APPENDICEAL MUCOCELE - A REVIEW OF LITERATURE WITH A CASE REPORT

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ABSTRACT

Appendiceal mucoceles represent neoplastic and non-neoplastic, dilatated, mucus filled appendix vermiformix. Appendectomy is obligatory due to a possible malignancy. It is crucially important to avoid rupturing of the mucocele because it can result in pseudomyxoma peritonei, with high morbidity and mortality. We presented a 52-year-old man with pain and palpable mass in the lower right quadrant of the abdomen. The mucocele was removed without a rupture, and the patient was discharged from the surgical department one day after the surgery without a complication. The resection must be done very carefully, because the rupture of a mucocele can cause pseudomyxoma peritonei, a very dangerous and often lethal condition. Due to the concern of rupture, we performed the classical resection through laparotomy. It is very important, especially for young, inexperienced surgeons to be aware of this rare diagnosis and perform a surgical intervention according to the guidelines of good clinical practice.

Keywords: *Appendiceal Mucocele, Pseudomixoma Peritonei, Case Report.*

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BACKGROUND

Appendiceal mucocele, first described in 1842 by Rokitansky, is a rare diagnosis, with less than 1% of pathologies related to the appendiceal disease. It represents a distended appendix filled with mucus, not some specific disease. Mucoceles of the appendix are diagnosed as incidentaloma in about 50% of cases. The symptoms can include the abdominal pain or mass, weight loss, nausea, changes in bowel habits, or they can be presented as acute appendicitis (1). Sometimes, appendiceal mucocele can be presented as acute appendicitis or an adnexal mass, mimicking the true nature of a disease (2). US and CT exams are usually sufficient imaging diagnostic tools. Colonoscopic findings may be characteristic with a "volcano sign", presenting the appendiceal orifice elevated and covered by normal mucosa or sometimes with a lipoma-like submucosal tissue (3).

CASE PRESENTATION

We presented an otherwise healthy 52-year-old man with the symptoms of the right-lower quadrant abdominal pain and palpable mass. The abdominal ultrasonography revealed suspicion of appendiceal mucocele, which was confirmed by MSCT of the abdomen (Figure 1). CEA, CA 19-9 and CA 125 were within a normal range.

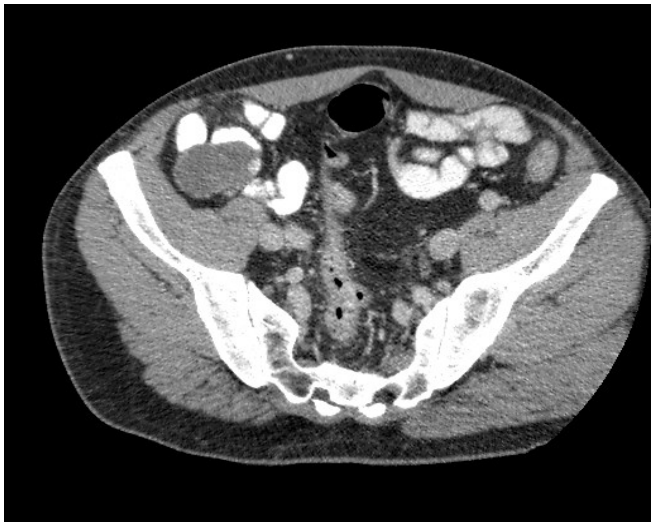


Figure 1. MSCT findings of appendiceal mucocele.

During the open access appendectomy, we clearly identified basis of the appendix, whose diameter was cc 5mm and after careful manipulation, the appendix and mucocele were removed intact, without a rupture of the mucocele wall or mucin leakage (Figure 2). A carefully maintained exploration of the peritoneal cavity excluded any mucin deposits. We performed the "ex tempore"- frozen section biopsy which revealed benign characteristics of the specimen and simple appendectomy was a sufficient treatment. The patient was discharged from the surgery department one day after the surgery without any complaint.

