

Retroperitoneal Giant Mixed Adenoneuroendocrine Carcinoma: Case Report

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ABSTRACT

Mucinous cystadenocarcinoma remains the second most frequent cause of appendiceal mucocele. Presently described is a case of mixed adenoneuroendocrine carcinoma (MANEC) located in the right retroperitoneal space. A 64-year-old male was admitted with a palpable mass in the right lower quadrant. An abdominal computed tomography scan revealed a cystic mass in the pelvic area 31x25x25 cm in size. A search of the literature yielded more than 50 similar cases, with mean diameter of 13 to 40 cm; however, only a few of these cases were mucinous cystadenocarcinoma. Usually, an appendectomy is sufficient therapy for cystadenoma and hyperplasia, while a right hemicolectomy is required for cystadenocarcinoma. To our knowledge, the present case is the first with a pathological diagnosis of MANEC (30/70 neuroendocrine carcinoma and adenocarcinoma) to be reported.

INTRODUCTION

Appendiceal mucocele is a rarely seen disease characterized by dilation of the appendiceal lumen with mucus. The accumulation of excessive mucin causes abnormal dilation, leading to the formation of a cystic mass.^[1,2] The second most prevalent form of appendiceal mucocele is mucinous cystadenocarcinoma.^[3,4] The incidence of adenocarcinoma in the histopathological examination of all appendectomy specimens has been reported to be 0.08%.^[5,6] Approximately 40% of all appendiceal adenocarcinomas are mucin-secreting mucinous adenocarcinoma.^[7] Mucoceles may rupture spontaneously or during surgery and develop into pseudomyxoma peritonei.^[8] This case report describes a giant (maximal diameter: 31 cm), rarely seen, appendiceal mixed adenoneuroendocrine carcinoma (MANEC) filling the right retroperitoneal cavity of a 64-year-old male patient.

CASE REPORT

A 64-year-old male patient had complaints of difficulty walking persisting for a year, but which had become aggravated in the previous 3 months, swelling of the right foot, pain in the right lower quadrant, weight loss, and pretibial edema. A physical examination revealed abdominal distension and a palpable mass in the right lower quadrant. His laboratory findings were within normal limits. Abdominal computed tomography (CT) revealed a multiseptated cystic mass (31x25x25 cm) with patchy areas of calcification that filled the right abdominal quadrant and extended from the lower edge of the liver to the pelvis and obscured the planes around the iliopsoas muscle. The mass was adjacent to the right kidney, right iliac artery, and the ureter (Fig. 1). Intraoperatively, a mass lesion extending from the liver to the pelvis and approaching the midline was observed. A giant mucocele was separated from the

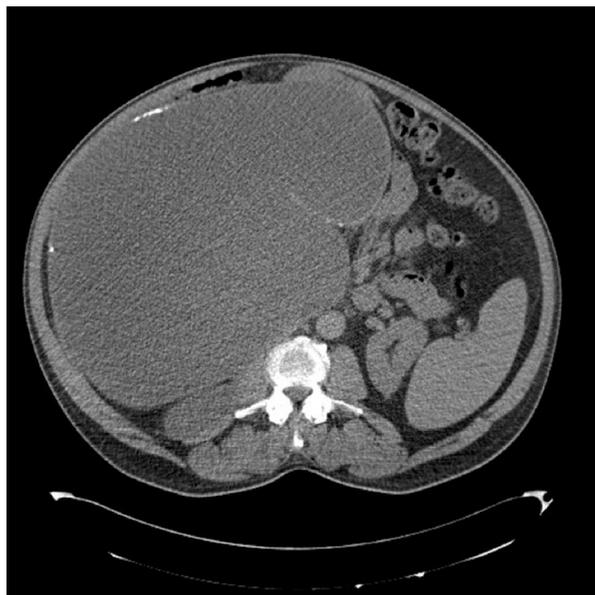


Figure 1. A multiseptated cystic mass measuring 31x25x25 cm that largely filled the right side of the upper abdominal cavity and extended from the subhepatic space to the right pelvis.

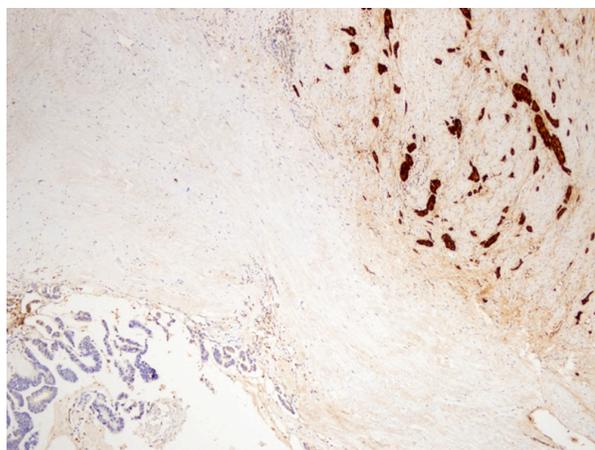


Figure 2. Diffuse pools of mucin consistent with mixed appendiceal adenoneuroendocrine carcinoma and the focal atypical glandular structures of neuroendocrine carcinoma/adenocarcinoma with a ratio of 30/70.

