

Mucinous cystadenoma of the appendix. A diagnostic dilemma?

E. Kelemouridou, S. AL Mogrampi, G. Tsavis, M. Verroiotou, T. Rallis, I. Fardellas

Surgical Clinic of General Hospital of Naousa Imathia, Greece

Rezumat

Chistadenomul mucinos apendicular. O dilemă diagnostică?

Mucocelul apendicular prin chistadenom mucinos este o descoperire clinică rară. Aproximativ jumătate dintre pacienți sunt asimptomatici. Este definit ca dilatație a lumenului apendicular datorată acumulării anormale de mucus la acest nivel. Pseudomixomul peritoneal care este consecința rupturii apendicelui reprezintă complicația cea mai gravă. Prezentăm două cazuri de pacienți care s-au prezentat în clinica noastră cu simptomatologie diferită. Primul pacient s-a prezentat cu simptome de apendicită acută, în timp ce al doilea prezenta simptome atipice ca dureri abdominale, masă palpabilă în fosa iliacă dreaptă și mucocel apendicular diagnosticat computer tomografic. La ambii pacienți s-a practicat apendicectomie, iar diagnosticul final histopatologic a confirmat prezența chistadenomului mucinos apendicular care a determinat formarea mucocelului. În concluzie mucocelul este o tumoră rară, care ar trebui luată în considerare în diagnosticul diferențial al unei mase tumorale situate în cadrul drept inferior al abdomenului.

Cuvinte cheie: chistadenom mucinos, apendice cecal

Abstract

Mucocele of the appendix due to mucinous cystadenoma is a rare clinical finding. Approximately half of the patients are asymptomatic. It's defined as the dilatation of the appendiceal lumen due to abnormal accumulation of mucus in to it. The pseudomyxoma peritonei, as a result of rupture of the appendix, is the most dangerous complication (1). We present two case reports of patients that were presented in our Department with different clinical findings. The first patient was presented with symptoms of acute appendicitis, while the second patient showed with atypical symptoms such as abdominal pain, a palpable mass in the right iliac fossa and a diagnosed mucocele of the appendix with the use of computerized tomography. In both patients was performed an appendicectomy and the final histopathology diagnosis confirmed the presence of the mucinous cystadenoma of the appendix which caused the creation of the mucocele. In conclusion mucocele is a rare tumor which must be considered in the differential diagnosis of a mass in the right lower quadrant of the abdomen.

Key words: mucinous cystadenoma, appendix

Corresponding author:

E. Kelemouridou M.D.
4 Kivelis Street, Naousa Imathia, Post
Code 59200, Greece
Tel.: 00306938676895
E-mail:efikelem@hotmail.com

Introduction

Mucocele of the appendix is an uncommon clinical finding due to abnormal accumulation of mucus in to it. There is prevalence in women middle aged or older than in men. Overall, appendiceal mucoceles make up about 0, 3% of appendix specimens (1). Four are the most common causes of mucocele: mucosal hyperplasia (25%), mucinous cystadenoma

(63%), mucinous cystadenocarcinoma (11%) and retention cyst (1%). The management of mucocoele is necessary because of the risk of rupture and associated pseudomyxoma peritonei, which consists the most dangerous complication. Usually are benign entities without pathologic significance and a survival which ranges from 25% up to 100% according to histological type. The patient is often asymptomatic (23-50%) or presents unspecific symptoms such as abdominal pain, abdominal mass, altered bowel habits, weight loss, acute appendicitis or rectal bleeding. Mucocoele may be discovered incidentally during surgery, endoscopic procedures or radiological evaluations (1-2).

Case report

We refer to two case reports of patients that were presented in our Department. In the first case a 67 year old man was presented to the Emergency Department with complaints of abdominal pain, which was located in the right lower quadrant, anorexia and fever. On examination there was tenderness in the palpation of the right iliaca fossa (positive test McBurney) and signs of peritoneal irritation. The laboratory test revealed a leukocytosis and an increased CRP, while the radiological evaluation was normal. In the second case a 72 year old woman was presented as an out-patient complaining of chronic abdominal pain for about 4 months and a palpable mass which was located to the right iliaca fossa. The patient was afebrile and hemodynamically stable. She had a history of an operated umbilical hernia with Mayo technique, while in the family her brother died from colon cancer. On examination was found a palpable mass in the right iliaca fossa without signs of acute abdomen. The laboratory tests were normal except an increased CEA (carcinoembryonic antigen). A computed tomography (CT) was obtained and revealed the presence of the mucocoele of the appendix.

Results

In the first case the patient with the diagnosis of acute appendicitis underwent a surgical operation. During the surgery the appendix was found behind the cecum with intense edema of the wall and length 10cm and dilatation of the distal part with length 7cm, amplitude 5cm close to the base of the appendix. Intraoperatively it wasn't easy to distinguish if the base was infiltrated, so a simple appendicectomy was performed. The specimen was full of mucinous material (Fig. 1). The histology showed mucinous cystadenoma of the appendix. The patient recovered uneventfully and was discharged from the hospital on the 8th day of admission. With the suspicion of infiltration of the base the patient underwent a colonoscopy and the biopsies were benign. In the second case the patient underwent an exploratory laparotomy and during the surgery was identified a large appendix (length 7,5 cm) with intense dilatation of the lumen (bigger diameter up to 3 cm) and full of mucinous material. The base of the appendix and the cecum were partially infiltrated and for this reason was performed incision of the cecum, the appendix was found in the lumen of the cecum and was performed an appendicectomy (Fig. 2). The final pathologic analysis of the specimen was mucinous cystadenoma of the appendix. The patient recovered without complications and was discharged from the hospital on the 10th day of admission.

