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Appendiceal mucocele - Case Report and Review of the Literature

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Rezumat

Mucocel apendicular - prezentare de caz și analiza literaturii

Mucocelul apendicular (MA) a fost descris pentru prima oară de către Rokitansky în 1842. (1) Incidența sa este între 0.2 – 0.4% din totalul apendicectomiilor efectuate și se întâlnește predominant la femei cu un raport de 4/1 comparat cu bărbații, iar cel mai frecvent la vârsta de 50 de ani. (2,3) Vă prezentăm cazul unei paciente de 64 de ani, îndrumată către investigații amănunțite după un control ginecologic de rutină. Mucocelul apendicular cu localizare retrocecală, învelit de membrane Jackson adiționale, a fost descoperit intraoperator. Formațiunea odată cu apendicele a fost excizată, fără a compromite integritatea mucocelului. Examenul histopatologic a determinat un chistadenom mucinos cu linii clare de rezecție, fără alte date sugerând afectarea țesutului apendicular. Pacienta a fost externată la 3 zile după operație fără alte complicații, iar următorul control medical este programat la 6 luni.

Cuvinte cheie: mucocel apendicular, celule caliciforme, apendice, tumori producătoare de mucus

Abstract

Appendiceal mucocele (AM) was described for the first time by Rokitansky in 1842.1 Its incidence is 0.2 – 0.4% of all apendectomies performed, as it is observed predominantly in women with the ratio of 4/1 versus men and most frequently at the age of 50.2,3 We present to your attention a 64-year-old woman, who was referred to diagnostic further clarification after a preventive gynaecologic exam. Appendiceal mucocele with retrocecal location, enveloped by additional Jackson's membranes was determined intraoperatively. The formation has been dissected and appendectomy was performed without mucocele integrity being compromised. The permanent histological specimen revealed mucinous cystadenoma with clear resection lines, without any data on the appendiceal base being affected. The patient was discharged on the third day after surgery without any complications, and further follow-up was scheduled in 6 months.

Key words: appendiceal mucocele, goblet cells, appendix, mucous-producing tumours

Introduction

Appendiceal mucocele (AM) was described for the first time by Rokitansky in 1842. (1) Its incidence is 0.2 – 0.4% of all apendectomies performed, as it is observed predominantly in women with the ratio of 4/1 versus men and most frequently at the age over 50. (2,3) Appendiceal mucocele represents a progressive appendix dilatation caused by intralumenal accumulation of mucous substance, as it may be a malignant or a benign process. (4,5) Appendix is covered by epithelium

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containing a large number of goblet cells, which are significantly more in comparison to colon, due to which most appendiceal epithelial tumours are mucinous and begin as mucocele. (6,7)

Mucocele is a heterogeneous group which, according to the component that causes it, is of neoplastic and non-neoplastic type (non-epithelial type). An underlying inflammatory process or a fecalith obturation was the reason for appendicocele formation in the last group. (8,9) In most cases appendiceal mucocele is a benign process. (8)

Case report

We present to your attention a 64-year-old woman, who was admitted to the Department of Surgery at University Hospital "Queen Joanna" and referred to diagnostic further clarification after a preventive gynaecologic exam, which revealed a tumour formation in the right lower abdominal quadrant. Computer tomography (CT) that has been performed determined a well-formed capsulated formation, located between bladder and cecum and lying on iliac vessels, without any data on being connected to them. Imaging test was not able to determine formation origin. There was no evidence of visible calcificates and enlarged lymph nodes. Fibrocolonoscopy (FCS) was performed in addition which did not reveal any pathological finding. A solid tumour formation, located in the ileocecal region, 8/4 cm in size, heterogenous and hypoechogenous structure with layered structure was echographically visualised, as a connection to intestine was not visualised, it was well circumscribed from surrounding tissues and had smooth walls – a suspected retroperitoneal cyst. The results of the complete blood count (CBC) and biochemistry tests performed revealed no abnormalities. Tumour markers - CEA, CA 19-9, CA 125, CA 15-3, CA 72-4 were tested in addition, which were within the reference range.

A decision on conventional (open) surgery was made due to the unclear diagnosis and the possible dense accretion to iliac vessels. Appendiceal mucocele with retrocecal location, enveloped by additional Jackson's membranes was determined intraoperatively. (Fig. 1)

The formation has been dissected and appendectomy was performed without mucocele integrity being compromised. A frozen section was sent – clear resection lines. No evidence of enlarged lymph nodes. The intraoperative finding is presented on the following two figures. (Fig. 1 and 2)

The permanent histological specimen (Fig. 3) confirmed the diagnosis according to the type of mucinous cystadenoma, but with clear resection lines, without any evidence of appendiceal base being affected.

The patient recovered quickly. Intestinal passage was recovered on the second day after surgery. She was discharged on the 3rd day after surgery. A follow-up exam was performed 6 months after discharge, as follow-up computer tomography, complete blood count and biochemistry – without any pathological finding.



