A case of giant appendiceal mucocele

Dev bir appendisial mukosel olgusu

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Appendiceal mucocele is a rare clinical condition that causes distension of the appendix lumen with mucus. A seventy-three-year-old female patient presented with complaints of abdominal pain, nausea, and vomiting. Abdominal examination revealed mild tenderness, right lower quadrant pain upon palpation, rebound tenderness and muscular rigidity, and a palpable mass. Abdominal ultrasonography and computed tomography scans demonstrated a cystic lesion in the right iliac fossa, adherent to the cecum, suggesting an abdominal abscess. An emergency operation was performed, during which a diagnosis of a mucocele of the appendix was made. Surgical treatment included appendicectomy, partial resection of the ileum, and resection of the cecum. Histopathologic examination confirmed the operative diagnosis. The role of imaging and clinical approach is emphasized in the treatment of an appendiceal mucocele, especially in emergency settings.

Key Words: Adenocarcinoma; appendiceal neoplasms; appendix; cystadenoma; intestinal obstruction; mucocele; tomography, X-ray computed.

Appendisial mukosel, apandiks lümeninin mukus ile distansiyonu sonucu oluşan nadir bir klinik durumdur. Yetmiş üç yaşında kadın hasta karın ağrısı, bulantı ve kusma şikayetleriyle kliniğimize başvurdu. Karın muayenesinde orta derecede hassasiyet, sağ alt kadranda palpasyonla ağrı, rebound hassasiyet, kas rijiditesi ve palpabl kitle saptandı. Abdominal ultrasonografi ve bilgisayarlı tomografi incelemelerinde sağ iliak fossada, çekuma yapışık, abdominal apse düşündüren kistik bir lezyon görüldü. Acil ameliyata alınan hastaya ameliyat sırasında apandiks mukoseli tanısı kondu. Hastaya cerrahi tedavi olarak apandektomi, parsiyel ileum rezeksiyonu ve çekum rezeksiyonu yapıldı. Histopatolojik inceleme ameliyat sırasında konan tanıyı doğruladı. Bu yazıda, özellikle acil koşullarında, appendisial mukosel tedavisinde görüntüleme yöntemlerinin rolü ve klinik yaklaşımın önemi vurgulandı.

Anahtar Sözcükler: Adenokarsinom; apandiks neoplazmları; apandiks; kistadenom; intestinal tıkanıklık; mukosel; bilgisayarlı tomografi.

Appendiceal mucocele is a rare clinical condition that causes the lumen of the appendix distended with mucus.[1,2] Twenty-five per cent of the cases is asymptomatic and incidentally discovered either during surgery or on radiologic examination.[2,3] It has a higher frequency in females (M/F: 1/4) and at ages beyond fifty years. Ultrasonography (US) or computed tomography (CT) may reveal a mucocele as a cystic mass and suggest the diagnosis.[1,3,4] In the presence of a mucocele, the appendix does not fill with contrast medium. A mucocele with abnormal mucus accumulation in the appendiceal lumen may pose severe problems; an accidental or iatrogenic rupture of the mass may result in pseudomyxoma peritonei.[1,2,4] To avoid surgery-induced ruptures, a carefully planned resection of the mass is required. Benign forms are treated by appendicectomy. However, in malignant mucoceles or cystadenocarcinoma, the treatment of choice is right hemicolectomy, if no mucosal involvement is present. Herein we report a patient with

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appendiceal giant mucocele, who presented with features of acute appendicitis.

