

MUCOCELE OF THE APENDIX - A CASE REPORT AND REVIEW OF THE REFERENCES

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ABSTRACT

Mucocele of the appendix is a rare clinical and pathological entity with a higher incidence in the female population, and non-specific symptomatology that includes more diagnoses including neoplastic and non-neoplastic causes of the appendix dilation and mucin proliferation. With obtained numerous variations in symptomatology, clinical picture, intraoperative presentation, surgical treatment and histopathological findings, the aim of this paper is an inspection into current knowledge of this disease, its histopathological characteristics, differential diagnosis and current treatment modalities. We present a patient with a clinical picture of the acute abdomen and numerous comorbidities. Ultrasound diagnostics indicated the presence of thin-walled dilatation of the small intestine, especially in the lower right quadrant, as well as the presence of free fluid in the peritoneal cavity. Appendectomy and removal of the mucocele and resection of the gangrenous sigmoid colon according to Hartmann were done. A definitive histopathological finding suggests ischemic colitis and retention mucocele of the appendix with normal mucosa. Histopathologically, four types of mucocele appendixes are distinguished, namely the retention mucinous cyst, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma. Surgical treatment is the method of choice in treating these tumors, either open or laparoscopic. Imperative is the preservation of the integrity of the entire mucocele, due to the possible dissemination of contents of the cyst and the subsequent development of pseudomyxoma peritonei.

Keywords: Mucocele of the appendix, surgical treatment, pseudomyxoma peritonei.



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INTRODUCTION

Mucocele of the appendix is a rare but well-known clinical and pathological entity with non-specific symptomatology which includes more diagnoses, including neoplastic and non-neoplastic causes of the appendix dilation and mucin proliferation. This disease is characterized by dilatation of a lumen as a result of an accumulation of a large amount of mucus (1, 2). A patient's clinical symptoms may include pain in the right lower quadrant of the abdomen, palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, and signs of intussusception of the intestines (1, 3). Although with the incidence of pathological changes of 0.2 to 0.3% in the general population, with a slightly more frequent incidence in women and people after the age of 50, usually can be detected during abdominal surgery, routine radiological diagnosis or during endoscopy (4-6). Approximately 23–50% of patients are asymptomatic (4, 7).

Ultrasound findings usually show a cystic mass that is filled with dense content and marginal calcification. The CT is the most accurate method of diagnostics. The CT can be used to discover the signs specific to mucocele with high accuracy: appendix lumen more than 1.3 cm, its cystic dilatation, and wall calcification (8).

In the case of appendiceal mucocele, resection is recommended for possible malignancy, and the imperative is to maintain an integrity of the mucocele wall during surgery to prevent peritoneal pseudomyxoma (9-11). However, although laparoscopic intervention is traditionally contraindicated in mucocele appendix due to the perforation risk, some studies indicate that possible benefits of the minimally invasive surgical treatment over classical appendectomy (12, 13). Histopathologically, four types of this entity are distinguished, namely retention mucinous cyst, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma (1, 3, 14).

With obtained numerous variations in symptomatology, clinical picture, surgical treatment and histopathological findings in the case of this clinical-pathological entity, the aim of this paper was to present the case of mucocele of the appendix, and to explain more closely the problems that may arise in clinical practice.

CASE REPORT

A patient was first referred to a surgeon due to abdominal pain and diarrhea. At the reception the patient's follower (son) states that the patient did not vomit, had a normal miction control, and that he had a stroke 4 months ago and that he was immobile and poorly communicative ever since. The companion also states that the patient operated the abdominal hernia 30 years ago. He was regularly used acetylsalicylic acid for cardiological and neurological purposes, and the follower negates patient's allergy to medicines and food.

Objectively, the patient on reception was conscious, non-communicative, afebrile, eupneic and with normal skin color. Locally, anterior abdominal wall was at the chest level, palpably painfully sensitive on the deep palpation to peritoneal reaction and with rigidity. External inguinal openings are free, and kidney boxes are insensitive to succussion. Laboratory findings showed that patient has granulocytosis (17.81) in the differential blood count, and the ultrasound of the abdomen because of meteorism was insufficient. The liver is seen in a minimal part, which is insufficient for interpretation, and the pancreas and spleen are covered by gases. The right kidney does not visualize, while the left is without a path. Urinary bladder was empty. The dilated intestinal convoluts are seen with the linear probe, at the right ilica fossa with diameter up to 38 mm. Also, on the right, perivesically, between the intestinal bruises a smaller amount of free liquid is observed. Based on the clinical picture and laboratory findings, the patient was hospitalized and initially treated conservatively.

