

An Unusual Cause of Abdominal Pain

by Juan Echavarria, Alfredo Camero, Allan Parker

INTRODUCTION

PPrimary appendiceal cancer is a very rare entity, constituting less than 0.5% of all gastrointestinal neoplasms.¹ First reported by Beger in 1882, it is found in 0.9–1.4% of appendectomy specimens. The International Classification of Diseases for Oncology (ICD-O) divides the malignant tumors of appendix into five categories: “colonic type” adenocarcinoma, mucinous adenocarcinoma, signet ring cell carcinoma (SRC), goblet cell carcinoid/adenocarcinoid and malignant carcinoid/adenocarcinoid.² In a previous study, Uihlein and McDonald reported a distribution for malignant tumors as follows: 88.2% carcinoid, 8.3% malignant mucocele and 3.5% adenocarcinoma.³ In a recent study, McCusker et al. reported an age-adjusted incidence of 0.12 cases per 1,000,000 people per year for primary appendiceal carcinoma. Mucinous adenocarcinoma was the most common histological type reported followed by colonic adenocarcinoma and malignant carcinoid. Signet ring cell adenocarcinoma was the least common type, accounting for less than 5% of all appendiceal malignancies.⁴

Juan Echavarria, MD¹ Alfredo Camero, MD² Allan Parker, DO¹ ¹University of Texas Health Science Center at San Antonio, Department of Gastroenterology and Nutrition ²University of Texas Health Science Center at San Antonio, Department of Medicine

CASE REPORT

A 63 year-old Hispanic man with a past medical history of hypertension, gastroesophageal reflux (GERD), chronic constipation, anxiety, and benign prostatic hypertrophy (BPH) presented with a three month history of right lower quadrant abdominal pain, described as dull, intermittent, non-radiating and worsening over the last several weeks. His pain was not related to food intake or fasting, was not worse with movement, was not improved with defecation and improved with common over-the-counter analgesic medications. The patient denied weight loss, fever, bleeding and dysphagia. He had no significant family medical history.

Routine laboratory studies were all within normal limits. Diagnostic colonoscopy revealed an irregular, ulcerated mass in the appendiceal orifice, without other lesions (Figure 1). A computed tomography (CT) scan showed a thickened appendix measuring up to 18 mm at the base, with periappendiceal inflammatory stranding and numerous sub-centimeter ileocolic mesenteric lymph nodes (Figure 2). The biopsy revealed a poorly differentiated tumor with cells that possessed an extensive mucinous component and pale-staining cytoplasm, indicating an appendiceal adenocarcinoma, with signet ring cell carcinoma (SRC) features (Figure 3a, 3b). Esophagogastroduodenoscopy (EGD) and chest CT did not locate another primary site of malignancy, thus the diagnosis was consistent with a primary appendiceal SRC adenocarcinoma. The

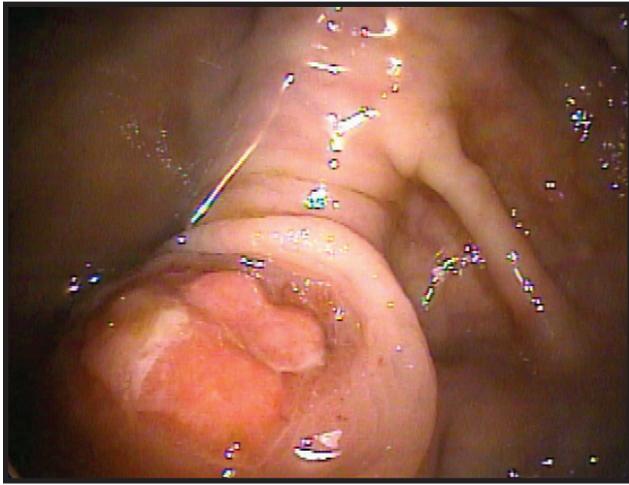


Figure 1. An endoscopic view of an irregular, ulcerated mass in the appendiceal orifice, without other lesions.