CASE REPORT

A seventy-three-year-old female patient suffering from abdominal pain, nausea, vomiting, and dyspeptic complaints over the past two days was admitted to the emergency department. She had a colicky abdominal pain around the umbilicus, which spread to the lower right abdominal quadrant in the last two days. She had no history of alcohol, drug ingestion, or smoking; but she underwent abdominal myomectomy in the past. Abdominal examination revealed mild tenderness, pain in the right lower quadrant and rebound tenderness with palpation, muscular rigidity, and a palpable mass. Digital rectal examination showed tenderness in the anterior part of the rectum. Laboratory findings were almost within normal limits except for leucocytosis (15,500/mm^3) and lactic dehydrogenase (567 IU/ml). Abdominal US demonstrated a mass located in the lower right abdomen, measuring 128 mm. On abdominal CT scans, a round cystic mass was noted located in the same area, with air bubbles in it (Fig. 1). Both abdominal CT and US demonstrated a right iliac cystic lesion adherent to the cecum. It was diagnosed as an abdominal abscess. Based on the radiologic, clinical, and laboratory findings, an emergency laparotomy was performed. Perioperative diagnosis was made as a mucocele of the appendix (Fig. 2a and 2b). Surgical treatment included appendicectomy, partial resection of the ileum, and resection of the cecum. Histopathologic examination confirmed the operative diagnosis. The lesion was reported to be a mucocele and acute suppurative appendicitis with localized peritonitis. The patient was discharged on the postoperative third day and no complications were encountered.

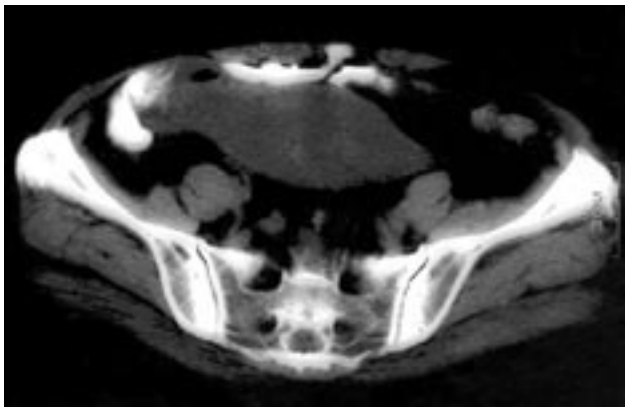

Fig. 1. The tomographic appearance of the giant mucocele located on the right side of the lower abdomen.

DISCUSSION

Appendiceal mucocele has been classified in three histopathologic groups ranging from focal or diffuse mucosal hyperplasia without epithelial atypia to mucinous cystadenocarcinoma.[1] Even larger lesions may be asymptomatic in 25% of the patients.[2,5] It may be incidentally detected at surgery or during radiologic examination for other causes.

In our case, we encountered a giant appendiceal mass consistent with cystadenoma. The possibility of a retention mucocele was excluded depending on the size of the lesion.[6-8] Pain in the right lower quadrant (approximately 60%) mimicking acute appendicitis is the most frequent sign. On physical examination, a mass in the right lower quadrant is detected in 50% of cases. The worst complication is

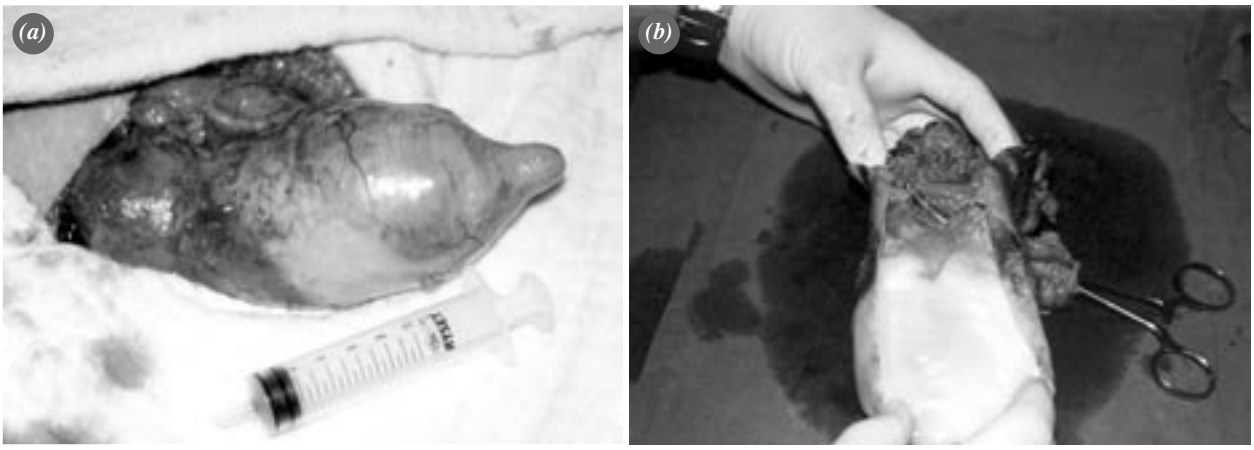

Fig. 2. **(a, b)** The gross operative and postoperative views of the giant mucocele.

