Carcinoid Tumor of the Ileoanal Pouch in a Patient with Ulcerative Colitis

SONJA CHEN, MBBS; MURRAY RESNICK, MD, PhD; VICTOR PRICOLO, MD

ABSTRACT

Carcinoid tumors have been reported to occur in various locations, particularly in the gastrointestinal tract. The relationship between the development of carcinoids and ulcerative colitis has been an unclear and controversial one. The association of ulcerative colitis and the development of ileal-pouch carcinoids has not, however, been well documented. We report a case of carcinoid tumor arising in an ileoanal pouch and discuss its unique diagnostic and therapeutic considerations.

KEYWORDS: Carcinoid tumor, ulcerative colitis, ileoanal pouch

INTRODUCTION

Carcinoid tumors of the gastrointestinal tract are receiving renewed attention, due to advances in understanding of their epidemiology and changes in pathologic classification to better define their metastatic potential, overall behavior and prognosis.

Although ileal-carcinoid tumors are relatively common to our knowledge no carcinoid in an ileal pouch had been described. We hereby describe one such case, which presented unique diagnostic and therapeutic challenges.

CASE REPORT

An 81-year-old woman experienced crampy, lower abdominal pain for about a month before being seen by her primary care physician. Her history was significant for a restorative proctocolectomy, transanal mucosectomy, and hand-sewn, ileal-pouch anal anastomosis for ulcerative colitis in 1989 at a different institution. Her functional outcome had been poor, with anal stenosis, fecal incontinence and up to 20 bowel movements a day. She was otherwise healthy and still working at a department store. Abdominal CT demonstrated a 1.5 cm mass in the wall of her ileal pouch.

The patient was referred to our institution and underwent an ileoscopy of her pelvic pouch, with findings of an umbilicated submucosal lesion at 8 cm from the anal verge. The biopsy yielded a diagnosis of carcinoid. An octreotide scan revealed no evidence of any octreotide-avid lesion. The patient's serum chromogranin A level was within normal

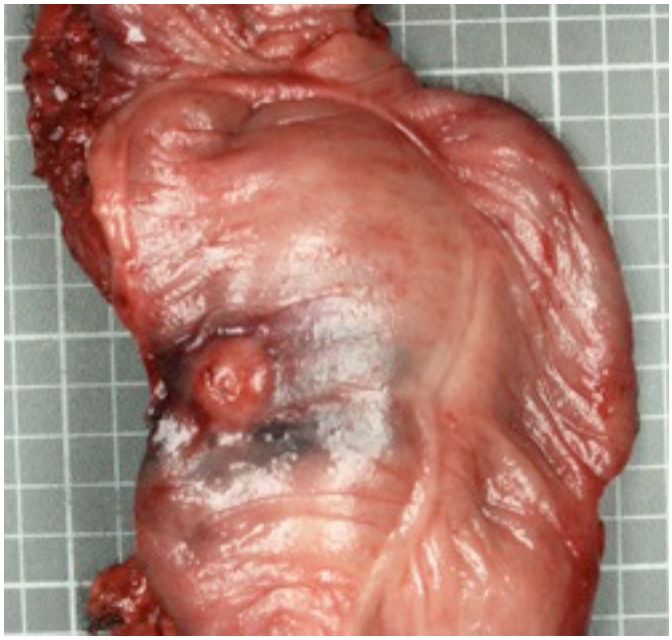

Figure 1. Gross photograph of umbilicated, pink to tan lesion with surrounding tattoo in the ileoanal pouch.

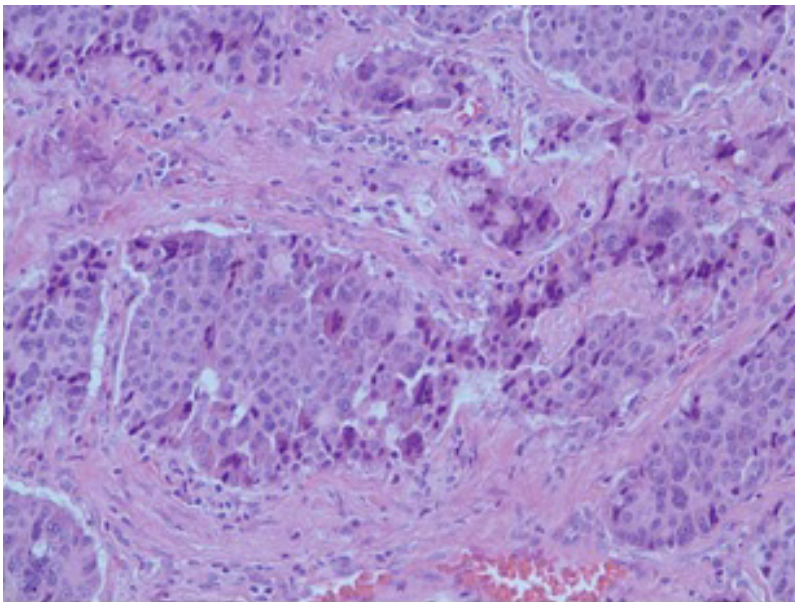

Figure 2. High-powered view demonstrating large nests of cells with the typical salt-and-pepper chromatin pattern characteristic of a neuroendocrine tumor (x 40).

range. In view of the patient's anal stenosis, a transanal excision was not technically feasible. In addition, in view of her poor quality of life with respect to fecal continence as well as the known metastatic potential of ileal carcinoids, the patient underwent a pouch excision with permanent ileostomy. She tolerated the procedure uneventfully and, six months postoperatively, she has adjusted well to her stoma.