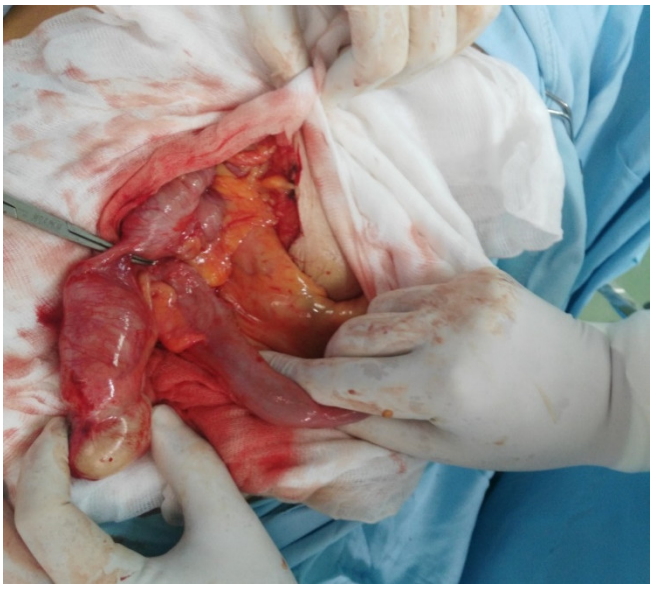


Figure 2. Surgical specimen of AM with an intact wall, basis of the appendix is not affected.

Consecutive pathology demonstrated appendiceal mucocele, with the wall covered by a single-line epithelium of benign histomorphological characteristics, and the lumen filled with a non-structural eosinophilic mucin (Figure 3 and 4).

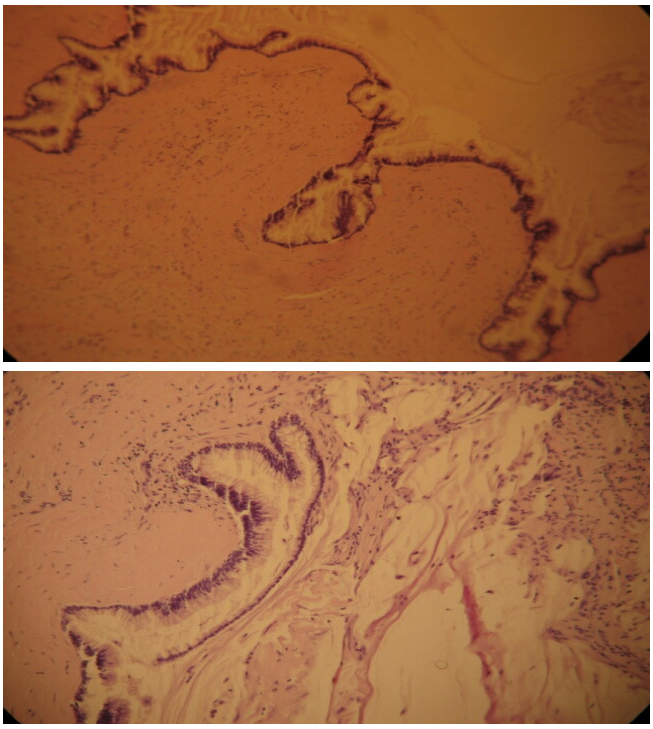


Figure 3 and 4. The wall of the mucocele coated with a single-stranded epithelium of benign histomorphological characteristics and the lumen filled with a non-structural eosinophilic mucin.

DISCUSSION

Historically, appendiceal mucoceles have been classified into four types: A. Non-neoplastic, including (1) a simple mucocele-retention cyst often caused by a fecalith or structure obstructing the appendiceal outflow, (2) mucosal hyperplasia, and B. neoplastic, including (3) a benign mucinous cystadenoma, and (4) a mucinous cystadenocarcinoma. The cystadenomas are typically referred to as low grade appendiceal mucinous neoplasms (LAMNs) (4).

