Mucinous cystadenoma of the appendix: a rare cause of acute abdomen

Nadir bir akut karın tablosu nedeni: Apendiks müsinöz kistadenomu

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BACKGROUND
We aimed to assess the acute abdominal conditions due to appendiceal mucinous cystadenomas.

METHODS
We retrospectively evaluated 11 patients with histopathologically confirmed appendiceal mucinous cystadenoma. Patient charts and data on patient demographics; clinical features; ultrasonography (US), colonoscopy and computed tomography (CT) findings; pathology reports; and operative and postoperative management were reviewed.

RESULTS
The incidence of appendiceal mucinous cystadenoma was 0.95% of all appendectomy specimens reviewed. In our review, there were 11 patients, five of whom were women. The median age was 70 years (50-85 years), and the most common presentation was abdominal pain (81.8%). On US in eight patients, findings were abdominal cystic mass and cyst wall calcification. The CT finding was well-encapsulated cystic mass in eight patients. In one case, a colonic mass was found in colonoscopic examinations. There was one patient with concomitant colon cancer. Appendectomy was performed in nine patients and right hemicolectomy was performed in two patients.

CONCLUSION
Colonoscopy, US, and CT are useful tools in diagnosing mucocele and synchronous cancer. However, diagnosis is usually made intraoperatively or postoperatively on histopathological examination. Appendectomy is the standard of care for mucinous cystadenoma. Furthermore, it is important to prevent spillage of the mucocele content.

Key Words: Acute abdomen; appendix; mucocele; mucinous cystadenoma.

AMAÇ
Akut karın bulguları oluşturan, apendiksin müsinöz kistadenomları araştırıldı.

GEREÇ VE YÖNTEM
Histopatolojik olarak apendiksin müsinöz kistadenomu tanısı alan 11 hasta geriye dönük olarak değerlendirildi. Hastaların, dosya taramalarından elde edilen, demografik veriler, klinik özellikler, ultrasonografi (USG), kolonoskopi ve bilgisayarlı tomografi (BT) bulguları, patoloji raporları, ameliyat öncesi, ameliyat ve sonrası bulguları incelendi.

BULGULAR
Patolojik incelemesi yapılan apendektomi parçalarında müsinöz kistadenom görülme sıklığı %0,95 olarak saptandı. Toplam 11 olgunun beşi kadındı. Ortalama yaş 70 (50-85) idi. Olgularda en sık görülen klinik bulgu ise karın ağrısıydı (%81,8). Karın USG'si yapılan sekiz olguda kistik kitle ve kist duvar kalsifikasyonları izlendi. BT incelemelerinde sekiz olguda düzgün kenarlı kistik kitle ve kist duvar kalsifikasyonu izlendi. Kolonoskopik inceleme yapılan bir olguda ise kolonda kitle izlendi. Diğer bir olguda ise eşlik eden kolon kanseri saptandı. Olguların dokuzuna apandektomi, ikisine sağ hemikolektomi ameliyatları yapıldı.

SONUÇ
Kolonoskopi, USG ve BT mukosel tanısı ve yandaş kanser olgularının araştırılmasında faydalı yöntemler olmasına rağmen kesin tanı, ameliyat bulguları ve ameliyat sonrası yapılan patolojik incelemelerle belirlenir. Apendiksin müsinöz kistadenomlarının tedavisi apandektomidir, ancak ameliyat esnasında mukosel içeriğinin batın içine dökülmemesine özen gösterilmelidir.

Anahtar Sözcükler: Akut karın; apendiks; mukosel; müsinöz kistadenom.

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Mucinous neoplasm of the appendix, an uncommon disease, is found in 0.3% of all appendectomy specimens.[1] It consists of a cystic mass filled with mucin (mucocele). The mucinous accumulation arises from the obstruction of the appendiceal lumen, and this may result from benign conditions such as hyperplastic growth or develop from malignant processes.[2] Histopathological lesions are classified as mucosal hyperplasia, mucinous cystadenoma, or mucinous cystadenocarcinoma.[3]

Patients may present with various clinical signs and symptoms. Preoperative colonoscopy, ultrasonography (US), and computed tomography (CT) are useful methods in diagnosing mucocele and distinguishing the mucocele from mimicking diseases.[4,5] However, the diagnosis is usually made intraoperatively or postoperatively on histopathological examination.[6] The aim of this review was to assess the clinical presentation, diagnosis, and planning treatment of appendiceal mucinous cystadenomas.

