until surgery and may then force the surgeon to make important decisions quickly. We report the case of a postmenopausal woman who underwent laparoscopy. Instead of the suspected ovarian cyst, a cystadenoma of the appendix was discovered intraoperatively.

CASE REPORT

A 64-year-old postmenopausal nulliparous woman was admitted to Landsklinikum Wolfsberg, a primary care center in Carinthia, Austria, because her gynecologist had detected a cystic lesion in the course of a routine gynecological examination. The patient did not suffer from any abdominal pain or digestive irregularities, such as obstipation or diarrhea. She had not undergone any gynecological operations so far.

On vaginal ultrasound, a dumbbell-shaped anechogenic cystic structure with a diameter of 70 x 32 x 22 mm was located in the region of the right adnexa. The structure showed a regular wall and no solid intracystic elements. Neither an infiltration of the surrounding tissues nor a connection to the uterus was found. No free fluid was detected, and the left ovary did not exhibit any pathological findings. Palpation neither revealed the pelvic mass nor caused any pain.

To help exclude a magnetic resonance tomography of the

Department of Gynaecologic Endocrinology and Reproductive Medicine, Medical University Vienna, Vienna, Austria (Drs. Nouri, Ott, Huber, Mayerhofer);

Department of Gynecology and Obstetrics, LKH Wolfsberg, Carinthia, Austria (Dr. Demmel);

Department of Surgery, Medical University Vienna, Vienna, Austria (Dr. Promberger).

Address correspondence to: Dr. Kazem Nouri, Department of Gynaecologic Endocrinology and Reproductive Medicine, Medical University of Vienna, Währinger Gürtel 18-20, 1090 Vienna, Austria. Telephone: +431 / 40400 2816, Fax: +431 / 40400 2817, E-mail: kazem.nouri@meduniwien.ac.at

DOI: 10.4293/108680810X12785289144917

lower abdomen was performed, revealing a dumbbell cystic structure (diameter, 77 x 32 x 22 mm) with liquid content in the right pelvis. The pelvic wall showed no signs of penetration. No definite connection to the ovary or to the uterine tube could be identified. The lesion was located ventral of the iliacal bifurcation. No signs of malignancy were detectable. Testing for tumor markers revealed elevations of CA -125U/mL to 71.7 U/mL (normal range, 0 to 33) and of TPS (Tissue polypeptide-specific Antigen) to 91.7U/L (normal range, 0 to 83).

A diagnostic laparoscopy was performed to assess the exact location, and the benign or malignant nature of the cystic lesion. The uterus and both adnexa displayed no indication of pathologies, however. In fact, the cystic lesion was a dumbbell distended appendix of rugged consistency with an estimated diameter of 70 x 40 x 30 mm **(Figure 1)**. The appendix wall seemed regular without any signs of inflammation or malignancy. There were no adhesions between the appendix and other pelvic structures. Judging from these findings, we decided to perform a laparoscopic appendectomy, because we have extensive experience in conducting this procedure. However, we did not consider the possibility malignancy.

The procedure was uncomplicated. After skin incision, we placed a 10-mm trocar in a suprapubic position and two 5-mm trocars at a suprailiac crest level on the left and right side. Because it was not possible to grasp the appendix directly with only one atraumatic grasper, we held it towards the abdominal wall between 2 graspers (ENDO GRASP 5mm, Covidien, Mansfield, MA). Subsequently, we created a mesenteric window, almost 1cm in diameter, beneath the base of the appendix with a dolphin nose grasper (ENDO DISSECT 5mm, Covidien, Mansfield, MA). The appendix was then transected at the base by using an endoscopic stapler (ENDO GIA, Universal Covidien, Norwalk, CT). Afterwards, the appendix was dissected from the gastrointestinal tract and removed with a laparoscopic retrieval pouch (ENDOBAG 3“ x 6”, Covidien, Mansfield, MA), which had been inserted through the suprapubic trocar.

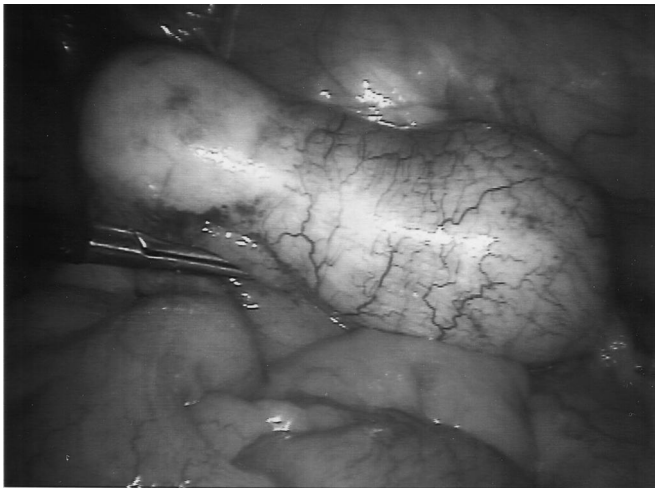

Figure 1. Dumbbell distended appendix.

Histology revealed a villous mucinous cystadenoma of the appendix with low-grade intraepithelial neoplasia.