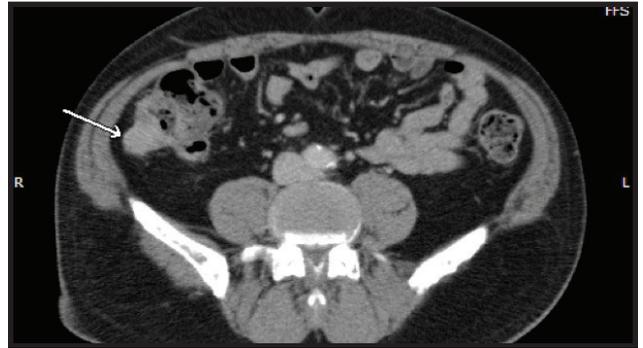


Figure 2. A computed tomography (CT) scan showed a thickened appendix measuring up to 18 mm at the base, with periappendiceal inflammatory stranding and numerous sub-centimeter ileocolic mesenteric lymph nodes.

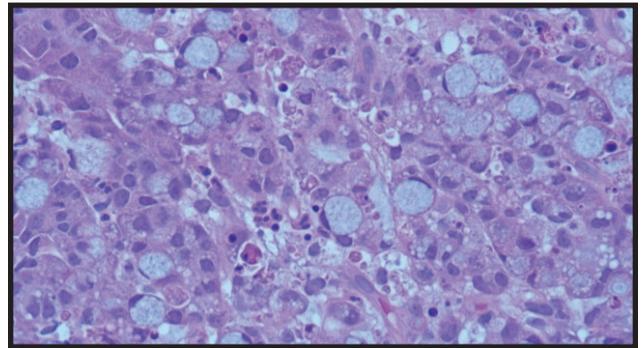
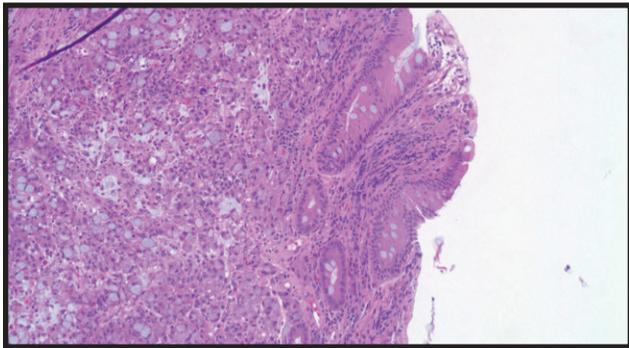


Figure 3a. and 3b. Biopsy showing a poorly differentiated tumor with cells with an extensive mucinous component and pale-staining cytoplasm, along with with SRC features. **3a** is low power and **3b** is higher power.

patient underwent a hand assisted laparoscopic right hemicolectomy (exploration of the abdomen showed no peritoneal implants), which later revealed a stage IIIc adenocarcinoma, with full-thickness penetration of muscularis propria into periappendiceal adipose tissue and extensive lymphovascular invasion (Figure 4). He was started on a modified FOLFOX6 regimen, and restaging CT chest/abdomen at 6 months showed no evidence metastatic disease.

Discussion

Primary appendiceal adenocarcinoma of the appendix is a rare event. In a 10 year retrospective study by Chen et al., the reported incidence was 0.28% identified from 2841 appendectomies, which is comparable to the incidence reported in similar studies.⁵ Several studies based on the Surveillance, Epidemiology and End

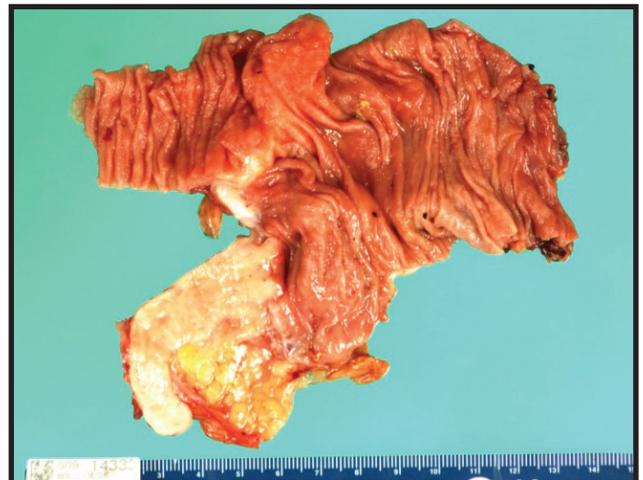


Figure 4. Gross specimen of the right hemi-colon along with the mass demonstrating full thickness invasion into periappendiceal adipose tissue.