MATERIALS AND METHODS

Between January 2000 and January 2005, 1156 appendectomies were performed in Ankara and Adana hospitals. We retrospectively reviewed the records of 11 patients with histopathologically confirmed appendiceal mucinous cystadenoma. Recorded data included patient demographics, clinical features, US, colonoscopy and CT findings, concomitant diseases, and conditions for which surgery was indicated. Pathology reports, operative and postoperative management, and information on last follow-up were also recorded. In this study, the descriptive variables of standard deviation and median were used.

RESULTS

A total of 1156 appendectomies were performed. Two hundred fifty-three patients were older than 50 years and 212 of these older patients were operated with a presumptive diagnosis of acute appendicitis.

We reviewed 11 patients (0.95%). The median age was 70 years (50-85 years). Histopathological diagnosis of all patients was appendiceal mucinous cystadenoma (Fig. 1).

Patients' clinical presentations included abdominal pain (n: 9), nausea/vomiting (n: 3), abdominal distention (n: 2), and weight loss (n: 1). Two patients were asymptomatic (18.2%). During the physical examination of 1 patient, a palpable mass localized at the pain site was determined. Both US and CT were performed in 8 patients, and appendiceal mucinous cystadenoma was detected as an abdominal cystic mass with wall calcification situated in the right lower quadrant. CT examinations revealed a well-encapsulated cystic mass with thick (n: 4), or thin (n: 4) wall in the pericecal area (Fig. 2). On postcontrast scan, cystic mass was presented as wall calcification in four patients (Fig. 3). Two patients were examined by colonoscopy and a colonic mass was found in 1.

Eight patients underwent surgery because of acute abdominal conditions (acute appendicitis 6 cases, intestinal obstruction 2 patients). In 3 patients, surgery was indicated for other conditions (colon cancer, pelvic and liver hydatid disease, and right adnexal mass) (Table 1).

Nine patients underwent appendectomy, and a right hemicolectomy was performed in 2 patients. In all patients, histopathological examination of specimens found mucinous cystadenoma of the appendix.

One patient died of septicemia in the third postoperative week. The other 10 patients recovered uneventfully with a median hospital stay of 5.8 days (range: 3-15 days). The 10 surviving patients had no evidence of disease at their last follow-up (median: 23.7 months; range: 10-48 months).

DISCUSSION

The incidence of mucocele ranges from 0.2% to 0.3% of all appendectomy specimens. Higa et al.[3] reported that mucinous cystadenomas of the appendix make up 63% of all mucinous lesions. However, we found 11 mucocele in 1156 appendectomy specimens (0.95%), and all of them were mucinous cystadenoma. There was no simple mucocele or mucinous cystadenocarcinoma.

Mucocele of the appendix is more frequent in women and is usually observed in patients older than 50 years.[7] In contrast with the literature, the group we reviewed included 5 women and 6 men; however, all patients were older than 50 years. The typical US finding is a cystic mass with variable internal echogenicity, layered wall, and calcification in the wall; we found this in 8 patients. The CT

Table 1. Clinical presentation, diagnosis and treatment in patients with appendiceal mucinous cystadenoma