As an adequate clinical response did not follow, the patient was prompted for surgical treatment on the same day. After a short and immediate operative preparation, the patient undergone general endotracheal anesthesia. After the opening of the abdomen, a gangrenally-altered part of the sigmoid column (length of about 20 cm) was discovered by exploration, with a slightly cloudy liquid paracolic and in the Douglas space. Further exploration found a cystic change of about 4x2 cm (a cystic expansion of the distal two-thirds of appendix), which was in close contact with the terminal ileum. After exploration, appendectomy (Figure 1) was performed, with complete removal of the cyst without perforation, and resection of the gangrenous part of the sigmoid colon by Hartmann procedure (Figure 2). Then the distal part of the colon was closed with the GIA stapler, and the proximal part of the left colon was derived on the lateral left abdominal wall – in the form of an unipolar colostomy. Local toilet and haemostasis were made. Douglas's pouch was clean. An abdominal drain was placed in the Douglas space, followed by the closure of the surgical space by layers. Appendix with cyst (Figure 3) and sigma were sent to a pathohistological analysis with a clinical diagnosis: Abdomen acutum pp gangrenae coli sigmoidei, Mucocela appendicis, Collitis, and as an operative procedure: Laparotomia mediana supra et infraumbilicalis, Appendectomia et extirpatio mucocoeellae appendicis, Resectio coli sigmoidei sec Hartmann, Drainage cavi Douglasi. The post-operative course worsened, due to current comorbidities, and the next day patient was prolonged further treatment in an adequate ICU.

A pathohistological examination has shown that it is ischemic colitis (peritonitis fibrinospurulent acuta focalis) and appendiceal mucocele, i.e. retention type of mucocele with normal mucosa (Figure 4).



Figures 1. i 2. Intraoperative finding of mucocele of the appendix. An intact cyst is seen as well as a partially altered gangrenous part of the sigmoid colon

Figure 1.



Figure 2.

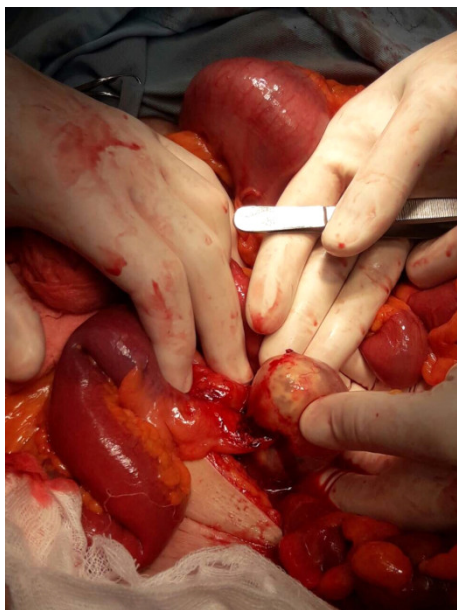
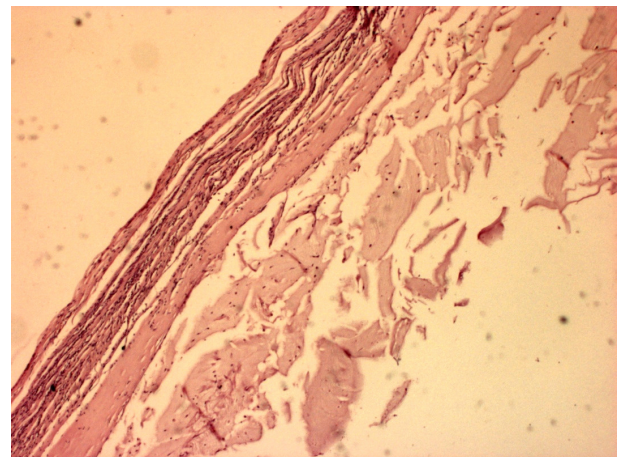


Figure 3. Material for biopsy - preparation of mucocele of the appendix (gross appearance)



Figure 4. Histopathological finding (microscopic slide) of mucocele of the appendix with normal mucosa



DISCUSSION

Mucocele of the appendix was recognized first by Karl Freiherr von Rokitsky in 1842. (4, 15,16) and was described by Virchow in 1863 (16). Feré applied the name “mucocele” in 1877 (16). The major pathological mechanisms that are responsible for the formation of appendiceal mucocele are elevated appendiceal pressures as sequelae of luminal obstruction caused by prior inflammation, mucosal

hyperplasia, or appendiceal lesions (fecaliths, endometriosis, diverticulae, polyps) and tumors of the appendix (carcinoid, cystadenoma, cystadenocarcinoma) (3, 7). Mucinous cystadenocarcinomas are less common than mucinous cystadenomas. This neoplasm rarely spreads through the lymphatic or vascular routes, but has an atypical tendency of penetration and spread well beyond the appendix to the peritoneum (3, 10). Moreover, in advanced cases the whole peritoneal cavity becomes distended with adhesive semi-solid mucous; a



condition termed pseudomyxoma peritonei (17, 18). Complications of appendicular mucocoele include: the intestinal bleeding/obstruction, melena, pyonephrosis, intussusception, perforation, peritonitis and pseudomyxoma peritonei (7, 19).