Pathologic examination of the ileal pouch (Figure 1) demonstrated a 1.2 cm well-differentiated neuroendocrine tumor (Figure 2) with intratumoral lymphatic invasion. The tumor demonstrated a 2% proliferative index with Ki-67 as well as Chromogranin A positivity (Figure 3). The regional lymph nodes in the excised specimen were not involved by tumor. The final pathologic stage was pT2 N0 Mx.

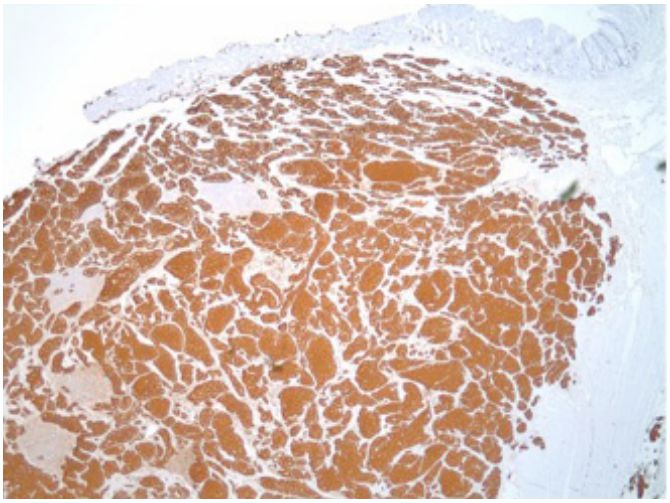

Figure 3. Staining immunopositivity for Chromogranin A (x 2).

DISCUSSION

Carcinoid tumors, or well-differentiated neuroendocrine tumors (NETs), most commonly arise in the gastrointestinal system. Their incidence is estimated to be approximately 1.5 cases per 100,000/year of the general population (i.e., approximately 2,500 cases/year in the United States). Nonetheless, they account for 13% to 34% of all tumors of the small bowel and 17% to 46% of all malignant tumors of the small bowel.[1]

Currently, about 22% of all NETs of the small bowel arise from the duodenum while the ileum remains the most frequent site of NETs in the small intestine (> 70%), where they tend to arise sporadically and are not associated with neurofibromatosis type 1 or MEN syndrome. With newer imaging and endoscopic modalities, NETs are being detected earlier than in the past. In the 1970s, advanced disease was present in 31.3% of patients at first presentation. This fell to 22% in the1980s and 1990s, to 18.5% in 2000-2004.[2]

Such findings beg the question of risk of metastatic disease. In this case, benign regional lymph nodes were noted. In general, approximately one third of patients exhibit regional nodal metastases only and another third show distant metastases.[3] The prevalence of distant metastases also increases with the size of the primary tumor. In published studies, the rate of metastases from tumors smaller than 1 cm was 2%; from tumors 1 to 2 cm, 50%; and from tumors larger than 2 cm, 80%.[4] However, five-year survival after the initial diagnosis of metastatic neuroendocrine tumor is approximately 75%.[5]

The association of ulcerative colitis and carcinoid tumors has been suggested, with up to 27 cases reported in the literature. However, most of these lesions have been documented in the rectum and are usually discovered incidentally. In ulcerative colitis patients, the incidence of these tumors arising in the jejunum and ileum is 0.67 per 100,000 per year [6], much less frequently than in the normal population. The association of ulcerative colitis and the development of ileal-pouch carcinoids, has not, however, been documented.

Other lesions which have been found to occur in the ileoanal pouch include, most commonly, adenocarcinoma, lymphoma and even squamous cell carcinoma. This case and these observations, in addition to the anal canal dysplasia-cancer risk, emphasize the need for continued endoscopic surveillance of the ileoanal pouch.

References

1. Hamilton SR, Aaltonen LA (Eds.): World Health Organization Classification of Tumours. Pathology and Genetics of Tumors of the Digestive System; IARC Press: Lyon 2000.
2. Scherübl H, Schwertner C, Steinberg J, Stölzel U, Pohl J, Dralle H, Klöppel G. Neuroendocrine tumors of the small bowels are on the rise: early tumors and their management. Z Gastroenterol. 2010;48(3):406-13.
3. Burke AP, Thomas RM, Elsayed AM, Sobin LH. Carcinoids of the jejunum and ileum: An immunohistochemical and clinicopathologic study of 167 cases. Cancer. 1997;79(6):1086-93.
4. Modlin IM, Kidd M, Latich I, Zikusoka MN, Shapiro, MD. Current status of gastrointestinal carcinoids. Gastroenterology. 2005;128(6):1717-51.
5. Strosberg J, Gardner N, Kvols L. Survival and prognostic factor analysis of 146 metastatic neuroendocrine tumors of the midgut. Neuroendocrinology. 2009;89(4):471-476.
6. Quinn P, Platell CF. Resolving microcarcinoids in ulcerative colitis: report of a case. Dis Colon Rectum. 2004;47(3):387-91.

Authors

Murray Resnick, MD, PhD, is a Professor of Pathology & Laboratory Medicine at the Warren Alpert Medical School of Brown University.

Victor Pricolo, MD, is a colorectal surgeon at the Rhode Island Hospital and The Miriam Hospital.

Sonja Chen, MBBS, is a PGY2 physician in the Department of Pathology, Rhode Island Hospital/ Warren Alpert School of Brown University.

Disclosures

The authors and/or their spouses/partners have no financial interests to disclose.

Correspondence

Murray Resnick, MD, PhD
Brown Alpert Medical School / Rhode Island Hospital,
593 Eddy Street, Providence RI 02903
401-444-4380
mresnick@lifespan.org