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Two concurrent appendiceal neoplasms in an elderly patient: A case report

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ABSTRACT

INTRODUCTION: Acute appendicitis, one of the commonest surgical diagnoses, is rare and more complex presentation in the elderly. Physicians must consider atypical causes appendicitis in this population, which could affect the management of the patient.

PRESENTATION OF CASE: An elderly female presented with a two-day history of lower abdominal pain, associated with low-grade fevers and chills. Studies showed leukocytosis and computed tomography (CT) findings consistent with appendicitis. She underwent laparoscopic appendectomy. Intra-operatively, the Appendix had an unusual appearance, so a frozen-section was obtained, suggestive of a mucinous neoplasm with grossly clear margins. Despite the possibility that she may require a more extensive cancer operation pending the final Pathology results, the decision was made to complete the operation at this stage, and return at a later date if needed, after completing the patient's work-up with a colonoscopy to rule out any synchronous colonic lesions that could alter her surgical management. Final Pathology revealed both a low-grade appendiceal mucinous neoplasm, as well as a tip carcinoid tumor, both of which were adequately treated with appendectomy alone.

DISCUSSION: Physicians treating elderly patients with appendicitis should suspect an atypical etiology, such as appendiceal cancer. Early identification and appropriate pre-operative counseling may alter the surgical management.

CONCLUSION: The role and timing of right hemicolectomy in treating appendiceal cancers remain controversial, and should be evaluated on a case by case basis.

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1. Introduction

Acute appendicitis, though very common in the adolescent and young adult population, is unusual in the elderly [1]. It is estimated that only 5%–10% of the cases occur in elderly patients [2]. In places with large geriatric populations, although a straightforward inflammatory appendicitis is possible, one must have a high index of suspicion for an atypical etiology such as cancer. This diagnosis, when made intra-operatively, could alter the immediate management of a patient. This work has been reported in accordance to the Surgical Case Report (SCARE) guidelines [3].

2. Case

A 67 year old Caucasian female with a history of hypertension, gastroesophageal reflux disease, and no prior abdominal surgeries,

presented to the hospital with a two day history of abdominal pain. The pain, initially diffuse, had localized to the right lower quadrant, associated with low grade fevers and chills. She denied any nausea, vomiting, diarrhea, or loss of appetite. Laboratory studies revealed a mild leukocytosis without left shift. CT imaging showed a moderately dilated fluid-filled Appendix with mild wall thickening and periappendiceal inflammatory changes consistent with appendicitis.

The patient was taken to the operating room that evening for a laparoscopic appendectomy. Upon entry into the abdomen, the abdominal contents were inspected, including the liver, and found to have no abnormalities except a large, firm, bulbous Appendix that was inflamed without evidence of perforation. The appendiceal tip appeared normal. The specimen was easily resected with no signs of spillage. Taking into consideration the patient's older than average age, prolonged time course of symptoms, and abnormal looking appendix, the decision was made to obtain a frozen section intraoperatively. Pathology findings were suspicious for a low grade mucinous neoplasm with clear surgical margins.

Given the tumor location at the base of the appendix, it was thought that the patient would likely require a completion right

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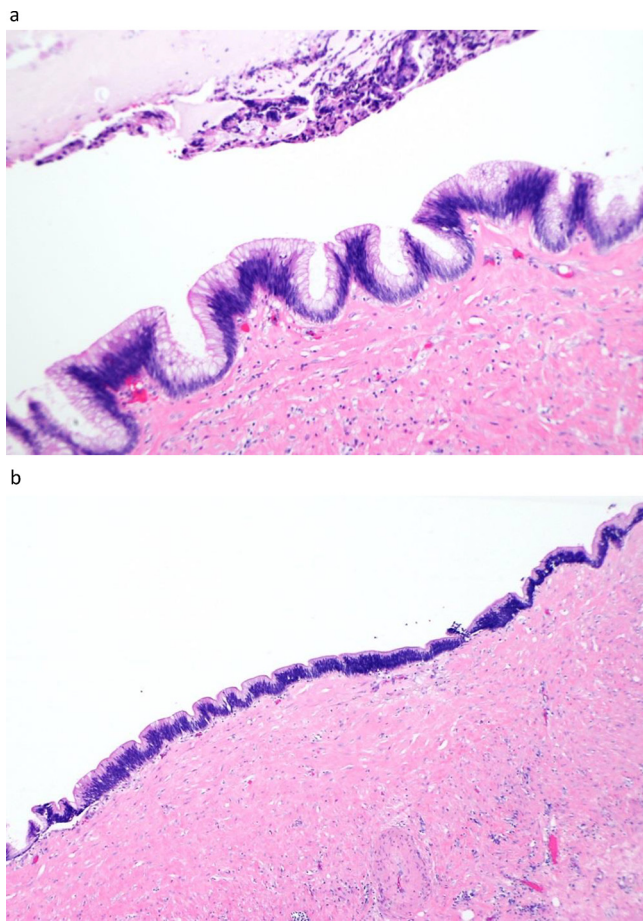


