Appendiceal Adenocarcinoma Presenting as a Rectal Polyp

Erin Fitzgerald^c Lilian Chen^c Moises Guelrud^a Harmony Allison^a
Tao Zuo^b Yvelisse Suarez^b James Yoo^c

^aDepartment of Medicine, Tufts Medical Center, Boston, Mass., USA; ^bDepartment of Pathology, Tufts Medical Center, Boston, Mass., USA; ^cDepartment of Surgery, Tufts Medical Center, Boston, Mass., USA

Keywords

Appendiceal adenocarcinoma · Rectal polyp · Hematochezia · En bloc resection

Abstract

Appendiceal adenocarcinoma typically presents as an incidentally noted appendiceal mass, or with symptoms of right lower quadrant pain that can mimic appendicitis, but local involvement of adjacent organs is uncommon, particularly as the presenting sign. We report on a case of a primary appendiceal cancer initially diagnosed as a rectal polyp based on its appearance in the rectal lumen. The management of the patient was in keeping with standard practice for a rectal polyp, and the diagnosis of appendiceal adenocarcinoma was made intraoperatively. The operative strategy had to be adjusted due to this unexpected finding. Although there are published cases of appendiceal adenocarcinoma inducing intussusception and thus mimicking a cecal polyp, there are no reports in the literature describing invasion of the appendix through the rectal wall and thus mimicking a rectal polyp. The patient is a 75-year-old female who presented with spontaneous hematochezia and, on colonoscopy, was noted to have a rectal polyp that appeared to be located within a diverticulum. When endoscopic mucosal resection was not successful, she was referred to colorectal surgery for a low anterior resection. Preoperative imaging was notable for an enlarged appendix adjacent to the rectum. Intraoperatively, the appendix was found to be densely adherent to the right lateral rectal wall. An en bloc resection of the distal sigmoid colon, proximal rectum and appendix was performed, with pathology demonstrating appendiceal adenocarcinoma that invaded through the rectal wall. The prognosis in this type of malignancy weighs heavily on

whether or not perforation and spread throughout the peritoneal cavity have occurred. In this unusual presentation, an en bloc resection is required for a complete resection and to minimize the risk of peritoneal spread. Unusual appearing polyps do not always originate from the bowel wall. Abnormal radiographic findings adjacent to an area of gastrointestinal pathology may signify locally advanced disease from a surrounding organ that secondarily involves the gastrointestinal tract. These findings warrant further investigation prior to any intervention to ensure appropriate treatment.

© 2016 The Author(s)
Published by S. Karger AG, Basel

Background

Appendiceal cancer is a rare malignancy with an incidence of around 0.1 in 1,000,000 making up only 0.5% of all gastrointestinal malignancies. The tumor itself is not aggressive but has the potential for rupture and spread throughout the peritoneum in a phenomenon called peritoneal carcinomatosis which carries a poor prognosis and is debilitating both in terms of effect on quality of life and required treatment. Five year disease-specific survival is dependent on the degree of spread at diagnosis as well as histological subtype and ranges from 93% for carcinoid to 27% for signet ring cell type [1]. The disease-specific survival for mucinous adenocarcinoma is 58%. This low survival can be attributed to advanced stage at diagnosis as well as the occurrence of spread throughout the peritoneal cavity secondary to rupture of the smaller diameter and thinner-walled appendix [2, 3]. This cancer is most often diagnosed following appendectomy for suspected acute appendicitis [4].

Appendiceal cancer is distinctly different from colorectal cancer despite their close anatomic proximity and use of the same staging system. The average age at diagnosis in appendiceal cancer is much younger at 58 years compared to 72 years in colorectal cancer [2]. Another difference is related to the ability to prognosticate based on lymphatic spread. Although the presence of metastasis to lymph nodes does mean a shorter survival in appendiceal cancer, it is not the only indicator of poor prognosis, and 20–40% of patients with invasion of cancer through the muscular wall indicating T2 or greater disease will have no nodal involvement and yet still go on to die of their disease [5].

Surgical treatment traditionally involves a right hemicolectomy either as the primary operation when appendiceal malignancy is suspected or as a secondary procedure based on pathological diagnosis following appendectomy for presumed acute appendicitis. However, there is also evidence that a right colectomy does not confer a survival advantage, and thus practices are varied [6–8].

We describe a case of appendiceal adenocarcinoma that was initially diagnosed as a rectal polyp based on its location and appearance in the rectal lumen on colonoscopy.