No	Age/ Sex	Clinical presentation	Suspected diagnosis	Surgery	Accompanying disease	Follow-up (months)
1	71/M	Asymptomatic	Right colon carcinoma	Right hemicolectomy	Adenocarcinoma of the right colon	30
2	49/F	RLQ pain	AA	Appendectomy	AA	14
3	52/F	RLQ pain	Right adnexal mass	Appendectomy + TAH-BSO	–	42
4	85/M	Abdominal distention	Intestinal obstruction	SB resection + appendectomy	SB necrosis	13
5	66/F	Asymptomatic	Liver and pelvic hydatid disease	Partial cystectomy-omentoplasty + appendectomy	Liver hydatid disease	12
6	82/M	Abdominal distention	Intestinal obstruction	SC resection + appendectomy	Ischemic colitis	Exitus
7	72/M	RLQ pain	AA	Appendectomy	AA	12
8	70/F	RLQ pain	AA	Appendectomy	AA	40
9	54/F	RLQ pain	AA	Appendectomy	AA	10
10	56/M	RLQ pain	AA	Right hemicolectomy	AA	20
11	70/M	RLQ pain	AA	Appendectomy	AA	36

M: Male; F: Female; RLQ: Right lower quadrant; AA: Acute appendicitis; SB: Small bowel; SC: Sigmoid colon; TAH: Total abdominal hysterectomy; BSO: Bilateral salpingo-oophorectomy.

findings were reported to consist of a well-encapsulated cystic mass with a wall of variable thickness.[4,5] Our patients' radiologic findings were similar to those reported in the literature.

Ultrasonography and CT are useful tools in diagnosing other pathologic processes and concomitant diseases. In our review, 2 patients were asymptomatic for mucocele; 1 had symptoms for colon cancer and the other for liver hydatid cyst. Appendiceal mucinous cystadenoma was detected during radiologic workup.

Colonoscopy is usually nondiagnostic, as

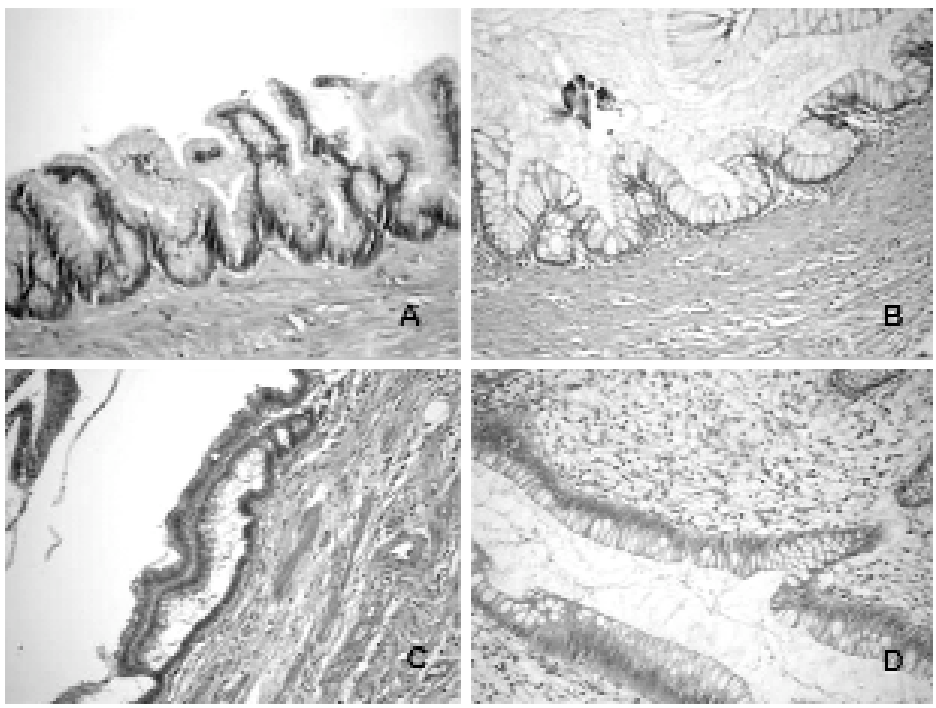

Fig. 1. (a, b) Proliferative and papillary configuration in mucinous cystadenoma of appendix (H-E x 200). **(c)** Mucinous epithelium showing undulating pattern (H-E x 200). **(d)** Mucinous cystadenoma is lined by single layer of mucinous cells (H-E x 400).