A CASE REPORT

Results (SEER) registry, have shown that mucinous adenocarcinoma is the most prevalent type of all malignant tumor of the appendix, followed by colonic type adenocarcinoma, malignant carcinoid, goblet cell carcinoid and lastly SRC adenocarcinoma. Upon diagnosis, 63% patients with mucinous adenocarcinoma and 76% with SRC adenocarcinoma have extension to adjacent organs or metastatic disease. Similarly, 64% of patients with SRC adenocarcinoma have lymph node involvement compared to 26% of patients with mucinous adenocarcinoma.³

Given their overall low incidence, primary appendiceal adenocarcinomas are seldom suspected before surgery and less than half of the cases are diagnosed preoperatively.⁵ These neoplasms tend to not have early signs or pathognomonic symptoms since the majority of them present with appendicitis.⁶ As opposed to other more typical gastrointestinal luminal neoplasms, this type of tumor does not classically present with occult blood, anemia, weight loss or change in bowel function. Rather, patients typically present with acute appendicitis or an abdominal mass. They are also frequently detected as an incidental finding during exploration for another surgical disorder.

Endoscopic detection of appendiceal adenocarcinoma has been reported sparingly through case reports and limited case series in the medical literature. The general consensus regarding appendiceal adenocarcinoma is that a colonoscopy is inconsequential, as a normal colonoscopy does not predict the absence of an appendiceal adenocarcinoma. However, colonoscopy is useful in detecting synchronous colonic polyps that may have a higher risk for malignant transformation in this patient population. Furthermore, findings of a smooth, submucosal lesion in the cecum near the appendiceal orifice or free-flowing mucin from the appendiceal orifice should raise concern for appendiceal adenocarcinoma.⁷

SRC carcinomas are highly virulent tumors associated with a very poor prognosis. One study showed a 5 year survival rate of 28.6% in patients with SRC of the colon.⁸ The patient we described was diagnosed with a primary SRC adenocarcinoma of the appendix with regional lymph node involvement without evidence of other metastatic disease, and was treated with a right hemicolectomy and a modified FOLFOX-6 regimen. This case highlights the rarity of SRC adenocarcinoma of the appendix and its implications on prognosis and treatment once discovered. ■

References

1. Deans GT, Spence RJ. Neoplastic lesions of the appendix. *British Journal of Surgery*. 1995; 82: 299-306.
2. Percy C, Van Holten V, Muir C, editors. International Classification of Diseases for Oncology (ICD-O). 2nd edition. Geneva: World Health Organization, 1990.
3. McCusker ME, Cote TR, Clegg LX, Sobin LH. Primary Malignant Neoplasms of the Appendix - A Population-Based Study from the Surveillance, Epidemiology and End-Results Program, 1973–1998. *Cancer*. 2002; 94:3307-3312.
4. Uihlein A, McDonald JR. Primary carcinoma of the appendix resembling carcinoma of the colon. *Surg Gynecol Obstet*. 1943; 76:711-714.
5. Chen HT, Lee YM, Chou AS, Wu YK, Yin WY, Lee MC, Hsu YH. Primary Appendiceal Malignancy: A Clinicopathologic Study. *Kaohsiung J Med Sci*. 2006; 22(12): 618-25
6. Connor SJ, Hanna GB, Frizelle FA. Appendiceal Tumors – Retrospective Clinicopathologic Analysis of Appendiceal Tumors from 7,970 Appendectomies. *Dis Colon Rectum*. 1998; 41(1): 75-80.
7. Trivedi AN, Levine, EA, Mishra G. Adenocarcinoma of the Appendix Is Rarely Detected by Colonoscopy. *J Gastrointest Surg*. 2009; 13: 668-675.
8. Kang H, O’Connell JB, Maggard MA, Sack J, Ko CY. A 10-Year Outcome Evaluation of Mucinous and Signet-Ring Cell Carcinoma of the Colon and Rectum. *Dis Colon Rectum*. 2005; 48(6): 1161-68.

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