Fig. 1. a. Low Grade Appendiceal Mucinous Neoplasm (LAMN), High Power Field. b. Low Grade Appendiceal Mucinous Neoplasm (LAMN), Low Power Field. a and b: LAMN – Sections of the mucinous cystic lesion reveal a mildly undulating epithelium, lacking underlying lamina propria, resting atop an attenuated fibrous stroma. Scattered papillary type tufting can be appreciated. Single cells overlap minimally, contain low grade slightly elongated nuclei, and abundant mucin-filled cytoplasm. Extravasated mucin can be seen within the appendiceal lumen.

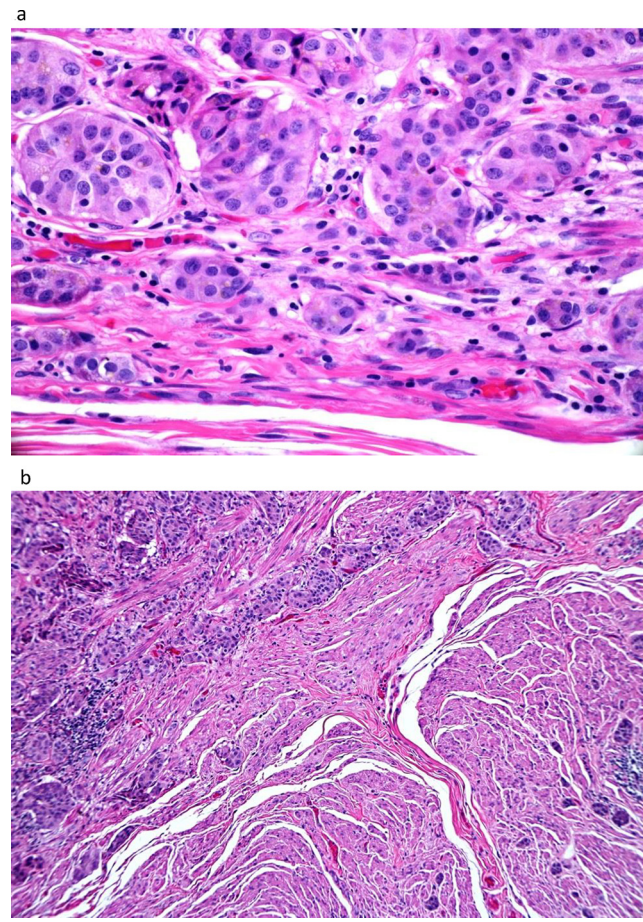


Fig. 2. a. Neuroendocrine Tumor (NET), High Power Field. b. Neuroendocrine Tumor (NET), Low Power Field. a and b: NET – Sections of the appendiceal tip reveal an infiltrating low grade neuroendocrine tumor extending into the muscularis. The tumor characteristically is composed of small nests of bland monotonous cells with small salt and pepper nuclei and somewhat amorphous eosinophilic cytoplasm. No mitotic figures are present.

hemicolectomy; however, the surgeon favored first obtaining a pre-op colonoscopy to rule out any synchronous lesions prior to proceeding with a colonic resection. The patient recovered well from the first operation and was discharged home on post-operative day #1. Histologic evaluation revealed an appendix that grossly measured 8 cm in length and was proximally dilated to a maximal diameter of 3 cm. There was no evidence of appendiceal rupture or serosal mucin extravasation. Serial sectioning revealed a proximally dilated appendix with copious intraluminal mucin. The appendiceal wall was fibrotic and mildly thickened up to 0.5 cm in this area. Within the wall of the distal appendix was a small slightly rubbery nodule measuring approximately 1 cm. The final pathology report revealed a low-grade appendiceal mucinous neoplasm (LAMN) without presence of invasion or dissecting mucin, as well as a 1.1 cm well differentiated grade 1 neuroendocrine tumor (“carcinoid”) at the distal tip of the specimen (Fig. 1).

3. Discussion

This case of appendicitis is interesting and rare in that it was precipitated by not one, but two concurrent, distinct appendiceal neoplasms. The incidence of appendiceal neoplasm is rare, usually found incidentally in about 1% of appendectomy specimens and appendiceal neoplasms constitute less than 0.4% of all gastrointestinal tract tumors [4]. A diagnosis of appendiceal cancer is

rarely made pre-operatively because of similar presenting symptoms to acute appendicitis; however risk factors for cancer include older age (55–65 years) [4], abnormally long duration of symptoms (>2 days), and low hematocrit (<38%) [5]. The patient in this case met all three of these criteria, suggesting a high relative potential cancer risk.

