Incidental Intramucosal Adenocarcinoma of the Appendix Presenting as Acute Appendicitis

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Abstract

Intramucosal adenocarcinoma arising in an adenomatous lesion of the appendix is an uncommon occurrence and has rarely been reported in a young patient per our literature search. This report is of a 29-year-old woman with this lesion and her course.

Learning Objectives:

- Routine histologic examination of appendectomy specimens is critical to detect rare cases of neoplasms.
- Right hemicolectomy is the suggested treatment for appendiceal adenocarcinomas per the ASCRS guidelines.

Case Description

The patient is a 29-year-old female who presented to an urgent care with complaints of nausea and right flank pain. She was subsequently referred to our emergency department, where a CT scan showed probable appendicitis characterized by enlargement, hyperenhancement, and wall thickening. Her medical history revealed that she was currently 3 months post-partum and taking Flagyl for bacterial vaginosis, and that she had a history of protein S deficiency, asthma, and remote liver lesions that were favored to represent hemangiomas and hepatic adenomas per imaging. Her vitals and labs were normal and physical exam revealed only abdominal and right flank tenderness.

She was then taken for a laparoscopic appendectomy, where the appendix was described as enlarged and inflamed without perforation. The resected appendix was sent to pathology where upon opening they discovered a 1.7 cm yellow-tan polypoid mass in the appendiceal tip. Histologic sections showed intramucosal adenocarcinoma arising in an adenomatous lesion with extensive high-grade dysplasia and features of both conventional and traditional serrated adenoma. No invasion was identified. The patient was discharged two days later and has since undergone a right hemicolectomy with node dissection that was negative for malignancy. Her follow up care entails surveillance colonoscopy in 1 year.

Introduction

Intramucosal adenocarcinoma arising in an adenomatous lesion of the appendix is an uncommon occurrence and has rarely been reported in a young patient per our literature search. This report is of a 29-year-old woman with this lesion and her course.

The rate of appendiceal cancer in acute appendicitis is 0.53% in the U.S. and hovers around 1% for all routine appendectomies. It is known that this rate increases in patients above age 50 and correlates with increasing age. Overall it is considered an orphan disease, meaning it affects less than 200,000 people nationwide, and is often discovered incidentally on routine histologic examination.[1] The likelihood of preoperative detection is low, ranging from 6.6% - 25%.[3]

Appendiceal neoplasms are broadly categorized into two histologic subtypes: neuroendocrine and epithelial, with the latter further classified into goblet cell adenocarcinomas, LAMNs, HAMNs, and adenocarcinomas including mucinous, non-mucinous, and signet ring cell subtypes.[5]

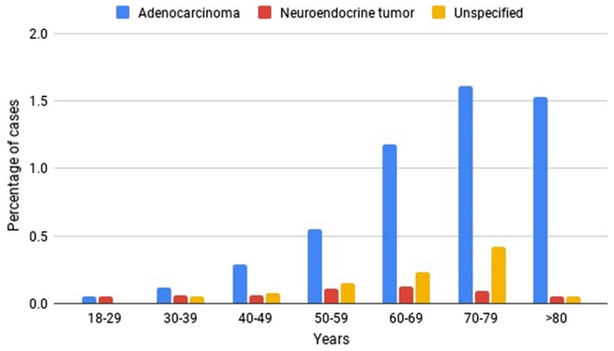

Figure 1. The incidence of adenocarcinoma, neuroendocrine, and low grade appendiceal mucinous neoplasm (LAMN) obtained from the National Surgical Quality Improvement Program (NSQIP) database from 2010 to 2018 categorized into 10 year age groups.[1]

Imaging

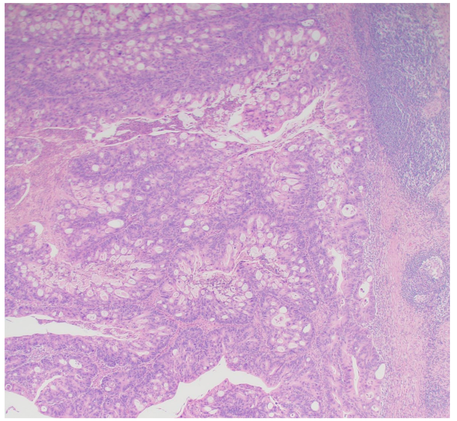

Figure 2. This 4x view reveals disrupted cellular architecture and features of serrated adenoma. The submucosa can be visualized in the upper right and demonstrates a lack of invasion into the deeper appendiceal structures.

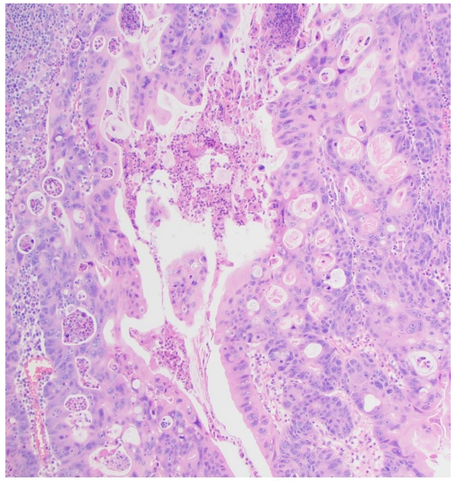

Figure 3. This view at 10x shows high grade cytologic atypia with multiple mitotic figures and necrotic debris within the gland lumen.

Discussion

This report has detailed the course of a young patient with intramucosal adenocarcinoma of the appendix whose presentation was consistent with acute appendicitis by symptoms, radiology, and physical exam. The incidence of appendiceal neoplasms rose in the early 2000s without a clear rationale.[5] Although this remains a very rare occurrence, this case serves as an important reminder that cancer can be found in anyone and may manifest in atypical presentations. This further supports the pathologist's role in the grossing and careful analysis of appendiceal specimens following appendectomy.

The increased risk of neoplasm in patients above 40 years of age should be a factor when discussing non-operative management of appendicitis in this population. In addition, it was found that appendectomy due to complicated appendicitis (involving abscess, perforation, or necrosis) carried a 3.24% risk of malignancy as compared to uncomplicated appendicitis with a 0.87% risk, a statistically significant difference.[2] This patient had a CT scan that showed a dilated appendix without evidence of perforation. When appendiceal adenocarcinomas are found, the 5-year survival is markedly improved in patients undergoing appendectomy + right hemicolectomy at 63% as compared to appendectomy alone at 20%.[3] Further treatment with adjuvant chemotherapy is indicated in nodal-positive adenocarcinomas.[5]

Of the classifications of appendiceal neoplasms, signet ring cell carcinoma and adenocarcinoma carry the worst prognoses.[4] 13-42% of patients with a primary epithelial appendiceal neoplasm will go on to develop concurrent colorectal cancer.[5] This patient underwent prophylactic right hemicolectomy as well as colonoscopy with negative findings for GI neoplasm. She was also followed with a CEA to monitor for recurrence. At this time there is little data on the benefits of trending serum tumor markers in appendiceal neoplasms, but it is reasonable to extrapolate data from their use in colorectal cancers.[5]

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