

Incidental Appendiceal Mucinous Neoplasms in Patients Who Underwent Right Hemicolectomy

Sağ Hemikolektomi Hastalarında İnsidental Olarak Saptanan Apendiksin Müsinöz Neoplazmları

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ABSTRACT

Objective: Subclinical appendiceal mucinous neoplasms (AMNs) may accompany abdominal malignancies which must be considered during colectomies. We aimed to evaluate the treatment and follow-up approach to the patients in whom incidentally appendiceal mucinous neoplasm detected following right hemicolectomy performed for abdominal malignancies.

Method: The records of the patients who underwent right hemicolectomy due to benign or malign conditions in our general surgery clinic between January 2016 and December 2019 were analyzed. After the exclusion of the patients who had undergone hemicolectomies due to benign causes and appendiceal pathologies detected before the operation, the records of remaining patients' were evaluated. The patient records who underwent right hemicolectomy for gastrointestinal or gynecologic malignancies were analyzed. The patients whose pathology results were reported as AMNs were included in the study. The incidence of AMN was analyzed and compared with the literature.

Results: Hundred and sixty-seven of the 214 patients with gastrointestinal or gynecologic malignancies were included in this study. Eighty patients were female and 87 were male. The mean age was 63 (23-95) years. The incidence of AMN was 2.9% in patients who underwent right hemicolectomy for malignity in three years period. According to the pathology reports; 3 patients (60%) had appendiceal mucinous neoplasia, one patient (20%) appendiceal mucinous cystadenocarcinoma and one (20%) appendiceal carcinoid tumor.

Conclusions: Incidentally detected AMN is a rare entity and should be considered during the right hemicolectomy operations of different malignancies. Resections must follow principles for malignancy surgery. The preoperative evaluation of the patients needs attention, their postoperative treatment must be planned according to the pathology reports and the patients must be followed for any potential accompanying colorectal tumors.

Keywords: appendectomy, incidental, tumor, appendiceal neoplasm

Öz

Amaç: Subklinik apendiks müsinöz neoplazmları (AMN) abdominal malignitelere eşlik edebilir ve kolektomiler sırasında göz önünde bulundurulmalıdır. Çalışmamızda, abdominal maligniteler sebebiyle sağ hemikolektomi uygulanan hastalarda insidental apendiks müsinöz patoloji saptanması halinde tedavi ve takip yaklaşımını değerlendirmeyi amaçladık.

Yöntem: Ocak 2016 - Aralık 2019 yılları arasında genel cerrahi kliniğimizde benign ya da malign nedenler sebebiyle sağ hemikolektomi uygulanan hastaların kayıtları incelendi. Benign nedenlere bağlı yapılan hemikolektomiler ve operasyon öncesi apendiks patolojisi saptanan hastalar çalışma dışına alındıktan sonra kalan olgular incelendi. Gastrointestinal veya jinekolojik malignite için sağ hemikolektomi uygulanan hasta dosyaları değerlendirildi. Patoloji sonucu AMN olarak raporlananlar çalışmaya alındı. İnsidental olarak saptanan AMN'ler değerlendirilerek literatürle karşılaştırıldı.

Bulgular: 214 hastanın gastrointestinal veya jinekolojik kaynaklı malignite sebebi ile sağ hemikolektomi uygulanan 167'si çalışmaya alındı. Hastaların 80'i kadın, 87'si erkek idi. Yaş ortalaması 63 (23-95) idi. Kliniğimizde üç yıllık sürede malignite sebebiyle sağ hemikolektomi operasyonu geçiren hastalarda AMN insidansı %2.9 idi. Patoloji raporlanmasında 3 hastada (%60) musinoz neoplazi, 1 hastada (%20) musinoz kistadeno kanser saptandı. 1 hastada ise (%20) apendiks karsinoid tümörüyle karşılaşıldı.

Sonuç: AMN nadir görülen bir antitedir ve malignite nedeni sağ hemikolektomilerde akılda olmalıdır. Yapılacak rezeksiyonlar malignite cerrahisi sonuçları gözetilerek gerçekleştirilmelidir. Hastaların preoperatif değerlendirmeleri dikkatle yapılmalı, tedavileri postoperatif patoloji sonuçlarına göre yönlendirilmeli ve hastalar gelişebilecek kolorektal tümörler açısından takip edilmelidir.