Discussion

The term mucocoele of the appendix implies a dilated appendiceal lumen caused by an abnormal accumulation of mucus in to it. It may be developed secondary to mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%) and retention cyst. Also can occur due to occlusion of the lumen by endometriosis or carcinoid tumor. Mucocoele of the appendix due to mucinous

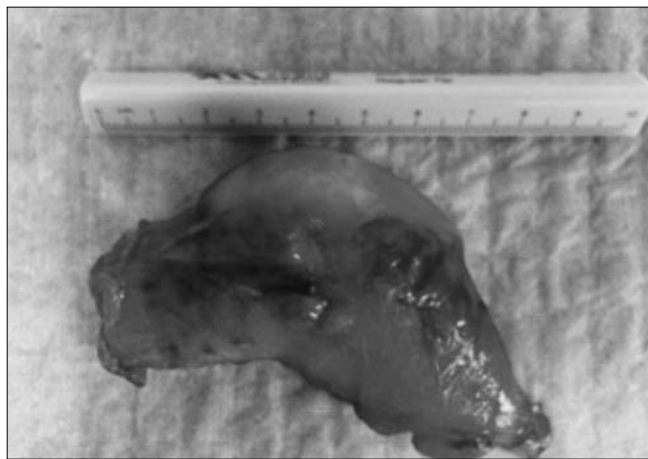
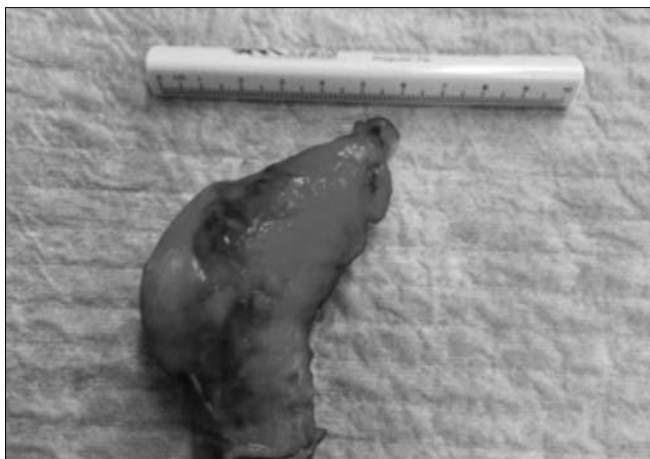


Figure 1. The operative specimen. A giant mucocoele of the appendix



Figure 2. The mucocoele of the appendix during the surgery and after the removal.

cystadenoma is a rare clinical finding and make up about 0, 3% of appendix specimens. Approximately 25% of patients are asymptomatic. The most common symptoms associated with mucinous cystadenoma are abdominal pain which is located in the right iliaca fossa and mimics the image of acute abdomen, palpable mass, weight loss, nausea and vomiting (1-2). Rarer manifestations include gastrointestinal bleeding, intussusception or urologic symptoms.

The differential diagnosis of a palpable mass in the right lower quadrant of the abdomen includes mucinous cystadenoma, mucinous cystadenocarcinoma, lymphoid hyperplasia, lymphoma, carcinoid tumors, periappendiceal abscess, primary adenocarcinoma and ovarian tumors (3).

The diagnosis of the mucocoele can be made either preoperative or intraoperative during a routine appendectomy. The preoperative diagnosis of the mucocoele can be difficult and tests such as ultrasound, computed tomography, barium enema and colonoscopy are necessary. With ultrasound can be seen cystic masses with various echogenity and a wall with calcification. CT is the modality of choice for the diagnosis of the mucocoele due to its ability to show well-encapsulated cystic masses with low attenuation that communicates with the cecum. The presence of calcifications because of the chronic inflammation is a typical finding in computerized tomography. On barium enema usually there is no filling of the appendix with contrast. The colonoscopy is also non diagnostic because the biopsies are most commonly normal. The pseudomyxoma peritonei due to rupture of the mucocoele is the most serious complication (4).

There have been mentioned simultaneous cases of mucocoele of the appendix and colon cancer with incidence up to 20% and for this reason it is recommended endoscopic control of the colon when there is suspicion of mucocoele of the appendix (5).

The treatment of mucocoele of the appendix is surgical and can either be by laparotomy or laparoscopy. A laparoscopic approach although provides a better evaluation of the entire

abdominal cavity, a more rapid recovery and a better cosmetic outcome is not recommended due to the increased risk of rupture of the mucocoele and the provocation of the pseudomyxoma peritonei. The conversion in laparotomy should be made in case of increased risk of injury and rupture or when the tumor extends beyond the appendix. A typical appendectomy is performed in mucinous cystadenoma and mucosal hyperplasia when the appendiceal base is intact, but in cases of local invasion the surgery should be completed with cecal excision. For the mucinous cystadenocarcinoma a right hemicolectomy is recommended. During the surgery should be made an exploration of the entire abdominal cavity due to the probability of coexistence of other tumors such as carcinoma of the colon or ovarian tumors (6-8).

The patients with simple or benign mucocoele of the appendix have a great postoperative survival of 5 years up to 90-100%. On the other hand in cases of mucinous cystadenocarcinoma the survival for 5 years is 25%.

The mucinous cystadenoma of the appendix is a rare clinical finding and the preoperative diagnosis remains difficult even with specific imaging methods. The treatment of choice is the surgical excision of the mucocoele of the appendix.

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