

# A rare occurrence of two synchronous carcinoid tumors in the same organ

Melanie Danielle Crispin, Andrew Busuttill, Chris Y. Lu

## ABSTRACT

**Introduction:** Carcinoid tumors are neuroendocrine cell tumors, most commonly found in the appendix and small bowel. Carcinoid tumors are often found together with another tumor of a different histological type. However, to find two synchronously occurring carcinoid tumors in one person is uncommon, and indeed to discover two synchronous carcinoid tumors in the one organ is extremely rare. **Case Report:** This case report describes a 57-year-old male presented with symptoms of a small bowel obstruction. After failing conservative management, he proceeded to undergo a laparotomy. At the time of operation, two distinct lesions were found in his jejunum which were the cause of his bowel obstruction. These were excised, and found to be synchronous carcinoid tumors of the small bowel. **Conclusion:** We describe a unique case of synchronous carcinoid tumors causing small bowel obstruction. Carcinoid tumors are a rare but important consideration when managing patients with small bowel obstructions. In addition, more research is required regarding the prognostic factors and treatment options for small bowel carcinoid

tumors. The clinical and prognostic significance of synchronous small bowel carcinoid tumors also requires further study.

**Keywords:** Cancer, Carcinoid, Neuroendocrine, Synchronous

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## INTRODUCTION

Carcinoid tumors are neuroendocrine cell tumors, most commonly found in the appendix and small bowel. Prevalence is higher in patients with MEN1 mutations [1]. Carcinoid tumors are not infrequently diagnosed in synchrony with cancers of another histological type, such as gastrointestinal adenocarcinoma, lung carcinoma or prostate carcinoma, with incidence ranging between 18–40% [2]. It is however, uncommon for one patient to have synchronous carcinoid tumors in two organs, thus to discover synchronous carcinoid tumors in the same organ is extremely rare. There are isolated reports of this occurring in the colon, rectum, and lung but at time of publication there are no reports of this occurring symptomatically in the small bowel [3–6]. Tse et al. [7] describes one case where two ileal carcinoid tumors were discovered incidentally following right hemicolectomy for colonic adenocarcinoma. We present a unique case of symptomatic, synchronous carcinoid tumors of the small bowel.

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## CASE REPORT

A 57-year-old male presented with a three-day history of cramping central-abdominal pain localizing to the right iliac fossa, associated with nausea and vomiting. The patient had a bowel movement on the day of presentation, but was not passing flatus. His past history was significant for ischemic heart disease and right hip arthroplasty. He was an ex-smoker and drank 30-standard-drinks per week. He had not previously undergone abdominal surgery.

On examination he appeared well, was afebrile, and hemodynamically stable. His abdomen was soft, but tender over the right iliac fossa with voluntary guarding. Bowel sounds were present. There were no hernias, and no abdominal scars. Blood tests revealed an elevated white cell count and mildly elevated gamma glutaminase; other liver function tests and lipase were normal.

Due to the undifferentiated nature of his presentation, computed tomography (CT) scan was performed, which showed a moderate small bowel obstruction with feces sign. A transition point was identified in the lower jejunum to the right of the midline, just below the umbilicus. (Figure 1). Surgical decision was made to proceed to laparotomy.

At laparotomy two five-millimeter nodular strictures, 10 cm apart, were identified at the distal jejunum transition point with fecalization in-between, indicating chronic closed loop small bowel obstruction. A palpable two centimeter mesenteric lymph node was also identified. Twenty centimeters of small bowel was resected en bloc with the palpable lymph node. An end to side anastomosis was created.

Histopathology revealed two synchronous carcinoid tumors excised with clear margins, measuring twelve millimeters and nine millimeters in diameter, infiltrating to serosa (Figures 2 and 3). Immunohistochemical stains confirmed neuroendocrine origin; positive for chromogranin, synaptophysin and neuron specific enlase (NSE). Ki-67 proliferative indices of the tumors were between 4–6% indicating grade 2 tumors. The lymph node was involved with metastatic tumor. Post-operative positron emission tomography (PET) scanning revealed two peritoneal nodal metastases posterior to the anastomosis site.

## DISCUSSION

Jejunal carcinoid tumors can present with small intestinal obstruction, ischemia, abdominal pain, distension, weight loss, nausea and vomiting [8]. The mean age of diagnosis for jejunal carcinoids is 65.4 years in both men and women. Surgical resection of the primary tumor and affected lymph nodes by mesenteric wedge resection remains gold-standard for gastrointestinal carcinoid tumors, offering approximately 20% of patients a chance of cure. Life-long surveillance is recommended [8].