Figure 1. *Intraoperative mobilization of the appenceal mucocele*

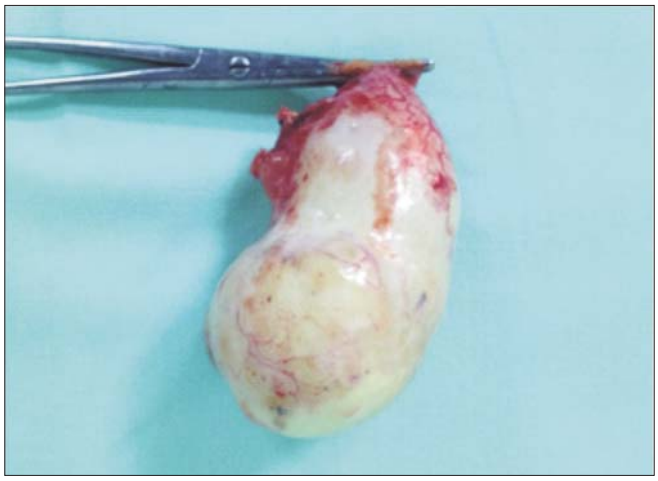


Figure 2. *Appendiceal mucocele after resection*

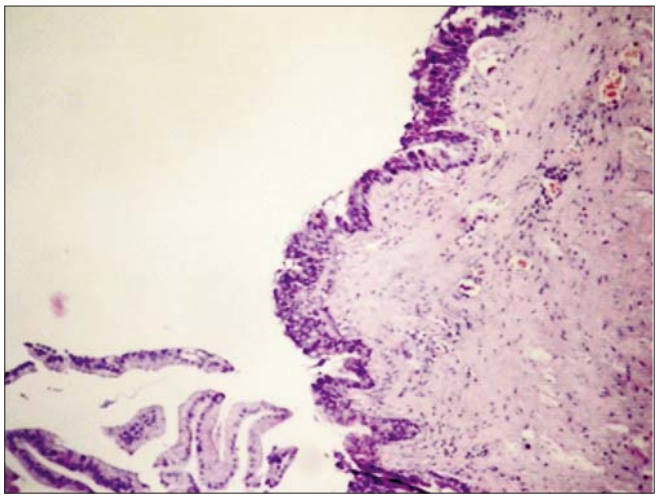


Figure 3. *Appendiceal mucocele*

Discussion

The incidence of appendiceal tumors is 0.5 - 2 %, as 8% of them present a mucocele. (8,11)

AM is divided into four subgroups: (10)

1. Retention cyst – intramural accumulation of mucoid substances, in rare cases bigger than 2 cm.
2. Mucous hyperplasia –dilatation of the appendix with presence of zones with hyperplasia of epithelial layer.
3. Mucinous cystadenoma – lumen dilatation exceeding 6 cm with low-level dysplasia
4. Mucinous cystadenocarcinoma – with stromal invasion and intraperitoneal dissemination, similar to ovarian mucinous cystadenocarcinoma

Clinical presentation of AM is non-typical. It may often be completely asymptomatic and can be diagnosed during accidental preventive imaging tests. The most common symptoms are pain in the right lower quadrant (27%), palpable abdominal mass (16%), weight loss (10%) and change in the intestinal function (5%). (12,13)

AM may take a symptomatic course, as well, as with appendicitis, which is one of the possible complications. Other complications include bleeding, peritonitis, intestinal obstuction, invagination, appendiceal torsion or fistula formation. (8,12) The most severe complication is spontaneous or iatrogenic rupture, with developing of peritoneal pseudomyxoma (PMP). This condition complicates significantly treatment and final outcome.

The typical image, called 'onion skin', is an important sonographic marker of appendiceal mucocele. (14) Visualisation of appendix and its diameter exceeding 15 mm is considered as specific appendiceal mucocele index with sensitivity of 83 % and specificity of 92 %. (2,15,16,17,18) Discontinuity of the appendiceal wall with leakage of the internal contents into surrounding tissues indicates rupture of the mucinous tumor. (19) In cases of developed pseudomyxoma peritonei (PMP) typical ultrasound findings are non-mobile, sonogenic ascites with multiple semi solid masses and scalloping of the hepatic and splenic margins. (20,21)

An important indication during FCS performance is the formation of the so called 'volcano sign'- a visible raised zone in cecum with an appendicular orificium located in its centre. (22) FCS performance is important in diagnostic aspect in order to exclude synchronous or metachronous colon tumour, which may be determined in up to 29 % of the cases. (2,15, 16,17,18)

In case of CT it is important to assess formation interrelation with surrounding organs, which may make diagnostication easier. CT is considered to be the most informative imaging method, although accurate diagnostication is often not possible. In most of the cases a well-capsulated cystic formation with calcificates in the wall is visualised, which is considered a specific index, as well. (23,24) Another important CT indication is the presence of appendiceal lumen exceeding 1,3 cm. (6) When the mucinous tumor is ruptured and pseudomyxoma peritonei is developed the most common findings on a CT scan are a large volume of mucinous ascites with the density of fat that displace the small bowel and the normal mesenteric fat. (20) Other characteristic findings are omental thickening, multiseptate lesions, scalloping of organs, and curvilinear calcifications (20,21,25,26,27).