Anahtar kelimeler: appendektomi, insidental, tümör, apendiks neoplazmı

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INTRODUCTION

Mucocele is a rare appendiceal pathology (0.2-0.7% of all appendectomy specimens) ⁽¹⁻³⁾. Appendiceal mucinous neoplasms (AMNs) are very rare with 1000 to 2000 cases diagnosed annually in the United States ⁽⁴⁾ and they are responsible about 1% of all cancers worldwide ⁽⁵⁾. AMNs include a heterogeneous group of diseases and their malignancy potential varies as seen in different classification systems ⁽⁶⁻⁸⁾. Different classifications with similar terminology were suggested to define lesions of variable biologic potential by several authors. Not only classification but also management of AMNs are also problematic for clinicians ^(7,8). A remarkable part of the lesions is asymptomatic, and they are incidental findings at surgeries AMNs are usually diagnosed incidentally during surgery performed for suspect cases of appendicitis ⁽⁹⁾. Advanced stage of AMN is observed with abdominal distension related to the accumulation of mucin in the peritoneal cavity. These tumors may disseminate throughout the peritoneal cavity in the form of gelatinous deposits; the worst complication of AMNs called peritoneal pseudomyxoma, caused by spontaneous or iatrogenic appendix perforation with high rate of morbidity and mortality ^(10,11). Also, there are no clear guidelines on the extent of surgical resection; therefore, many reports on surgical procedures have been published ^(3,12). Considering uncertain potential malignant progression, an early and accurate preoperative identification of AMNs confined to the appendix is crucial for prediction of prognosis and decision on treatment strategy ⁽¹³⁾.

In our study we aimed to evaluate the treatment and follow-up approach to the patients who had incidentally detected appendiceal mucinous pathology following right hemicolectomy performed for abdominal malignancies.

MATERIAL and METHODS

Patients, inclusion and exclusion criteria:

We have analyzed the records of the patients who underwent right hemicolectomy performed due to benign or malign conditions in our general surgery clinic between January 2016 and December 2019. The records were analyzed for demographics, sur-

gery reports, pathology results and long-term outcomes.

Patients with a history of a previous operation for benign conditions, such as acute appendicitis or with a radiographically or pathologically confirmed appendicitis before the primary surgery were excluded from the study. After exclusion of patients who had undergone hemicolectomies due to benign causes and appendiceal pathologies detected before the operation, records of remaining patients were evaluated.

We have identified the cases matching the inclusion criteria that was pre-described as malignancy. Physical examination notes, operation, and the pathology records as well as clinical data of all included patients were carefully reviewed.

The patients with a final pathological diagnosis of appendiceal mucinous neoplasm were also included in the study. The incidence of appendiceal mucinous pathology was analyzed and compared with the literature. The AMN cases were re-examined and evaluated based on pathology reports recorded during the study.

Pathologic evaluation:

Appendiceal mucinous pathology (intracytoplasmic mucine containing nuclear atypia) was identified in histological specimens (Figure 1).

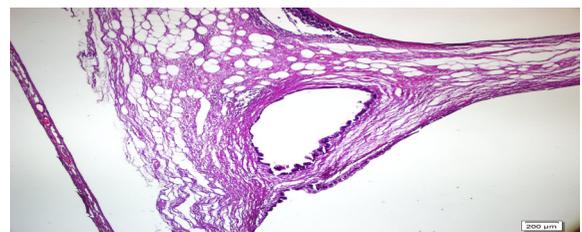


Figure 1. Neoplastic columnar cells with mild intracytoplasmic mucine containing nuclear atypia (HE X40, X200).

Ethics committee approval was obtained. All patients were evaluated in follow-up visits after surgery.

RESULTS

There were 214 patients who underwent right hemicolectomy due to benign or malign conditions in our general surgery clinic in three years period. A total of

167 (78%) patients were included in the study who had medical history of a gastrointestinal or gynecologic malignancy and underwent primary surgery.

A total of 167 cases who underwent right hemicolectomy due to malignancy; consisted of 80 (47.9%) female and 87 (52.1%) male patients. The mean age of the patients was 63 (23-95) years.

In the present study, 5 of the 167 cases evaluated were diagnosed with appendiceal mucinous neoplasms by histomorphological and architectural findings and two of them had lesions of >2 cm in diameter. The study population consisted of four women and one man. The median age of these patients was 52 (23-63) years. The incidence of appendiceal mucinous neoplasm was 2.9% in patients who underwent right hemicolectomy for gastrointestinal or gynecologic malignancies from January 2016 to December 2019 in surgery department (Figure 2).



Figure 2. Distribution of appendiceal mucinous pathology among patients who underwent right hemicolectomy.