LAMN, also known as mucinous cystadenoma, is a benign tumor of the appendix. It is the most common benign appendiceal neoplasm, accounting for 0.3% of appendectomy specimens, or about 60% of benign appendiceal tumors [6,7]. If resected with clear margins, appendectomy alone is sufficient treatment. However, this condition can predispose to appendiceal mucocele formation, as well as the development of pseudomyxoma peritonei if perforation occurs [6]. This complication can lead to need for future surgical debulking, or the consideration of other controversial therapeutic options such as intraperitoneal chemotherapy.

Carcinoid is the most common malignant tumor of the appendix, accounting for about 0.5% of appendectomy specimens, or 66% of appendiceal cancers [6]. Appendectomy alone is performed for lesions <2 cm in size, located within the tip or body of the appendix, and without evidence for metastasis. If any of these parameters are violated, a right hemicolectomy is indicated (Fig. 2).

Because of the rare nature of appendiceal cancers, there is no clear consensus regarding the timing of pursuing a definitive operation. In instances when an appendiceal mass violates the base or invades into the cecum, it is widely accepted that an immediate

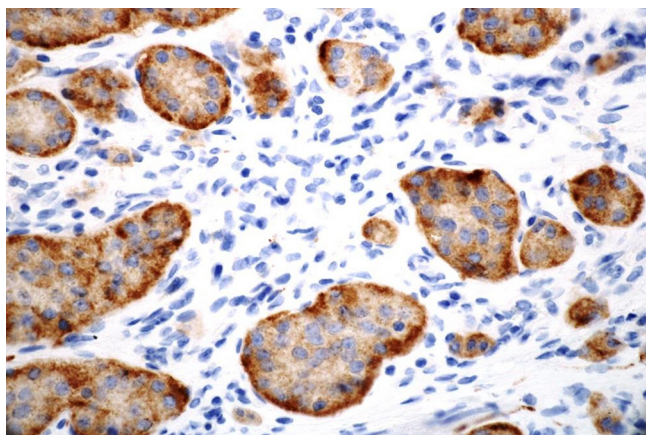


Fig. 3. A synaptophysin immunohistochemical stain highlights the neuroendocrine cells.

partial cecectomy or ileocecectomy should be performed to avoid impaired healing or seeding of the staple line. In the absence of an obvious mass or a mass in the appendiceal body or tip, however, the majority of cancer diagnoses are not made until the final pathology can be determined several days after the initial operation, after which the patient is counseled to return for an interval right hemicolectomy.

In this case, the patient had an obvious appendiceal mass that did not seem to violate the base of the appendix. Given our high suspicion for cancer both pre- and intraoperatively, we considered moving forward with immediate right hemicolectomy at the same operation. Benefits would have included subjecting an elderly patient to a single operation rather than multiple stresses to the body, as well as avoiding a delay in care if the pathology did indicate cancer. On the other hand, about 10% of appendiceal neoplasms can be attributed to a secondary malignancy, most commonly from colon cancer (55%), as well as there being a high incidence of synchronous and metachronous colorectal cancer in all appendiceal tumors [3]. In these instances, a right hemicolectomy may not encompass the diseased areas, and a second more extensive operation may still be needed for definitive treatment. Despite her older age, this patient's relatively low number of comorbid conditions made it seem more appropriate to hold off on further surgical intervention until more diagnostic information could be obtained.

4. Conclusions

Risk factors for appendiceal cancer include old age, longer duration of symptoms, and low hematocrit. When working with a predominantly geriatric population, attention to these details can aid in establishing a pre-operative index of suspicion for cancer, as well as guide intra-operative practice that may alter treatment decisions. Elderly patients presenting with acute appendicitis should uniformly be counseled on the potential need for a more extensive cancer operation based on their risk factors. Although appendiceal neoplasms are rare overall, this case report demonstrates a patient with multiple risk factors for cancer, who

ultimately presented with two synchronous appendiceal tumors, both of which turned out to be benign. Despite current guidelines, there continues to be disagreement regarding the survival benefit of a right hemicolectomy for appendiceal cancer, and each case should be evaluated based on clinical judgement on an individual basis (Fig. 3).

Conflicts of interest

None.

Funding

None.

Ethical approval

We did not seek IRB approval for this case report, which contains only retrospective, deidentified patient information. The writing or publication of this case report did not affect this patient's treatment or outcomes in any way. There are no ethical dilemmas with this case.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors contribution

Riva Das – Primary author, researcher, resident surgeon for case.
Joshua Cantor – Pathologist, provided pictures and captions.
Thai Vu – Attending surgeon for case.

Guarantors

Riva Das.
Thai Vu.

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