A 58-year-old female with history of recurrent gastrointestinal bleeding presented with three days of melena. The patient underwent colonoscopy in 2007 at an outside institution for hematochezia and Hemoglobin of 6.7 mg/dL. Gross blood was seen from the terminal ileum to the rectum, but no lesions were visualized. An upper endoscopy was performed in 2008, revealing gastritis. In 2009, the patient presented to our institution with occult blood in stool and underwent capsule endoscopy. A polypoid mass with ulcer was seen in the mid small bowel (Figure 1-Polypoid mass; Figure 2-Ulcer in ileum), but the patient was lost to follow up. She returned in 2010 with recurrent gastrointestinal bleed.

Retrograde double balloon enteroscopy revealed three sessile 6–8 mm submucosal nodules in the distal, mid, and proximal ileum (Figure 3-Submucosal nodules), a flat 1 cm ulcer in the mid ileum (Figure 4), and an umbilicated 1.5 cm polyp with central ulceration in the proximal ileum. Biopsies of all lesions revealed carcinoid tumor (Figure 5-H and E stain; Figure 6-Chromogranin immunostain).

Preoperatively, a CT scan of the abdomen revealed focal thickening of a short segment of small bowel and lymphadenopathy (Figure 7). An Octreotide scan revealed abnormal uptake in the pelvic mesentery at 24 hours (Figure 8). The serum chromogranin A level was normal.

The patient subsequently underwent surgery with resection of 100 cm of the small intestine, appendix, and surrounding lymph nodes. Six separate carcinoid tumors, all invading the muscularis propria, were identified. Three out of the twenty-two lymph nodes were found to be positive for disease. The resection margins were negative.

The overall incidence of carcinoid tumors in the United States is estimated to be 1–2/100,000 but the true incidence may be even higher, as tumors are indolent. Diarrhea and flushing are the hallmarks of carcinoid syndrome, however only 5–7% of small bowel carcinoids present with carcinoid syndrome. The
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Figure 2.
sensitivity of the diagnostic markers (24-hr urine 5-HIAA, serum chromogranin, or serum 5-hydroxytryptamine) diminishes in patients with nonmetastatic or asymptomatic carcinoid tumors. At presentation, many of these tumors are found to be metastatic to the lymph nodes or liver. A review of 145 patients with carcinoid syndrome found presenting symptoms to include uncharacteristic abdominal pain (50%), flushes (13%), and overt gastrointestinal hemorrhage (14%). At diagnosis, 44% of these patients had regional metastases and 40% had distant metastases. The 5-year survival with local disease is estimated to

Figure 3.

Figure 4.

Figure 5.

Figure 6.

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be 65% and 40% with distant metastases. Given the indolent disease course and good prognosis, these patients should be referred for surgical resection. Thus, the clinician needs to be wary of the diagnosis in order to find the disease at its earliest stages.

References