



DIAGNOSIS AND TREATMENT OF APPENDICEAL MUCINOUS NEOPLASM AFTER THE HYSTERECTOMY AND ADNEXECTOMY PERFORMED PRIOR

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ABSTRACT

Mucinous appendiceal neoplasms are rare entity with different prognostic value. Surgery offers the optimal treatment for mucinous neoplasms in patient with no metastatic disease. We present the case of patient with mucinous appendiceal neoplasm that had prior been undergone to hysterectomy and adnexectomy. Considering the intraoperative finding and no signs of perforation or metastatic disease appendiceal resection with the resection margin high on appendiceal basis was performed. No further oncological treatment was necessary.

Introduction

Appendiceal mucinous neoplasms (AMN) represent 0.2–0.7% of all appendix specimens. Appendiceal mucoceles (AM) or mucinous neoplasms are rare lesions characterized by a distended and mucus-filled appendix. The appendix epithelium contains many goblet cells and thus the accumulation of mucus is a typical finding.^[1]

The general surgeon should be aware of the diversity of its clinical manifestations and biological behaviors along with the significance of the surgical treatment on the progression of the illness and the prognosis of the patients. The AMN are usually detected as an unexpected surgical finding.^[2]

They present a heterogeneous disease depending on the presence of mucinous histology, histologic grade, and stage.^[3]

Appendiceal mucinous neoplasms can be presented as acute appendicitis in only 8% to 14% of the cases.^[1]

But the clinical presentation is rather unspecific. Most of these AM are asymptomatic but can become symptomatic because of inflammation, presenting as an acute appendicitis or by causing nonspecific abdominal pain. The preoperative diagnosis of AM helps to avoid accidental iatrogenic perforation during surgery.

Other symptoms included abdominal pain, abdominal mass, weight loss, nausea or vomiting, obstipation, and changes in bowel habits. In the emergency setting AM can also be presented as intestinal strangulation, appendiceal intussusception, or generalised abdominal pain. Approximately 30% of patients may present with perforated appendicitis or extravasation of mucus during surgery and this can result in pseudomyxoma peritonei. Although both the benign and malignant variants of AM may cause pseudomyxoma peritonei, this is more frequent and with worse prognosis for malignant cases.

Ultrasound (US) and computed tomography (CT) imaging studies are valuable for the detection of AMN and can be easily performed in the emergency setting. Ultrasound examination can detect AMN with a sensitivity of 83% and a specificity of 92%. Although the lesion size is not associated with malignancy AMN smaller than 2 cm are rarely malignant. Simple mucoceles have a mean diameter of 4.1 cm while cystadenomas have 8.1 cm.^[1]

The distinction between primary ovarian mucinous tumors and appendiceal mucinous neoplasms metastatic to the ovary can be challenging given the overlap of morphologic features and immunohistochemical expression of traditional markers.^[4]

There are reported cases of pseudomyxoma peritonei associated with low-grade appendiceal mucinous neoplasms where there was colonization of the endometrium and cervical mucosa by low-grade atypical enteric-type mucinous epithelium. Pseudomyxoma peritonei (PMP) may rarely result in endometrial and cervical mucosal involvement, presumably secondary to transtubal spread.^[5]

There is also a case report of the squamous cell carcinoma of the cervix metastasizing to the appendix and presenting as appendicitis.^[6]

We present the patient with the AMN after the hysterectomy and bilateral adnexectomy was performed prior due to cervical carcinoma gradus CIN III.

Case report

A 53-year-old woman presented to the Department of surgery with abdominal pains, weakness and feeling of intraabdominal pressure. One year prior the admission she underwent the laparotomy due to cervical carcinoma gradus III when the hysterectomy and adnexectomy were performed. Clinical examination revealed tenderness in the lower abdominal part while laboratory tests showed no abnormalities.

Abdominal US revealed the hypoechoic mass in the lower right abdominal part. The abdominal CT confirmed existing of intraabdominal cystic formation which seem to be related to the right hemicolon. (Figure 1).