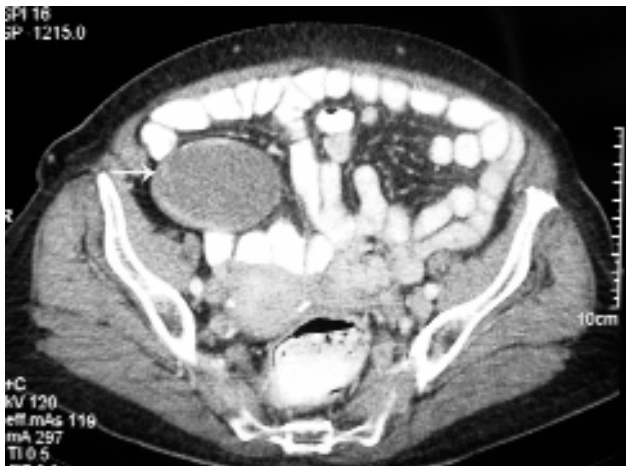

Fig. 2. CT scan shows oval shaped cystic mass (arrow) in the pericecal area.

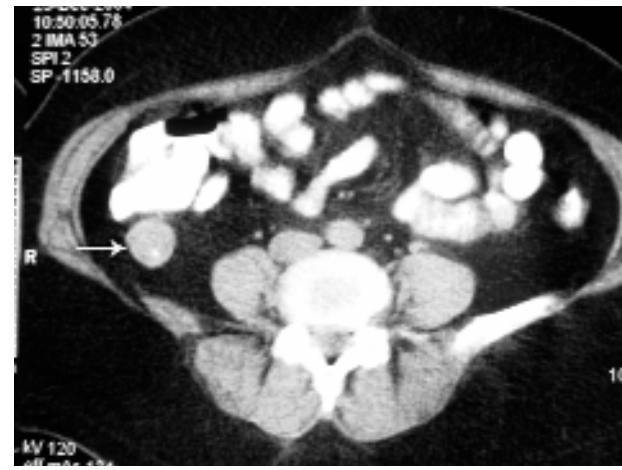

Fig. 3. Precontrast CT scan shows round cystic mass (arrow), with a punctuate calcification.

mucosal biopsies will often be normal. However, extrinsic compression or mass protrusion of the appendiceal orifice can be helpful.[3] Two patients diagnosed with intra-abdominal mass through radiological methods underwent colonoscopy. Right colon adenocarcinoma was detected in 1 patient, and extrinsic compression in the other.

Mucinous cystadenomas of the appendix can cause acute abdominal conditions, particularly in elderly patients.[1] Complications of mucocele include intussusception, intestinal bleeding, urethral or intestinal obstruction, hematuria, and rupture resulting in pseudomyxoma peritonei.[8-11] In our cases, 6 patients had surgery for acute appendicitis. One patient with intestinal obstruction was found to have gangrenous small bowel loops caused by strangulation by a mucinous cystadenoma of the appendix. Mucinous cystadenomas of the appendix were associated with colon adenocarcinomas in 20% of cases.[3,12] In our review, 1 patient had concomitant colon cancer (9.2%).

Appendectomy is the standard of care for mucinous cystadenoma, while cystadenocarcinoma requires a right hemicolectomy. It is important to prevent rupture and spillage of its contents during surgery; otherwise, pseudomyxoma peritonei may result.[3,6] In our group, there was no mucocele perforation.

Because of the high association of mucinous cystadenoma with colon and ovarian malignancy, follow-up CT, US and colonoscopy examinations must be performed during the postoperative period. During the follow-up period, there was no evidence of disease in our group.

In conclusion, mucocele of the appendix is a rare condition and may cause acute abdominal conditions; mucinous cystadenoma is the most common histopathologic type. It is more frequent in elderly patients and may cause acute appendicitis (2.8%). US, CT and sometimes colonoscopy are useful tools in diagnosing mucocele and concomitant cancer. However, actual diagnosis is usually made intraoperatively or during histopathologic examination of the excised specimen. Inadvertent spillage of the mucocele content is the most troublesome complication. Finally, it should be kept in mind that these lesions may coexist with other neoplasms.

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