A recent international Delphi consensus has suggested that the term "cystadenoma" should no longer be used for appendiceal tumors. "Mucinous adenocarcinoma" should be used to describe mucinous tumors with infiltrative invasion, "low-grade appendiceal mucinous neoplasm" for describing those with spreading growth but without destruction, "high-grade appendiceal mucinous neoplasm" describes tumors with low-grade architectural features but high-grade cytological features, and term "signet ring carcinoma" for those with over half of the cells with the signet ring morphology. Pseudomyxoma peritonei can be of low-grade, high-grade, and signet ring cell histological type (5).

Mucoceles <2 cm are usually benign, while sizes >6 cm are more often associated with malignancy and a higher rate of perforation (6).

A mucocele is usually diagnosed by the abdominal CT scan. The typical finding of appendiceal mucocele is a low attenuated, well-encapsulated, thin-walled cystic mass in the right lower quadrant of the abdomen (7).

The most feared complication, occurring secondary to a spontaneous or iatrogenic rupture, is pseudomyxoma peritonei (PMP), which represents mucinous deposits within the peritoneal cavity. PMP is poorly understood; however, it is known to develop insidiously as a result of mucin producing neoplastic, epithelial goblet cells forming mucinous implants throughout the abdominopelvic peritoneum and it is associated with significant morbidity and mortality (8).

Both benign and malignant mucoceles can produce mucinous peritoneal deposits known as pseudomixoma peritonei (PMP). A 5-year survival rate in the first case is about 91–100%, while the prognosis for a malignant mucocele is much worse, with only 25% of a 5-year survival rate (9).

With some aggressive regimens, such as HIPEC (heated intraperitoneal chemotherapy) and cytoreductive surgery, 10-year survival can raise close to 50% (10).

For a long time, laparoscopy has been considered as a contraindication for the surgical treatment of appendiceal mucocele. Also, some authorities suggested laparotomy for better exploration of the peritoneal cavity searching for mucin deposits (11).

However, with technical development and general improvement in surgical skills, there is a raising number of recent reports describing safe laparoscopic or robotic assisted removal of appendiceal mucocele (12, 13).

Apart from the dilemma regarding the surgical approach (open vs laparoscopic surgery), there is still a debate concerning the extent of resection, from appendectomy as the only treatment to a right hemicolectomy. To determine indications for the right hemicolectomy, González Moreno and Sugarbaker recommended the use of a sentinel lymph node biopsy, with a frozen section examination. According to them, the indications for the right hemicolectomy are: (1) inability to achieve total removal of the primary tumor or complete cytoreduction by appendectomy, (2) appendiceal or ileocolic lymph nodes involvement, and (3) a non-mucinous neoplasm confirmed by the histopathological examination (14).

A useful practical algorithm, proposed by Filho and collaborators, suggests appendectomy with excision of the mesoappendiceal fat and lymph nodes if appendiceal base is not affected. If the base is involved, typhlectomy or partial right hemicolectomy is indicated. The frozen section is mandatory in both cases, and if a specimen is benign, the right colectomy is not indicated. A malignant specimen indicates an oncologic right hemicolectomy (15).

If a rupture of AM occurs, followed by adenomucinosis, a 5-yearfollow-up period with CT scans and CEA and CA19-9 levels monitored every six months are recommended. In the case of AM perforation with a confirmed diagnosis of mucinous adenocarcinoma, a second-look surgery is recommended, six months after the initial intervention (16).

CONCLUSION

The diagnosis of AM should always be considered in all cases of cystic lesions of the right lower quadrant. The surgical resection of a mucocele is the therapy of choice, open approach or laparoscopy, depending on the surgeon's skills. Benign and especially malignant AMs rupture must be avoided due to PMP and a consequent high mortality rate. The management of appendiceal mucocele is simple, ranging from appendectomy only to the right hemicolectomy followed by HIPEC, depending on the intraoperative and pathological findings. Clear understanding of this rare condition and optimal surgical treatment based on algorithms of good clinical practice are critical.

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CONFLICT OF INTEREST

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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