

Multiple carcinoids of the ileum: case reports

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Intestinal carcinoids are an uncommon neoplasm, and multiple carcinoids are extremely rare. Carcinoids of the ileum account for 15.4% of all gastrointestinal carcinoids¹ and are multicentric in 20%–30% of patients.² We present the first published report of patients with more than 40 carcinoids in the terminal ileum. In both patients, the carcinoids were associated with extraintestinal neoplasms. Our findings support the association of carcinoids with other neoplastic conditions.

Case 1

A 43-year-old woman underwent a laparoscopic left ovarian cystectomy for what proved to be an incompletely excised invasive mucinous carcinoma with a small focus of endometriosis. She subsequently underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy, peritoneal cytology and a lymphadenectomy. At laparotomy, multiple nodules along an extensive length of ileum were incidentally noted. Examination of a frozen section revealed a carcinoid neoplasm. A small bowel resection was performed, encompassing the affected portion of the bowel and an adjacent enlarged and hard lymph node in the small bowel mesentery (Fig. 1). The final pathology revealed more than 40 carcinoids in mucosa, submucosa, muscularis propria and serosa of the small bowel (Fig. 1). The histologic pattern was predominantly insular; however, rare trabecular and glandular patterns were also seen. Lymphatic invasion was identified in

the small bowel wall and the surrounding fat. The proximal and distal margins were negative for neoplasm, and 9 of 13 mesenteric lymph nodes were positive for metastatic carcinoid. The appendix was grossly and histologically unremarkable. The patient was disease-free on follow-up almost 3 years after her operation.

Case 2

A 77-year-old woman presented with a history of multiple emergency visits and admissions to the hospital for recurrent small bowel obstruction over a 2-year period. A

small bowel follow-through examination was normal. Abdominal ultrasound revealed a multilobulated terminal ileal mesenteric mass measuring 3 × 3.2 cm, likely representing a lymph node that was very vascular. A computed tomography scan of the abdomen and pelvis with contrast showed no evidence of small bowel obstruction, with focal cecal wall thickening and 2 large mesenteric masses measuring 3.5 × 3.4 cm and 1.7 × 1.9 cm. Colonoscopy revealed a sigmoid tubular adenoma, which was removed. Exploratory laparotomy findings included a normal cecum and multiple ileal lesions

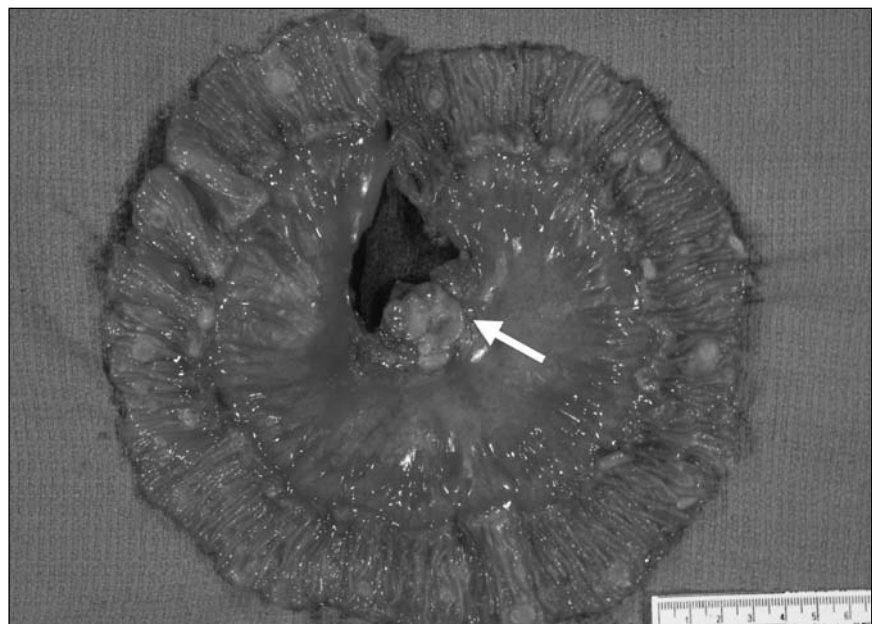


FIG. 1. Resected segment of ileum containing multiple carcinoids and a necrotic metastatic lymph node (arrow).

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visible on the serosal surface, one of which was almost completely obstructing the lumen. There were other ileal lesions that were intramural, firm and mobile and scattered along the small bowel segment. In addition, there was a 4.5-cm mass in the mesentery of the terminal ileal segment that contained the previously mentioned lesions. The segment of ileum containing the lesions and the mesenteric mass were successfully resected. The numerous lesions were reported as neuroendocrine (carcinoid) carcinoma with vascular, lymphatic and perineural invasion. Metastatic disease was identified in 6 of 13 regional lymph nodes retrieved.

Discussion

Among carcinoids, 70%–80% are asymptomatic and found incidentally at the time of operation for symptoms of bowel obstruction or during exploration of the small bowel in search of a primary neoplasm after distant metastases have been found.² Multicentricity has been reported as low as 2%–4%³ for rectal carcinoids and as high as 40% for small bowel carcinoids.⁴ The clinical significance of the multiplicity of small bowel carcinoids and their association with other neoplasms are not fully understood.

Yantiss and colleagues⁵ compared the clinical and pathological features and prognosis of 50 patients with solitary

carcinoids and 18 patients with multiple carcinoids of the ileum. At the time of diagnosis, patients with multiple carcinoids were significantly younger than patients with solitary lesions, there was a high association between multiple carcinoids and the carcinoid syndrome, and there was an association between multiplicity and venous invasion, but this relation was not statistically significant.

Another unusual observation was the frequent coexistence of a second primary malignant neoplasm of a different histologic type.⁶ Usually, this is a synchronous adenocarcinoma (most commonly in the large intestine), and it can occur in 10%–20% of patients with carcinoids. In Morgan and colleagues⁴ review of 135 patients with gastrointestinal carcinoids, associated malignant neoplasms occurred in 26% of patients.

With respect to therapy in both cases, the extent of resection necessary to encompass the disease process, along with its regional metastases, was of concern, especially in case 2, because of the proximity of a large metastatic deposit to the superior mesenteric artery. Nevertheless, the extensive resection was performed to rid the patients of all their visible disease. It is well recognized that operative resection is the mainstay of treatment of intestinal carcinoids.

Competing interests: None declared.

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Correction

The list of online case notes published in the April 2008 issue contained an error.¹ For the case note “Role of the endo-GIA stapler in transanal excision of rectal tumours,” the first author’s name was spelled incorrectly. The correct spelling is G. Montalto. *CJS* apologizes for this error and any inconvenience it may have caused.

Reference

1. Montalto G, Polinari U, Ausania F, et al. Role of the endo-GIA stapler in transanal excision of rectal tumours. *Can J Surg* 2008;51(2):155.