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

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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
range. In view of the patient’s anal stenosis, a transanal ex- 
icision 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 il-
estomy. 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 tu-
mors (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]. None-
theless, 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 fre-
quently 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 the 1980s and 1990s, to 18.5% in 2000-2004.2

Such findings beg the question of risk of metastatic dis-
ease. In this case, benign regional lymph nodes were noted. 
In general, approximately one third of patients exhibit re-

gional nodal metastases only and another third show dis-
tant metastases.3 The prevalence of distant metastases also 
increases with the size of the primary tumor. In published 


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