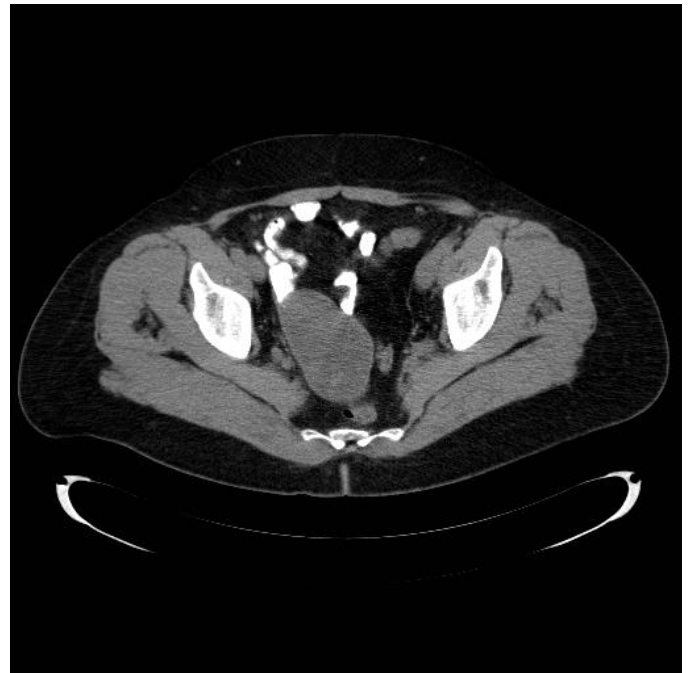


Figure 1. Abdominal computed tomography showing the appendiceal cystic formation

After the usual preparation the partial relaparotomy was performed. Abdominal cavity exploration revealed the appendiceal neoplastic formation measuring 12 cm x 8 cm. (Figure 2). Considering the intraabdominal finding, no signs of perforation nor metastatic disease, we performed the appendectomy as an urgent operation so the neoplastic formation was extracted together with the resection margin high on appendiceal basis.

Early postoperative course went fine, the patient was after five days discharged and had no complications.

The pathological analysis revealed low grade AMN. The patient was afterwards presented to the onkologist.

There was no adjuvant therapy needed and the control examinations showed no signs of metastatic disease.

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Discussion

The operative findings and, especially, tumor histology, determine the type of surgery. Intestinal histologic subtype behaves and should be treated similarly to the right colon neoplasms; while mucinous tumors, often discordant between histology and its aggressiveness, can be treated with a simple appendectomy or require complex oncological surgeries. Mucinous tumors are often associated with the presence of mucin or tumor implants in the abdominal cavity, being the clinical syndrome known as PMP. PMP tends to present an indolent but deadly evolution and requires a multimodal approach as a single treatment with curative potential.^[7]

The optimal surgical approach for treating an appendiceal mucocele remains controversial. Traditionally neoplasms of the appendix more than 2 cm in diameter are managed by right colon resection. The rationale for this approach is the resection of occult lymph nodal metastases within the ileocolic lymphatic system. At the time of appendectomy in the emergency setting, gross examination and the assessment of the size of the mucocele cannot reveal the malignancy of the lesion. In these cases it is important to consider every mucocele of the appendix as malignant. The laparoscopic approach has been described for the management of the appendiceal mucocele and is still recommended by some authors in selected patients. Single-port laparoscopic surgery for appendiceal mucoceles has also been reported to be safe and feasible. However, González Moreno et al. suggest conversion to open appendectomy in case of mucocele revealed during laparoscopic appendectomy. The open approach permits a safe and gentle surgical manipulation of the lesion. Furthermore port site recurrence after laparoscopic approach has been reported.

After an initial urgent operation if the histological diagnosis reveals positive lymph nodes, adenocarcinoma of the intestine, mucinous adenocarcinoma, carcinoid or adenocarcinoid tumors larger than 2.0 cm, or high mitotic rate, a right hemicolectomy should be performed. Patients with perforated AM in the initial surgery but with negative lymph nodes or margins in the histological diagnosis should not be submitted for a right hemicolectomy as they present lower survival rates when compared to those who only had an appendectomy at the time of the primary surgery. If the histological exam shows the presence of mucinous peritoneal carcinomatosis, then the patient will need cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) with the prospective of a long-term survival. Low-grade tumors have the maximum survival benefit from these locoregional treatments.^[1]

Previous studies have demonstrated that the prognosis of disseminated mucinous appendiceal neoplasms is highly dependent upon tumor grade. Reflecting this, the 7th edition of the American Joint Committee on Cancer (AJCC) staging system now incorporates a three-tier grading system for prognostic staging of mucinous appendiceal tumors. However, the grading criteria are not well described.^[8]

Conclusion

The surgery plays the main role in the treatment of mucinous appendiceal neoplasms. The optimal surgical approach for treating an appendiceal mucocele remains controversial. The operative findings and, especially, tumor histology, determine the type of surgery. Low-grade tumors have the maximum survival benefit from the locoregional treatments.

Since the rare cases are described and prognostic factors and grading criteria are not still well described, we believe that more research about appendiceal neoplasms is necessary.

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