One of basic principles of the patients' surgical treatment with mucocoele of the appendix is that an intact mucocoele is not a danger to the patient. However, if it is perforated, and consequently its content is spread into the peritoneum, that finally leads to the development of peritoneal pseudomyxoma, whose surgical treatment is difficult, and the prognosis for the patient is not always favorable (9, 20). However, in this case, the patient was primarily operated due to an advanced gangrenous process on the sigmoidal part of the colon, while the appendiceal mucocoele was an incidental finding. For that reason we did not perform any extensive surgical procedure on the right colon, but only appendectomy with intact cyst excision.

In the references it has traditionally been indicated that in the case of mucocoele appendix the advantage is given to open surgical techniques, in comparison with laparoscopic methods. If, however, a laparoscopic technique is applied and an appendiceal mucocoele is detected, it is necessary to continue the surgery with an open surgical technique, for the following: 1) prevention of perforation of the cyst during surgery, and preventing the spillage of content in the peritoneal cavity; 2) an open surgical method, the transparency of the operating field is better, and palpation is enabled, as well as a direct examination of the site in the abdomen where the mucocoele is localized to (18, 20, 21). However, in recent years, there have been numerous studies that have indicated the safety of laparoscopy in the surgical treatment of this disease (13, 22-25). Very important algorithm for the selection of the type of surgery has been proposed by Dhage-Ivatury and Sugarbaker (26). It pointed out three main factors: (1) Is there or not a perforation of a mucocoele i.e. wall integrity; (2) Involvement of the base of an appendix (margins of resection); (3) Is there any positive lymph nodes of mesoappendix and ileocolic. These criteria determine further treatment modalities: appendectomy to the right colectomy, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy. Simple appendectomy is the choice for patients with benign mucocoele as suggested by the presence of a normal caecum and appendicular base and no evidence of perforation (20, 27). Right hemicolectomy is recommended when malignant mucocoele is suspected by the presence of a perforated mucocoele, enlarged mesenteric lymph node or a positive cytology. Due to the well-known association between the appendicular mucocoele and other mucin-secreting cells cancers, such as colon and ovarian cancers an accurate exploration of the abdomen is advised (4, 28).

In the case of mucocoele of the appendix, a histopathological finding is of a great importance. Namely, histopathologically, there are four types of this rare entity – the retention mucinous cyst or lumen occlusion by carcinoid tumor (18%), mucosal hyperplasia (20%), mucinous

cystadenoma (52%) and mucinous cystadenocarcinoma (10%) (3, 14, 17). In this specific case, retention mucocoele with a normal mucosa was obtained histopathologically. The reference data indicate that appendiceal mucocoele represent about 8% of appendiceal tumors, because if there is a condition of pseudomyxoma peritonei, there is probably a malignant component, or cystadenocarcinoma of appendix (1, 29). Most of these tumors are detected incidentally, presenting as the acute appendicitis in as much as 40% of cases, and a definitive diagnosis is made by the postoperative pathohistological finding. Patients with benign mucinous cystadenomas have an excellent with 5-year survival rates of those greater than 90%. Taking into account that malignant cystadenocarcinoma peritonei and pseudomyxoma have a 5-year survival rate of early 25%, despite recent advances in treatment modalities, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy, it is obvious that there is need for recognition, management and follow-up (11, 21). In conclusion, mucocoele of the appendix are treated exclusively surgically, and preoperative diagnosis assists in planning the careful mobilization and resection, in order to prevent the occurrence of possible peritoneal contamination. If there is a suspicion of malignancy, the method of choice is the right hemicolectomy. In the concrete case, there was no suspicion of malignancy, and this entity was an incidental finding. However, since the risk of colorectal adenocarcinoma is a significantly higher in patients with appendiceal mucocoele compared to those who do not have it, regular controls are needed in the postoperative period.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The study was conducted in accordance with the ethical standards of the committee responsible for human experimentation (institutional and national) and the Helsinki Declaration of 1975, as revised in 2013. Voluntary written and informed consent was obtained prior to enrollment in the study.

CONFLICT OF INTERESTS

The authors declare no conflict of interest.

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