

The Appendix Mucocele in the Distinctive Diagnosis of Adnexial Mass: Case Report

ADNEKSİYAL KİTLE AYIRICI TANISINDA APPENDİKS MUKOSELİ

Özgür AKBAYIR, MD,^a Ceyhun NUMANOĞLU, MD,^a Kemal GÜNGÖRDÜK, MD,^a
Engin ODABAŞ, MD,^a Ahmet GÜLKILIK, MD^a

^aClinic of Gynecological Oncology, İstanbul Bakırköy Hospital of Women and Children, İSTANBUL

Abstract

Appendiceal mucoceles are uncommon cystic neoplasms characterized by distension of the appendiceal lumen with mucus. Most of them are caused by mucinous cystadenomas and rarely cystadenocarcinomas.

A possible rupture of the mucocele, either spontaneous or accidental, during surgery may result in the clinical condition of pseudomyxoma peritonei, a spread of mucin producing cells throughout the entire peritoneal cavity in the form of multiple mucin deposit. Therefore preoperative diagnosis and careful resection to remove mucocele of the appendix is important in preventing pseudomyxoma peritonei and predicting malignant transformation.

In this case, we present a patient who has been investigated for the etiology of adnexial mass and was diagnosed to appendix mucocele.

We report one case and present the clinical and diagnostic aspects of this disease.

Key Words: Mucocele; appendix; adnexial mass

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Özet

Appendiks mukoseli, appendiks lümeninin anormal mukus birikimi nedeniyle genişleyip kistik bir kitle haline gelmesi sonucu oluşur. Genellikle müsinöz kistadenom ve daha nadiren de kistadenokarsinom sonucu oluşur.

Preoperatif spontan ya da operasyon sırasında iatrojenik rüptürü, müsinöz materyalin batına yayılmasıyla oluşan psödomiksoma peritonei'ye sebep olabileceği gibi, batin içinde müsinöz depositler de neden olabilir. Preoperatif ön tanısı operasyon sırasında kitlenin bütünlüğünün bozulmadan çıkarılması açısından önemlidir. Böylece olası bir psödomiksoma peritonei ve malign transformasyon tablosunun önüne geçilmiş olur.

Burada, kliniğimize adneksial kitle etiyolojisi açısından araştırılan ve sonuçta appendiks mukoseli tanısı konan bir olgu sunulmuştur.

Anahtar Kelimeler: Mukosel; appendiks; adneksiyal kitle

Mucocele of the appendix means an enlarged appendix with obstructive luminal dilatation by mucus.¹ It is the sequela of several kinds of benign and malignant neoplasms. In the latter, pseudomyxoma peritonei is the worst complication. Symptoms and signs are frequently nonspecific. Therefore, proper diagnosis and avoidance of iatrogenic rupture during surgery are of paramount importance.

Case Report

A 56-year-old was referred to our hospital for pain located in the right flank and the right lower quadrant. This pain was intermittent for eight years. Her prior medical history was significant for diabetes and hypertension. Her prior surgical history included tubal ligation operation in 1981 and cholecystectomy operation in 1989.

The physical examination, her abdomen was flat, soft, and with normoactive bowel sounds. Tenderness was noted in the right upper and lower abdomen without rebound pain or muscle guarding. On pelvic examination, hard, mobile mass approximately 8x6 cm was palpated in the right adnexial area. Transvaginal ultrasonography revealed 51 x 30 x 86.1 mm heterogenic lesion in the right adnexial

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Yazışma Adresi/Correspondence: Kemal GÜNGÖRDÜK, MD
İstanbul Bakırköy Hospital of Women and Children,
Clinic of Gynecological Oncology, İSTANBUL
maidenkemal@yahoo.com

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area had a capsule thickness less than 2 mm and had no septation. The inferior of the mass was echoing in thin planes that had the view of onion membranes (Figure 1). There was no free fluid in douglas cavity, left ovary and uterus were viewed as atrophic. Endometrial thickness was less than 5 mm.

Tumor markers were in normal ranges; CA 125: 10.11 U/ml , Ca 19-9: 7.79 U/ml, Beta Hcg: 2.31 mIU/ml, AFP: 3.4 IU/ml.

Laparotomy was planned with the diagnosis of pelvik mass. Abdomen was entered under general anesthesia with median incision under umblicus. Uterus and ovaries were atrophic. A soft cystic mass with a diameter of approximately 80 x 50 mm with a smooth surface origminating from the appendix in the pelvic region was identified (Figure 2). Abdominal irrigation was performed. The abdominal viscera was evaluated as normal in exploration. The mass was extirpated thick, highly viscous mucous like material was identified in macroscopic examination (Figure 3). The sample was sent to frozen examination, after the frozen examination resulted as benign appendix mucocel, bleeding control was performed following abdominal cleaning and the abdominal layers were closed regarding to the anatomy.