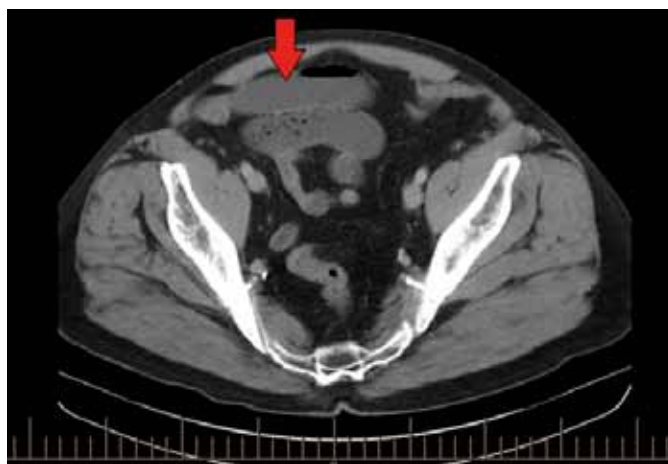


Figure 1: Axial computed tomography image demonstrating a closed loop small bowel obstruction (red arrow).

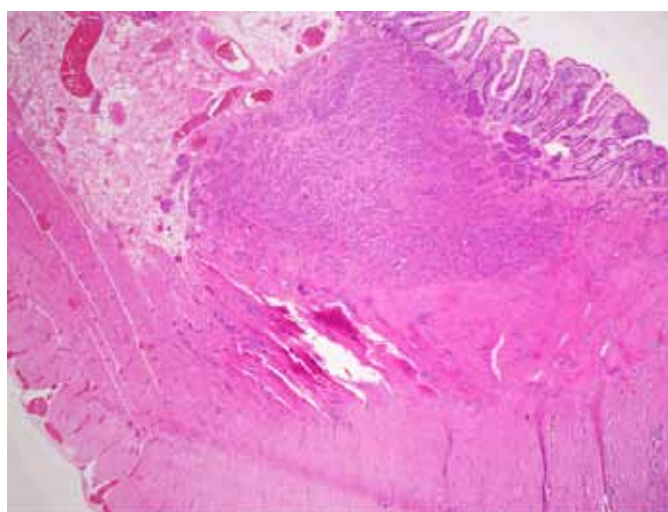


Figure 2: Histopathology slide of tumor one displaying typical neuroendocrine (carcinoid) tumor architectural patterns with cords and trabeculae of cells which comprise round nuclei with a salt-and-pepper nuclear chromatin pattern (H&E stain, x100).

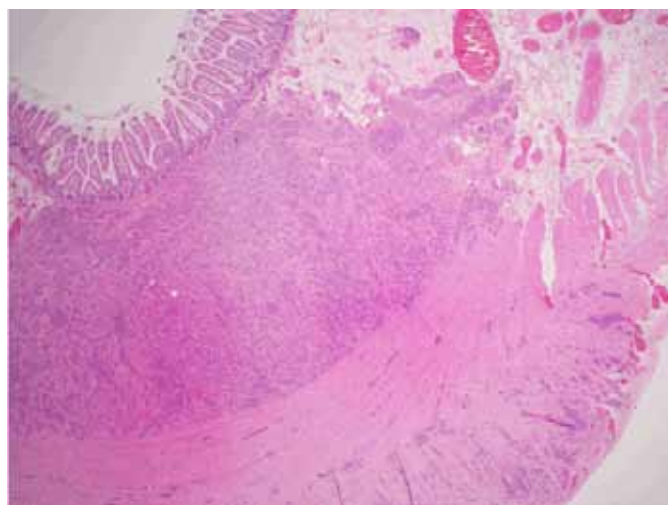


Figure 3: Histopathology slide of tumor two. This second tumor mass was found to lie 115 mm from the first tumor, and like the first tumor, showed transmurial infiltration by nests and cords of tumor cells with typical neuroendocrine nuclear features (H&E stain, x100).

Small bowel carcinoid tumors are classified using the TNM staging system. Our patient unfortunately had Stage IV disease [9]. Jejunal carcinoids commonly metastasize to lymph nodes and to the liver; 58–64% of patients have metastases at time of diagnosis. It is possible that our patient had a single primary jejunal carcinoid tumor which had metastasized to the small bowel and peritoneum, rather than synchronous jejunal tumors.

If hepatic detoxification of serotonin is compromised by metastases, carcinoids have the potential to generate carcinoid syndrome via argentaffin-positive, substance P-containing and serotonin-producing EC-cells. This occurs in approximately 35% of patients [8].

Carcinoid tumors of the appendix and rectum are generally slow-growing and amenable to surgical excision. Carcinoid tumors of the small bowel tend to metastasize early, and carry a poorer prognosis. Prognostic factors include tumor site and burden, presence of carcinoid syndrome, carcinoid heart disease, and high concentrations of tumor markers urinary 5-HIAA and plasma chromogranin A [8]. Certain antigens have been identified; these require more research regarding their importance. An octreotide trial to treat tumor progression demonstrated no difference in survival. Patients with inoperable liver metastases have a five-year survival rate of 50%; those with inoperable mesenteric and liver metastases have a five-year survival rate of 40%.

## CONCLUSION

More research is needed into the prognostic factors and treatment options for small bowel carcinoid tumors. The clinical and prognostic significance of synchronous small bowel carcinoid tumors also requires further study.

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## Author Contributions

Melanie Crispin – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Andrew Busittil – Substantial contributions to conception and design, Acquisition of data, Revising the article critically for important intellectual content, Final approval of the version to be published

Chris Lu – Substantial contributions to conception and design, Acquisition of data, Revising the article critically for important intellectual content, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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