Case Presentation

The patient is a 75-year-old female with a history of irritable bowel syndrome as well as a history of a previously normal colonoscopy who presented with spontaneous hematochezia. She was noted during colonoscopy to have a fungating rectal polyp. The polyp had multiple fingerlike projections and did not saline lift. Multiple biopsies were taken and pathology was consistent with fragments of tubulovillous adenoma and granulation tissue. A follow-up colonoscopy was then performed 2 months later which noted a single frond-like, villous, broad-based rectal polyp that appeared to be located within a diverticulum (fig. 1a),

and which had an area of distorted mucosal pattern consistent with malignancy (fig. 1b). A solution of saline and SPOT was injected into the submucosa but, similar to the previous procedure, the polyp did not lift. Endoscopic mucosal resection was attempted but was unsuccessful. At that point the patient was referred to Colorectal Surgery for a surgical evaluation. Based on the location of the polyp, the recommendation was for a low anterior resection.

A preoperative CT scan demonstrated a dilated appendix with its tip near the rectum which was of unclear significance at that time (fig. 2). During surgery, the appendix was found to be densely adherent to the right anterior rectal wall at the area that had been inked during colonoscopy. En bloc resection of the distal sigmoid colon and proximal rectum together with the appendix was performed (fig. 3). At that time, frozen sections of both the distal rectal resection margin as well as of the appendix were performed. The distal margin was negative for carcinoma. There was a mass at the tip of the appendix with mucin in the lumen which extended proximally but did not extend beyond the distal one half of the appendix. The proximal appendix was normal in appearance (fig. 4). The decision was made not to proceed with additional resection at that time but to await final pathology in order to guide further treatment. The patient's postoperative course was uncomplicated.

Pathology showed adenocarcinoma arising focally within an appendiceal tubulovillous adenoma with direct invasion into the rectum (fig. 5). The tumor size was 2.5 cm at its greatest dimension and occupied the distal one half of the appendix. Histologically it was grade 1. Microscopically, the tumor penetrated through the surface of the serosa with direct invasion into the rectal lumen. The proximal margin and the mesenteric margin were uninvolved by invasive carcinoma. There was no lympho-vascular invasion identified and no lymph node involvement after examination of 12 lymph nodes. Final pathological stage was pT4bN0.

Conclusions

Appendiceal cancers can be mistaken for other processes but often those are intraperitoneal, due to the risk of appendicitis and the potential for peritoneal carcinomatosis. As such, these processes are typically evaluated initially by CT scan, increasing the likelihood of identification of an appendiceal process. Intraluminal involvement of adjacent organs by appendiceal cancer is rare, especially as a presenting symptom, but this can occur due to local invasion or secondary to a contained perforation. When a colorectal polyp is identified with atypical features, it is important to consider other etiologies and additional radiographic imaging may be helpful. Local invasion of appendiceal cancer into adjacent organs requires an en bloc surgical resection for complete removal.

Statement of Ethics

Written informed consent was obtained from the patient's health care proxy for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Disclosure Statement

The authors declare that they have no competing interests.

References

- 1 Turaga KK, Pappas SG, Gamblin TC: Importance of histologic subtype in the staging of appendiceal tumors. *Ann Surg Oncol* 2012;19:1379–1385.
- 2 Marmor S, Portschi PR, Tuttle TM, Virnig BA: The rise in appendiceal cancer incidence: 2000–2009. *J Gastrointest Surg* 2015;19:743–750.
- 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 Misdraji J: Mucinous epithelial neoplasms of the appendix and pseudomyxoma peritonei. *Mod Pathol* 2015;28:S67–S79.
- 5 Bunni J, Arnold D, Shelley-Fraser G, Shabbir J: Multidisciplinary perspective on the management of appendiceal adenocarcinoma: case review of 10 patients from a university hospital and current considerations. *Clin Colorectal Cancer* 2015;14:58–62.
- 6 Nitecki SS, Wolff BG, Schlinkert R, Sarr MG: The natural history of surgically treated primary adenocarcinoma of the appendix. *Ann Surg* 1994;219:51–57.
- 7 Gonzalez-Moreno S, Sugarbaker PH: Right hemicolectomy does not confer a survival advantage in patients with mucinous carcinoma of the appendix and peritoneal seeding. *Br J Surg* 2004;91:304–311.
- 8 Proulx GM, Willett CG, Daley W, Shellito PC: Appendiceal carcinoma: patterns of failure following surgery and implications for adjuvant therapy. *J Surg Oncol* 1997;66:51–53.

Fig. 1. Endoscopic appearance of the appendiceal adenocarcinoma. **a** A single frond-like villous polyp was present in the rectal lumen and appeared to be located within a diverticulum. **b** The mucosal pattern appeared distorted, consistent with malignancy.

Fig. 2. Preoperative CT scan demonstrating a dilated appendix abutting the right side of the rectal wall.

Fig. 3. An en bloc resection of the appendix and rectum was performed. The appendix is densely adherent to the rectal wall along the right side and is dilated at its tip.

Fig. 4. Anatomic evaluation of the resected specimen. A mass lesion was identified in the tip of the appendix, with direct extension into the lumen of the rectum at the site of the presumed rectal polyp.

Fig. 5. Histological evaluation of the resected specimen. Direct invasion of the appendiceal adenocarcinoma into the rectal lumen was confirmed histologically.