Appendiceal Adenocarcinoma: The Great Pretender

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Introduction

- Appendiceal adenocarcinoma (AA) is a rare primary malignant neoplasm that arises from the cells lining the appendix[1]
- Appendiceal neoplasms can be divided into carcinoid, mucinous, goblet cell, and adenocarcinoma based on cytologic examination[1]
- Incidence is 0.12 per 100,000 people per year[2]
- AA accounts for 0.5% of all gastrointestinal neoplasms per year[2]
- Most common presentation of appendiceal neoplasms includes signs and symptoms of acute appendicitis including RLQ pain, nausea, anorexia, and vomiting[4-6]
- Definitive diagnosis is established through biopsy and pathology report[1]
- Past medical history, computed tomography, and ultrasound are tools to help establish appendiceal adenocarcinoma as part of the differential diagnosis[1]
- Treatment options depend on the histologic characteristics of the neoplasm and include appendectomy, hemicolectomy, cytoreductive surgery, peritonectomy, chemotherapy, and hyperthermic intraperitoneal chemotherapy[1,7-9]

Case Description

- 53-year-old female presented to the emergency department with a 3-week history of dull abdominal pain radiating to her RLQ, fatigue, sore throat, nausea, and vomiting. Denied changes in bowel movements, weight loss, blood in stool, or gynecologic symptoms.
- Vitals: febrile (101°F), hypertensive (162/83 mmHg), all other vitals within normal limits.
- Physical exam only notable for diffuse abdominal tenderness to palpation with tenderness over McBurney's point.

Medical History

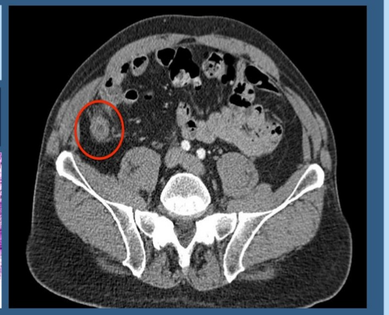
- **Past Medical History**: colonic polyps, abnormal uterine bleeding
- **Past Surgical History:** excision of tubular adenomas from the cecum and sigmoid, removal of hyperplastic polyps in the sigmoid colon
- **Social History**: former tobacco user (10.4 pack year), marijuana use
- **Family History**: prostate cancer, melanoma
- **Review of Systems**: non-contributory besides abdominal pain

Diagnostic Testing

- CMP and CBC within normal limits

Histology[11]

Imaging[10]



- Abdominal CT scan revealed acute appendicitis with a 21 mm thickened appendix
- RUQ ultrasound within normal limits

Table 1. Pathology Report

Tissue	Appendix
Histologic Type	Adenocarcinoma
Margins	All margins negative
TNM Staging	pT: pT3 (through the muscularis propria into the subserosa) pN: pN0 - no tumor involvement in 7 regional lymph nodes examined pM: not applicable – no metastasis

Hospital Course

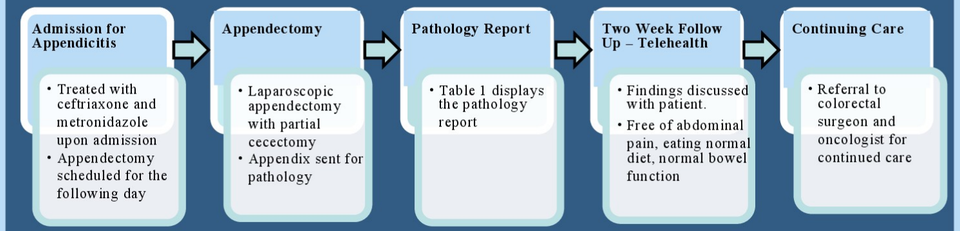
•Post-operatively, ceftriaxone and metronidazole continued, subcutaneous heparin started for deep vein thrombosis prophylaxis

•Discharged home that same day with prescriptions for oxycodone, amoxicillin/clavulanate, acetaminophen, and polyethene glycol

•Tumor markers CEA, CA19-9, and CA125 were ordered

•Referral to colorectal surgeon and oncologist was made for further staging, right hemicolectomy, and close follow up

Figure 1. Hospital Course



Discussion

•AA is a rare appendiceal neoplasm that most often clinically presents as acute appendicitis[1,4-6]

•Age greater than 50, family history of colon cancer, unexplained anemia, smoking history, and male gender are all risk factors[1]

•AA is typically diagnosed incidentally during pathologic examination, many patients require a secondary procedure[1,7-9]

•Regardless of nodal involvement, patients with AA should undergo oncologic resection with formal right hemicolectomy[1,7,8]

•In patients with node positive disease or distant metastasis, adjunctive chemotherapy with fluoropyrimidine and oxaliplatin is prescribed[9,12]

•Low grade adenocarcinoma without spread has excellent survivability[13,14]

•Higher grade tumors with KRAS mutations or a signet ring cell subtype tend to have poorer outcomes[13,14]

•Female gender has been linked to increased survivability even with less favorable histopathologic subtypes[13]

Conclusion

- Appendiceal adenocarcinoma is a rare appendiceal neoplasm that presents similarly to acute appendicitis
- Diagnosis requires histologic examination
- Right hemicolectomy is standard of care
- Advanced disease with nodal involvement or metastasis requires adjuvant chemotherapy
- Providers should be be cognizant of appendiceal neoplasms when evaluating patients with suspected acute appendicitis as it can affect management

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