Three out of five cases (60%) had been diagnosed with low-grade appendiceal mucinous neoplasms according to pathological reports of specimens during the study. One out of five patients (20%) was diagnosed with mucinous cystadenocarcinoma and one out of five patients (20%) was diagnosed with an appendiceal carcinoid tumor.

DISCUSSION

Primary neoplasms of appendix are rarely seen (less than 2% of surgical appendectomy specimens) and their major categories include epithelial tumors, mesenchymal tumors, and lymphomas. AMNs is a complex, diverse group of epithelial neoplasms often causing cystic dilation of the appendix due to the

accumulation of gelatinous material, morphologically referred to as mucoceles⁽¹⁴⁾.

AMN is an infrequently seen adenoma which can either be in the appendix or the surrounding appendiceal mucosa wall. Mucoceles are also very rare seen (0.2% and 0.7%) among all appendectomy specimens. They affect women 4 times more than men. Appendiceal mucinous lesions (both benign and neoplastic) have also a slight female predominance^(15,16). In our study, 80% of the cases were female. AMNs are usually diagnosed in patients in their 50s and 60s^(15,16). In our case series the median age of these patients was 52 years.

Patients with AMN can present with intussusception, abdominal pain and obstruction. However, these neoplasms are often found incidentally in asymptomatic patients. AMNs are frequently misdiagnosed as acute appendicitis, retroperitoneal tumors of the right iliac fossa, or an adnexal mass of⁽¹⁷⁾. Ultrasound and CT (the most commonly used radiographic interpretation for preoperative diagnosis) are the main imaging techniques in terms of diagnosis^(13,18). Cystic dilation of appendiceal lumen, calcifications of appendiceal wall and irregular appendiceal wall thickening are common abdominal CT findings.

In pathological examination; appendiceal wall hyalinization and fibrosis with a grossly swollen appendix secondary to mucinous accumulation⁽¹⁹⁾ can be seen. Histological characteristics of AMNs usually present as atypical glandular cells and epithelial cells with “pushing invasion” of malignant cells creeping into the appendiceal wall with possible diverticular formation. Mucinous cells, goblet cells among colon cells are frequently identified within AMNs⁽²⁰⁾. AMNs of <2 cm diameter are rarely malignant and usually classified as benign simple or retention mucoceles. AMNs of >6 cm represent high risk of malignancy, appendiceal perforation, and development of pseudomyxoma peritonei⁽²¹⁾. Two of our cases had lesions of >2 cm in diameter. One of them was diagnosed with mucinous cystadenocarcinoma while the other patient was diagnosed with appendiceal carcinoid tumor.

Controversies remain as to the best surgical approach (laparoscopic vs open), adjuvant therapy, duration of

follow-up, and imaging technique. The goal of the AMN management includes the prevention from rupture, seeding, and pseudomyxoma peritonei⁽²²⁾. Right hemicolectomy in the absence of lymph node metastasis has been replaced by appendectomy which is the only approach used for the treatment of benign appendiceal mucocoeles. Upon the discovery of submucosal malignancy, infiltration or lymph node metastasis, right hemicolectomy with or without omentectomy may be performed.

In our study, incidentally discovered appendiceal mucinous pathologies were evaluated in right hemicolectomy operations of the patients who underwent surgery for gastrointestinal or gynecologic malignancies. Incidentally detected appendiceal mucinous pathology is a rare entity.

AMNs can mimick appendicitis and be diagnosed pathologically in less than 2% of surgical appendectomy specimens⁽¹⁴⁾. The incidence of AMN in colon resection operations is not well defined. In our study, the AMN incidence in right hemicolectomy malignancies was 2.9 percent.

AMNs of > 2 cm are considered to have malignancy potential and right hemicolectomy is the recommended surgical treatment in such cases⁽²³⁾. Any appendiceal abnormality was not detected before surgery in our patients. We identified 5 AMN cases for whom we performed right hemicolectomy for gastrointestinal or gynecologic malignancies in our records.

Pathology reports revealed appendiceal mucinous pathologies in these patients. Thus, any further surgical and adjuvant therapies were not required in these cases.

CONCLUSIONS

Overall, further studies are needed to identify the best diagnostic method and treatment approach for appendiceal mucinous pathology. There remains a lack of standardization for post-treatment surveillance. AMNs should be considered during the colectomy operations of different malignancies and resections must follow principles for malignancy surgery. We recommend exploring the appendix for any

potential AMNs while performing colectomies due to any reason.

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Informed Consent: Not obtained since the study is retrospective.

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