Discussion

Mucocele of the appendix is a relatively uncommon pathology with a reported incidence of



Figure 1. The ultrasonographic view of appendix mucocel.

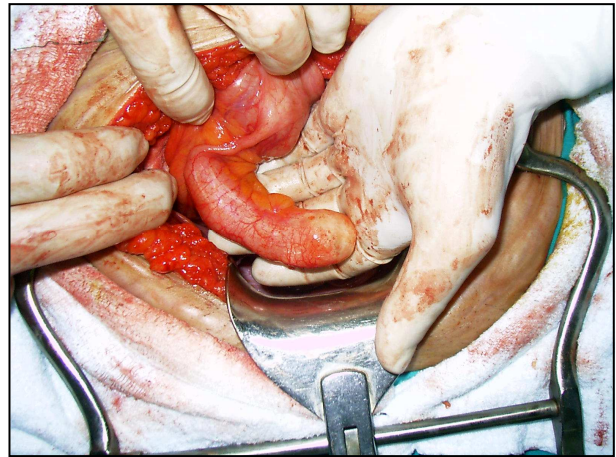


Figure 2. The view of appendix during the operation.

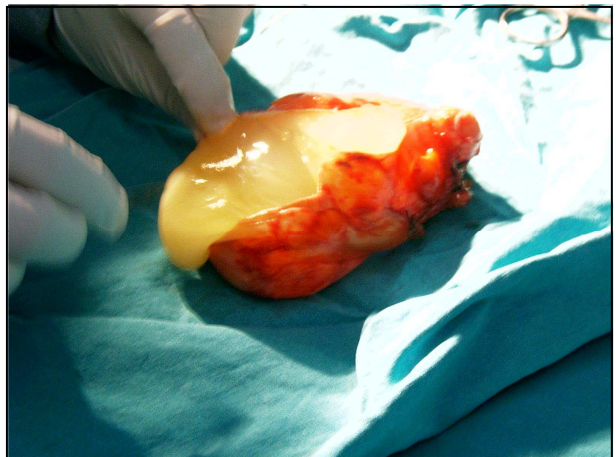


Figure 3. The resected view of appendix during the operation.

0.2–0.3% in all appendectomy specimens² and is often not considered when problems of the right lower quadrant of the abdomen present to the gynaecologist.

Appendiceal mucocele is not a specific diagnosis, but rather, is a descriptive term for dilatation of the lumen of the vermiform appendix by an abnormal accumulation of mucous. Two major pathological mechanisms are thought to be responsible for the formation of appendiceal mucocele; first elevated appendiceal pressures as sequelae of luminal obstruction caused by prior inflammation, mucosal hyperplasia, or appendiceal lesions (e.g., fecaliths, endometriosis, diverticulae, polyps) and second tumors of the appendix (i.e., carcinoid, cystadenoma, cystadenocarcinoma).³

Histopathologic classification of appendiceal mucoceles is dependent on the characteristics of their lining epithelium. These include retention cysts (18%), mucoceles with mucosal hyperplasia (20%), mucinous cystadenomas (32%), and mucinous cystadenocarcinomas (10%). Classification is important, because the course of the disease and prognosis are related to these subtypes.⁴ Simple mucoceles (also referred to as inflammatory or obstructive mucoceles, or as simple or retention cysts) are characterized by degenerative epithelial changes and may result from appendiceal obstruction and distension. There is no evidence of hyperplasia or neoplasia of the mucosa.⁵ Hyperplastic mucoceles are sessile or pedunculated lesions that represent hyperplastic polyps of the colon and are not known to have any malignant potential. Mucinous cystadenomas also have been referred to as low-grade appendiceal mucinous neoplasms. They typically are circumferential cystic lesions composed of mucinrich epithelium, which may form villous structures. The nuclei show at least focal stratification and crowding consistent with dysplastic epithelium, generally low grade. While mucinous cystadenomas can be considered the equivalent of adenomatous colon polyps, rupture and the spread of mucin and/or dysplastic epithelium into the abdominal cavity can occur. Deaths have been reported secondary to advanced disease/pseudomyxoma peritonei in these patients.^{5,6} Mucinous cystadenocarcinomas (also mucinous adenocarcinomas) demonstrate high-grade cellular dysplasia and, often, stromal invasion beyond the muscularis mucosae. Given the poorly defined criteria for invasion in mucinous neoplasms, a further category of lesions, "mucinous tumors of uncertain malignant potential" has been used by some investigators, reflecting the difficulty in classifying some lesions as clearly benign or malignant with respect to their clinical behavior.^{5,6}