DISCUSSION

A mucocele of the appendix is a distended appendix caused by intraluminal accumulation of mucus, which is subsequent to 1 of 4 primary conditions, as follows: (1) a simple retention cyst, characterized by a normal epithelium, usually associated with mild luminal dilatation not exceeding a diameter of 2cm. Mucoceles with a diameter of >2cm are likely caused by one of the other 3 diseases. (2) mucosal hyperplasia is found in 5% to 25% of mucocele cases. (3) a mucinous cystadenoma, as found in our patient, accounts for 63% to 84% of mucoceles and is characterized by low-grade epithelial dysplasia. Its histological appearance is similar to villous adenomas and adenomatous polyps of the colon. (4) 11% to 20% of mucoceles are caused by mucinous cystadenocarcinomas. Their characteristic neoplastic alterations of the epithelium are similar to those found in adenocarcinomas of the colon.[2-5] Spontaneous rupture reportedly occurs in up to 6% of cases due to severe luminal distension.

Appendectomy is known as a cure for benign diseases even after rupture has occurred, leading to mucinous ascites. Cystadenocarcinomas, however, carry the risk of peritoneal tumor implantation caused by rupture or laparoscopic resection. This may lead to pseudomyxoma peritonei characterized by the presence of mucin-secreting cellular elements in the peritoneal cavity. As cystadenocarcinoma can be the underlying disease, Matthews and Hodin[6] mention mucocele of the appendix as a contraindication for laparoscopic appendectomy. Survival at 5 years is reported as low as 50%.[6] Adenocarcinoma of stage Duke A, ie, early lesions confined to the mucosa and submucosa, may be treated by simple appendectomy. However, for stages Duke B and C, right hemicolectomy has to be performed, followed by adjuvant therapy.[6]

In our case, the decision on laparoscopic resection was made based on our extensive experience in laparoscopic appendectomy. Consulting a general surgeon was not

possible during the operation. The operating team was not aware of the fact that laparoscopic resection would have been a grave error in treatment in case of a mucinous cystadenocarcinoma. The reported case is an example of what is called a "near miss" in terms of quality control and risk management. Thus, we consider it necessary to raise gynecologists' awareness of this disease pattern.

Because the disease is of a nonspecific nature, preoperative diagnosis is difficult. Pelvic masses are usually preoperatively diagnosed by ultrasound, in which mucoceles of the appendix often mimic adnexal masses and thus may easily be misjudged. Computed tomography is mentioned as the imaging modality of choice for diagnosis and evaluation of appendiceal tumors. Thus, other imaging techniques than ultrasound may be of great value when mucocele of the appendix is suspected. A magnetic resonance tomography performed on our patient, though not for a suspected mucocele of the appendix, gave no useful additional information.

A palpable mass is found in up to 50% of appendiceal mucoceles[3] but was not evident in our case. The apparent cystic structure was a coincidental finding in the course of a routine gynecological check-up; our patient was completely asymptomatic as are about 25% of patients with mucocele of the appendix.[7,8,9] If present, pain in the right lower quadrant, although a common sign for appendiceal mucoceles, is unspecific. Other frequent symptoms are change in bowel habits and rectal bleeding.[2,8]

CONCLUSION

Appendiceal mucoceles are typically found in middle-aged patients with a higher incidence in women than in men. Thus gynecologists should routinely consider this disease in the differential diagnosis of right lower dumbbell abdominal cysts. Additionally, if a mucocele of the appendix is found during gynecological laparoscopy, a general surgeon should be involved in the procedure.

References:

1. Bartlett C, Manoharan M, Jackson A. Mucocele of the appendix - a diagnostic dilemma: a case report. *J Med Case Reports*. 2007;1:183.

2. Persaud T, Swan N, Torreggiani WC. Giant mucinous cystadenoma of the appendix. *Radiographics*. 2007;27:553-557.

3. Rampone B, Roviello F, Marrelli D, Pinto E. Giant appendiceal mucocele: report of a case and brief review. *World J Gastroenterol*. 2005;11:4761-4763.

4. Deans GT, Spence RA. Neoplastic lesions of the appendix. *Br J Surg*. 1995;82:299-306.

5. 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;32:1525-1541.

6. Matthews JB, Hodin RA. Acute abdomen and appendix. In: Greenfield LJ, Mulholland MW, Keith TO, Zelenock GB, Lillemoe KD, eds. *Surgery. Scientific Principles and Practice*. 3rd ed. Philadelphia, USA: Lippincott Williams and Wilkins. 2001;1224-1236.

7. Macek D, Jafri SZ, Madrazo BL. Ultrasound case of the day. Mucocele of the appendix. *Radiographics*. 1992;12:1247-1249.

8. Aho AJ, Heinonen R, Laurén P. Benign and malignant mucocele of the appendix. Histological types and prognosis. *Acta Chir Scand*. 1973;139:392-400.

9. González Moreno S, Shmookler BM, Sugarbaker PH. Appendiceal mucocele. Contraindication to laparoscopic appendectomy. *Surg Endosc*. 1998;12:1177-1179.