pseudomyxoma peritonei, which occurs due to peritoneal dissemination resulting from spontaneous or iatrogenic perforation of the appendix.[9] Intestinal obstruction or intestinal bleeding are rare complications. In neoplastic conditions, tumor markers may indicate cystadenocarcinoma. Retroperitoneal or pleural implants have been reported in malignant cases.[10-12]

Imaging techniques such as CT or US may allow recognition of the lesion preoperatively. Barium enema examination may reveal a failure in the filling of contrast medium into the appendix and a vertical appearance may be observed in the folds of the cecal mucosa. The cecum is often laterally displaced. Colonoscopy may show a "volcano sign" described as an erythematous mass with a crater, through which the mucus is discharged. Computed tomography shows an ellipsoid, thin-walled mass of low-density, with low levels of attenuation and lacking contrast enhancement. The presence of punctuate wall calcifications suggests the diagnosis of an appendiceal mucocele.[13,14] The role of fine-needle aspiration cytology is not clear due to the risk of pseudomyxoma that may be secondary to this technique.[15]

The case presented had an intractable right lower abdominal pain over at least two months. In the last 36 hours the pain became very severe with leucocytosis (16,400/mm^3), vomiting, and nausea. Computed tomography showed a thick-walled cystic mass with liquid content nearby the cecum (Fig. 1). There was no calcification on its wall. Radiologically, the lesion was defined as an abdominal abscess around the appendix. Abdominal US also demonstrated a right iliac cystic lesion adherent to the cecum, consistent with an abdominal abscess.

With the help of appropriate imaging techniques, an accurate preoperative diagnosis is needed to perform a careful resection in order to avoid surgery-induced pseudomyxoma peritonei.[15] A laparoscopic approach may not be appropriate due to the risk of rupture.[16] As far as the type of surgery concerned, appendectomy is performed for simple mucoceles and cystadenoma when the radix of the appendix is intact. For cystadenoma with a large base, cecal resection should be performed. A right hemicolectomy is recommended for cystadenocarcinoma to ensure the surgical margins free of tumor. Radiotherapy and chemotherapy have been shown to improve prognosis. The association with other solid organ tumors of the gastrointestinal tract, breast, kidney, and ovary should be kept in mind, so that an abdominal exploration may be necessary during laparotomy. In particular, appendiceal mucoceles are associated with colonic cancers in 11% to 20% of cases.[1,10]

The five-year survival rate is 100% for benign mucoceles, whereas it is as low as 45% for malignant forms. Pseudomyxoma peritonei, if localized, is usually treated by excision of local mucin deposits.[16] In our case, during exploration, we realized that the lesion was not an abscess from an appendiceal rupture, but a simple appendiceal mucocele or cystadenocarcinoma (Fig. 2a and 2b). We excised the gigantic mass without any rupture. The resected specimen consisted of the cecum and an ileal segment. Then an ileocolostomy was performed. After surgery, care was given to the abdominal exploration. There was no evidence for any other mass lesions in the abdominal organs, nor were there any signs of pseudomyxoma peritonei. The lesion was reported to be a giant appendiceal mucocele (cystadenoma) with a large base. No epithelial atypia was found. The patient was discharged on the postoperative third day without any complications.

It is concluded that the role of imaging and clinical approach is of particular importance in the treatment of appendiceal mucoceles, especially in emergency settings, since some lesions may be caused by mucinous cystadenoma and cystadenocarcinoma.

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