adjacent organs and tissues with sharp dissection, and a tumoral mass originating in the retrocecal appendix was detected. The mass was removed en bloc with a right hemicolectomy. Histopathological examination of a sample revealed appendiceal MANEC (30/70 neuroendocrine carcinoma/adenocarcinoma) (Fig. 2). The diagnosis of this case was consistent with the 2010 World Health Organization classification of MANEC.^[9] The patient underwent a regimen of oxaliplatin, folinic acid and infusional 5-fluorouracil (FOLFOX-4) as postoperative adjuvant chemotherapy. No local recurrence was detected in 2 years of follow-up.

DISCUSSION

Appendiceal mucocele is rare. It is a cystic mass that emerges following neoplastic changes to the appendiceal mucosa; the lumen becomes enlarged due to an abnormal accumulation of mucus.^[1,2] Mucocele of the appendix was first defined by Rokitansky.^[4] There are 4 histological types: simple mucocele or retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.^[10,11] Mucinous cystadenocarcinoma is the most frequently seen type after mucinous cystadenoma, and constitutes 11% to 20% of appendiceal mucoceles. A review of the literature revealed some 50 cases of large mucoceles, with a mean diameter of between 13 and 40 cm. However only a very few of these were mucinous cystadenocarcinoma.^[12,13] Cases of MANEC are very rare.

Abdominal CT aids in the differentiation between appendiceal mucocele and other lesions that may mimic appendiceal mucocele. The presence of a nodule on the wall of the mucocele may be considered suggestive of mucinous cystadenocarcinoma.^[1] In our case, in images, the wall of the cystic lesion in the right lower quadrant resembled the intestinal wall. The lesion extended to the cecum, and at the junction with the cecum, concentric layers typical of mucocele were detected. All of these radiological findings established the diagnosis of appendiceal mucocele. However, it is very difficult to differentiate between cystadenoma and cystadenocarcinoma preoperatively.^[7,14] Tumor markers, such as carcinoembryonic antigen and cancer antigen 19.9, are not very valuable in this instance.^[15,16]

Surgical strategies for these patients differ widely. As a standard approach, although an appendectomy is considered sufficient in cases of cystadenoma or hyperplasia, in patients with cystadenocarcinoma it should be combined with a right hemicolectomy.^[17,18] If there is no involvement of a neighboring mesenteric organ or peritoneal involvement, then removal of the appendix with its mesentery will suffice in cases of cystadenocarcinoma.^[19] However, if there is invasion of the surrounding organs or tissues, then a right hemicolectomy is advised. Although there is controversy about a preference between open and laparoscopic surgery in cases of appendiceal mucocele, open surgery is recommended in cases with cystadenocarcinoma due to the large size of the lesion and the risk of rupture. In our case, we performed a right hemicolectomy using an open approach.

In conclusion, these rare cases can grow to tremendous size and push away the adjacent organs. Preoperative evaluation and determination of the appropriate surgery strategy is very valuable.

Informed Consent

Approval was obtained from the patients.

Peer-review

Internally peer-reviewed.

Authorship Contributions

Concept: S.K., Y.E.A., Ö.A.; Design: S.K., Y.E.A.; Data collection &/or processing: S.K., Y.E.A., Ö.A.; Analysis and/or interpretation: S.K., Y.E.A., Ö.A.; Literature search: S.K., Y.E.A.; Writing: S.K., Y.E.A., Ö.A.; Critical review: S.K., Y.E.A., Ö.A.

Conflict of Interest

None declared.

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Retroperitoneal Dev Mikst Adenöroendokrin Karsinom: Olgu Sunumu

Apendiks mukoselinin en yaygın ikinci görülme şekli musinöz kistadenokarsinomdur. Sağ retroperitoneal boşluğun tamamına yakını dolduran nadir görülen mikst adenöroendokrin karsinom (MANEC) olgusu sunulmaktadır. Altmış dört yaşında erkek hasta; sağ alt kadranda palpabl kitlesi mevcuttu. Bilgisarlı batin tomografisinde (BBT) 31x25x25 cm boyutlarına ulaşan multiseptalı kistik kitle görülmüştür. Literatürde 50 olgu rapor edilmiş olup; büyük ve dev mukosellerin ortalama çapları 13 ile 40 cm arasında değişmesine rağmen bu olguların sadece bir kaçında musinöz kistadenokarsinom rapor edilmiştir. Operasyonda; kistadenom ya da hiperplazide apendektomi yeterli tedavi olarak görülse de kistadenokarsinomda sağ hemikolektomi yapılmalıdır. Bu olgumuz patolojik tanısı mikst adenöroendokrin karsinom (nöroendokrin karsinom/adenokarsinom oranı 30/70) olması dolayısıyla literatürde tek olgudur.

Anahtar Sözcükler: Görüntüleme tetkikleri; kistadenokarsinom; mukosel.