Four mucinous cystadenoma of the appendix treated by different approaches

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ABSTRACT

Appendiceal mucocele is a rare clinical entity characterized by dilatation of the appendiceal lumen due to abnormal mucinous secretion. It may be of inflammatory or tumoral origin. It may occur as a result of mucoceles, mucosal hyperplasia, mucinous cystadenoma or mucinous cystadenocarcinoma. Mucinous cystadenoma of the appendix is the most common form, although it manifests itself in many different clinical presentations. It is detected in 0.6% of appendectomy specimens. Preoperative diagnosis is difficult, and it is often detected at laparotomy. Despite existing reservations due to the risk of rupture, laparoscopic surgery is gaining acceptance. Although there is no consensus on the choice of surgery, either appendectomy or right hemicolectomy is applied. In this article, we discussed four patients with appendix mucinous cystadenoma where different surgical methods were applied in light of the literature.

Key Words: Mucocele, appendix, cystadenoma

INTRODUCTION

Appendiceal mucocele is a rare clinical condition that is characterized by cystic dilation of the appendix due to abnormal appendiceal mucinous secretion that fills the lumen (1). It may be inflammatory or tumoral in origin. Mucoceles may occur as a result of mucosal hyperplasia, mucinous cystadenoma or mucinous cystadenocarcinoma. Cystadenoma of the appendix is the most common form and manifests itself in different clinical situations. It is detected in 0.6% of appendectomy specimens (2). Preoperative diagnosis is difficult, and it is often detected at laparotomy. There is no agreement on the choice of surgery. Generally, either appendectomy or right hemicolectomy is performed. Laparoscopic techniques are more frequently applied in recent years. We present four mucinous cystadenoma cases treated by different surgical approaches.

CASE PRESENTATIONS

Case 1

Forty-four year old female patient was admitted due to abdominal pain present for one week. On physical examination, she had right lower quadrant tenderness, guarding and rebound tenderness. In addition to leukocytosis, a 86x30 mm mildly vascularized cystic lesion compatible with a mucocele was observed adjacent to the cecum on abdominal ultrasonography (Figure 1). In laparoscopy, the appendix was 8 cm in size and had a cystic structure, the adjacent cecum wall was firm, edematous and appeared as a mass. The mass that originated from the base of the appendix was submucosal and invaginated into the cecum (Figure 2). An infra-umbilical median incision was made by extending the umbilical trocar site, the right colon was mobilized, and appendectomy was performed along with a partial cecum resection (Figure 3). The distal ileum and ileocecal valve were normal. The results of the frozen section evaluation were reported as benign, thus the operation was completed. The patient was discharged on the 5th postoperative day without any problems. Definitive histopathological examination revealed an 8x3 cm appendix, with its lumen filled with mucin invaginated into the intestine, the cecum did not show signs of epithelial dysplasia. The diagnosis was reported as appendiceal mucinous cystadenoma.

Case 2

A fifty-two-year-old female patient was admitted with intermittent abdominal pain. On physical examination, she had right lower quadrant tenderness, defense and rebound tenderness, and the patient underwent surgery with an initial diagnosis of acute appendicitis. The appendix was 6 cm, with hyperemia and edema. Laparoscopic appendectomy was performed. The patient was discharged on postoperative 2nd day without any complications. The pathology evaluation revealed mucinous neoplasm with low malignancy potential. On microscopic examination, mucinous epithelium showing moderate atypia signs, without invasion, was observed. Partial-thickness mucin islands were observed in the appendiceal lumen and wall, with mucinous epithelium within these islands. Desmoplasia or invasion was not observed in either the appendix wall or serosa (Figures 4, 5).

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Received: 01.10.2013
Accepted: 20.11.2013

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Figure 1. 86x30 mm mucocoele next to the cecum

Figure 2. Submucosal mass invaginating into the cecum

Figure 3. Appendectomy along with partial cecum resection

Case 3

A sixty-six-year-old woman was admitted with complaints of intermittent abdominal pain for the last year. An 80x38 mm in size hypodense cyst with calcifications on its wall was detected on abdominal computerized tomography. The lesion was located medial to the cecum-appendix base, and extended to the right ovary, and it did not show contrast enhancement (Figure 6). At laparotomy, a cystic mass localized to the appendix was observed and appendectomy was performed. Histopathological examination showed 9x4 cm in size cystic mass and the lumen was filled with mucin. The inner surface was lined with stratified mucinous epithelium, with punctate dystrophic calcifications in the lumen.