In cases of appendiceal mucocele Magnetic Resonance Imaging (MRI) reveals well-capsulated lesions, cystic masses – hypertensive to T2 and hypo - or isotensive to T1 series. MRI have comparable sensitivity to the CT scan.

In the end echography and CT are non-specific diagnostic methods, as in differential and diagnostic aspect one must think of appendicitis, benign neoplastic process, cystic formation, carcinoid, lymphoma, mesenteric cyst, ovarian tumour or another malignant neoplasia of appendix. (2)

In differential and diagnostic aspect, in most of the cases appendiceal mucocele may not be easily distinguished from ovarian cyst or tumour. Cases of simultaneous development of ovarian epithelial tumour of ovary with appendicocele and endometrial adenocarcinoma, have been reported but no genetic relation was established so far. (10)

A basic aspect of distinguishing between appendicitis and mucocele is to consider wall thickening exceeding 6 mm. (10) The clinical signs between acute appendicitis and appendiceal mucocele especially when it is ruptured with mucin leakage often are indistinguishable. Because Appendiceal mucocele and pseudomyxoma peritonei are usually diagnosed after the age of 50 (28), we think that is mandatory to perform CT / MRI scan after this age when patient with clinical signs of appendicitis is presented.

Tumor tissue should be subjected to CK20 and CK7 immunohistochemistry. CK20 is a cytokeratin and intestinal tumor marker while CK7 is also a cytokeratin and marker of gynecological malignancies. (20,29) Several tumor markers-CEA, CA 19,9 and CA125 have been reported to have diagnostic and prognostic value for patients with mucinous neoplasms. These tumor markers also can be used for post-operative fallow-up.

Surgery is the only treatment with curative potential. Surgical treatment depends on the dimensions and histology of the mucinous neoplasm, as well as the clinical presentation. (19) During the operation we must be cautious while handling the mucocele to avoid rupture and dispersion of mucus or epithelial cells into the peritoneal cavity as this is associated with a poorer prognosis. (19,30) Conventional surgery is preferred rather than laparoscopic approaches for the treatment. (2) Laparoscopic approach has an increased risk of rupture and subsequent pseudomyxoma peritonei formation. (2) In general, laparotomy, is advised to permit careful exploration of other viscera, in particular the colon and ovaries, because of the reported association between appendiceal mucocele and colonic and ovarian tumors in 11_20% of cases.(19,31,32,33)

Few authors still recommend a minimally invasive approach. (34,35,36) If, however, the mucocele appears to be a homogeneous cyst, with no nodularity and no sign of perforation, then careful laparoscopic handling may be acceptable. (37)

Appendectomy is considered to be sufficient in case of

mucous hyperplasia or cystadenoma with an intact appendiceal base, as no long-term follow-up is required in this case. The prognosis for retention cysts and adenomas is excellent: 5-year survival is 91-100%. (31) No long term follow-up is needed for these patients. (2)

In case of cystadenoma with positive boundaries – base affection, cecum resection is necessary. If there are data for cystadenocarcinoma with or without lymph nodes, hemicolectomy with thorough lymph dissection must be performed 10 Long term follow-up is obligatory for these patients. (2)

The entire abdomen should be inspected for signs of mucin and peritoneal dissemination. If mucin is present then it must be collected completely and sent for cytology, because the presence of cells in mucin localized to the right lower quadrant provides significant prognostic information. (19,38)

The traditional approach to psudomyxoma peritoni is based on repeated surgical debulking procedures. Due to the presence of tumor deposits after the first debulking surgery, this approach could results in short term palliation with imminent recurrence or progression, hence redo procedures and a shorter 5- to 10-year overall survival (OS) rate of approximately 50%. (39)

In recent years more authors prefer the aggressive strategy developed by Dhage-Ivatury and Sugarbaker (2006) and it is based on the present perforation or rupture, clarity of boundaries and the presence of positive lymph nodes (LN). (40) This surgical approach includes peritonectomy and visceral resections- also called cytoreductive surgery (CRS). The goal of the procedure is complete removal of the tumor. Surgery is followed by local drug administration aimed at eliminating microscopic and/or minimal residual disease left in the abdominal cavity following surgical manipulations. (41,42) The additional effects of hyperthermia, through the use of a special pump, increase local tissue drug concentration and consequently antiblastic drug activity. (41,43) This technique has been defined as hyperthermic intraperitoneal chemotherapy (HIPEC). (41)

As a result of the approach of Sugarbaker 5-year survival rate of 86% has been reported for some patients. (44)

Conclusion

AM is a rare finding which must be taken into consideration in case of any formation in the right ileocecal region. We recommend conventional surgical treatment having in mind the exclusive importance of mucocele intactness preservation and the possibility of better field isolation. The case report presented by us concerns appendiceal mucocele according to the type of mucinous cystadenoma, without any evidence of appendiceal base being affected, due to which we considered appendectomy to be a surgical intervention of sufficient volume.

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