Appendiceal mucoceles reportedly show a female predominance of four to one. The average age at the time of diagnosis is 54 years for benign mucoceles and 64 years for malignant disease.⁷

Appendiceal mucocele is not clinically suspected in 50% of cases where it is diagnosed, and

25% of appendiceal mucocele patients are asymptomatic at the time of diagnosis.⁸ The most common presentation of symptomatic appendiceal mucocele patients is acute or chronic right lower quadrant abdominal pain, as occurred with our patient.⁹ Cyclical or colicky pain can occur when appendiceal mucocele is associated with intussusception or endometriosis.^{9,10} An intra-abdominal mass is palpated by the examining physician in half of cases and is also occasionally palpated by the patient.⁸ Nausea and vomiting, as well as altered bowel habits (e.g., diarrhea, constipation) are often reported, and evidence of gastrointestinal bleeding is noted if intussusception is present.⁴⁻¹¹

An association between appendiceal mucocele and synchronous colon neoplasms has been previously noted. The most common synchronous neoplasms occur in the large bowel (19.5%-21%), although they can also be found in other locations, such as the ovary (2-24%) gallbladder, breast, kidney and thyroid.¹²

A correct pre-operative diagnosis of appendiceal mucocele is difficult due to the nonspecific symptoms, but it is of great importance. Emphasis should be placed to avoid tumor rupture during surgery and formation of pseudomyxoma peritonei in case malignancy exists.

We should differentiate other diseases from appendiceal mucocele. Benign lesions of the appendix include appendicitis, mucinous cystadenoma of the appendix, and epithelial hyperplasia of the appendix. Others include Meckel's diverticulum, colitis cystitica profunda, colonic diverticulum with abscess formation, hydrosalpinx, ovarian cyst, mesenteric cyst, and enteric duplication cyst.¹ Differential diagnosis of malignant lesions of the right lower quadrant should include appendiceal cystadenocarcinoma, ruptured colonic mucinous adenocarcinoma, and mucinous tumors of the ovary with pseudomyxoma peritonei.¹³

Computed tomography (CT), ultrasonography (US), barium enema and colonoscopy have all been used to describe these tumours. Unfortunately none of these is entirely conclusive.⁷

The reported findings of mucocele of the appendix at US include a purely cystic mass with anechoic fluid, hypoechoic mass with variable internal echogenicity according to internal content (watery or thick gelatinous), a thin inner echogenic rim and outer echolucent layer of the wall representing bowel wall, curvilinear or punctate wall calcification due to dystrophic response to a chronic inflammatory process, intussusception, and pseudomyxoma peritonei caused by rupture of mucocele.¹⁴ In some patients, multiple echogenic layers along the dilated appendix produce the appearance of “onion skin-like” circles and may be pathognomonic for mucocele.¹⁵ The US findings in our patient correlate well with these previously reported findings.

The typical CT aspect of appendiceal mucocele is a cystic, well-encapsulated mass, sometimes with mural calcifications in expected location of the appendix and causing extrinsic pressure on the cecal wall without any sign of surrounding inflammatory reaction.¹⁴

Fine needle aspiration of the appendiceal mucocele is generally avoided by most surgeons because of fear of rupture or seeding of neoplastic cells, which may lead to localized or diffuse pseudomyxoma peritonei.⁷

Therapy is surgical. However, the laparoscopic approach should be avoided because of the increased risk of rupture and subsequent pseudomyxoma peritonei. Simple appendectomy is curative of non-neoplastic appendiceal mucocele and simple cystadenomas, whereas more extensive cystadenomas and cystadenocarcinomas require more extensive surgery (e.g., cecum resection, right hemicolectomy). At the time of surgery, the abdomen and pelvis are evaluated for the presence of any associated pathology (e.g., ovarian, colonic, etc.).¹⁶

The 5-year survival rate for simple or benign neoplastic mucocele ranges from 91% to 100%, but is 25% in mucinous cystadenocarcinomas. The 10-year survival rate for mucinous cystadenocarcinoma is 65% among patients treated with hemi-

colectomy, but only 37% among patients who undergo an appendectomy alone.¹⁻⁷⁻¹⁶

In conclusion, mucocele of the appendix is seen rare. It is very difficult to diagnose preoperatively. Preoperative diagnosis of the appendix mucocele is important for excising out the whole tumor without interfering with its integrity. Preoperative spontaneous or iatrogenic rupture may cause pseudomyxoma peritonei by spreading of the mucinous material into the abdominal cavity. To be able to prevent this clinically malignant condition, preoperative diagnosis is very important. The mucocele of the appendix must be considered in patients with advanced age; especially female gender, with atypical ultrasonografi appearance; or adnexial mass at the right side.

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