Case 4

A 49-year-old patient was planned for surgery with diagnosis of appendiceal mass and uterine myoma. The appendix was

Figure 4. Appendix wall lined with mucinous epithelium showing proliferations into the appendix lumen. HEx20

Figure 5. Extracellular mucin ponds (arrow) showing transition to the superficial muscle layer in the appendix wall. HEx40

Figure 6. An 80x38 mm in size hypodense cyst with calcifications on its wall, located medial to the cecum-appendix base, and extending to the right ovary, without contrast enhancement

approximately 8x3 cm in size, dilated and firm. The preoperative MRI showed the appendix to be 57 mm in length, 24 mm in thickness, with dilation in the distal part. The lumen was filled with material having an intensity equal to liquids. The patient underwent an appendectomy. The pathology result was reported as mucinous cystadenoma with high-grade dysplasia. Informed consent was obtained from all four patients.

DISCUSSION

Appendiceal mucocoeles are cystic masses that are formed by dilation of the appendix due to abnormal mucinous secre-

tion. They are more frequent in women over the age of fifty years (3). Most of these tumors are clinically silent, however they may present with acute or chronic right lower quadrant pain, bowel obstruction, bleeding and intussusception. Preoperative diagnosis is difficult. Most cases are detected during surgery or after pathological evaluation. On the other hand, when malignant mucinous cystadenomas invade other abdominal organs or lead to pseudomyxoma peritonei (PP), it can cause symptoms such as weight loss, nausea and vomiting. PP is the result of rupture of the appendix, and is the most dangerous complication. Adequate preoperative assessment and careful intraoperative dissection are very important in preventing rupture and malignant transformation that may be associated (2). There are four histological subtypes of appendiceal mucocoeles: simple retention cysts are characterized by normal epithelium and often smaller than 2 cm, mucosal hyperplasia, mucinous cystadenoma that constitutes the majority of cases and mucinous cystadenocarcinomas. Misdraji et al. (4) classified mucinous appendicular tumors into two groups: low-grade appendicular mucinous neoplasms (LAMN) characterized by villous and/or flat-mucinous epithelial proliferation and a low degree of atypia, and mucinous adenocarcinoma defined by tumoral invasion of the bowel wall, complex epithelial proliferation and high-grade nuclear atypia. McDonald et al. (5) also identified two different categories of low-grade appendicular tumors: Type 1 refers to disease limited to the appendix lumen, while type 2 defines presence of mucin and neoplastic epithelium in the appendiceal wall or periautendicular tissues.

There is no agreement on the surgical treatment of appendiceal mucocoele. It is generally accepted that appendectomy is appropriate in unruptured benign mucocoeles localized to the appendix (6, 7). Zagrodnik stated local invasion and involvement of the cecum as the two indications for right hemicolectomy (8). Nevertheless, Kleemann reported that resection of the cecum is indicated in cystadenomas at the base of the appendix (9). We treated the appendiceal mucocoele in the first case with an unusual partial cecum resection. The frozen section examination revealed a benign mass, thus the cecum was primarily closed and the operation was completed. In this manner, we were able to avoid right colon resection that has a higher morbidity rate. Although right hemicolectomy is usually recommended in mucinous cystadenocarcinomas, it is expressed that appendectomy may be sufficient in early lesions limited to the mucosa or submucosa (10). It should be kept in mind that 20% of mucinous cystadenomas may be associated with synchronous or metachronous colon cancer (11). Therefore, postoperative follow-up of patients should be done carefully.

Although some studies do not recommend laparoscopic method in the surgical treatment of mucocoeles due to the risk of rupture (6, 8), Chiu reported that laparoscopic appendectomy is appropriate when performed carefully (12). In the second case, we preferred to perform laparoscopic appendectomy and close follow-up.

CONCLUSION

Detailed preoperative evaluation of appendicular masses has an important role in the selection of surgical technique, and prevention of serious complications such as pseudomyxoma peritonei. Intraoperative frozen section evaluation of patients with mucocoele, may reduce the rate of more extensive surgeries with resection in many cases. In addition, we believe that diligently performed laparoscopic appendectomy is a safe method and does not increase the complication rate.

Informed Consent: Written informed consent was obtained from patients who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - M.Ö.K., M.B.; Design - M.Ö.K.; Supervision - A.İ., M.Ö.K.; Funding - M.Ö.K.; Data Collection and/or Processing - M.Ö.K.; Analysis and/or Interpretation - M.Ö.K.; Literature Review - M.Ö.K.; Writer - M.Ö.K.; Critical Review - M.B., A.İ.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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