Case in Point

Acute Obstructive Manifestation of a Jejunal Carcinoid

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This patient reported no herald symptoms of a carcinoid tumor, yet such a tumor was found when a pepper shell lodged in his intestine.

nteric obstruction is a mechanical or functional impeding of the normal transit of products of digestion. It may involve any level distal to the duodenum and represents a medical emergency. It is usually managed surgically. The major causes include postoperative adhesions, Crohn disease, tumor, and intestinal hernia. Neoplasms, either benign or malignant (including carcinoid), also may result in intestinal obstruction.^{1,2}

The following report describes a case in which a malignant carcinoid presented with acute obstructive signs and symptoms. These signs and symptoms were initiated by an entrapped pepper shell in the partial jejunal narrowing caused by the tumor.

INITIAL EXAM

A 77-year-old, white man presented to the emergency department with progressive nausea and vomiting of one day's duration. Pertinent physical examination findings included early signs of dehydration; a soft, distended, tympanitic abdomen; and hypoactive bowel sounds. He reported no history of inflammatory bowel disease or previous abdominal surgery and no symptoms leading up to his progressive nausea and vomiting.

Guaiac test results were negative, although laboratory data suggested signs of prerenal failure. With that clinical impression, a computed tomography (CT) scan of the abdomen was performed, revealing a nonspecific, small bowel obstruction below the level of the pancreas.

TREATMENT COURSE

The patient underwent emergency exploratory laparatomy and segmental bowel resection. No intraoperative frozen section evaluation was performed. The resected, 39-cm segment of small and large intestine exhibited grey-red serosal discoloration, with a firm and somewhat narrowed small bowel portion that measured 3.6 cm, located about 23.4 cm proximal to the ileocecal valve and 7.6 cm from the proximal resection margin. On opening, the intestine was found to contain a whole, intact, and fairly preserved pepper shell firmly entrapped in the partial enteric narrowing, which had only a 1.3-cm luminal circumferential dimension. The small bowel wall in this area was thickened to 7 mm and appeared solid, pale tan, and fibrotic (Figure 1). The rest of the small and large intestine were normal, with luminal circumferences

of 6.3 and 7.5 cm, respectively, and a thin (2- to 3-mm), soft, and pliable bowel wall.

Microscopically, the narrowed enteric wall revealed an ulcerated, poorly circumscribed, poorly demarcated, fibrotic, and transmurally invasive carcinoid that showed a classic insular growth pattern (Figure 2). Immunostaining results were positive for synaptophysin, chromogranin, and serotonin but negative for calcitonin, somatostatin,³ and epithelial membrane antigen. The serosal membrane was focally infiltrated but no regional lymph node showed metastasis.

The patient tolerated the surgical procedure well, and his postoperative course was unremarkable. Follow-up evaluations showed no recurrence of the carcinoid tumor at five years.

ABOUT THE CONDITION

A review of medical literature reveals that a variety of artifacts, such as coins, magnets, various materials used or inserted during surgical procedures, and a variety of foods have caused intestinal obstruction.^{4–10} Bezoars also have been associated with small bowel obstruction.

The term "bezoar" refers to a concretion of myriad compositions that are sometimes found in the stomach or intestines and are classifiable

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into trichobezoar (hair), phytobezoar (fruit and vegetable fibers), or trichophytobezoar (a mixture of hair and fruit and vegetable fibers). It has been suggested that phytobezoars be considered preoperatively as a cause of obstruction in patients with previous ulcer surgery.2 Reported bezoar treatment includes enzymatic therapy, milking of a phytobezoar to the caecum, emergency surgical exploration with bezoar removal, gastrotomy or gastrotomy with bezoar removal, and wedge resection of the gastric ulcer when present. Careful assessment for other phytobezoars also is important for prevention and management of phytobezoars.4-10

Although a pepper shell has not previously been associated with obstructive enteric carcinoid tumor, there have been two reports of bezoars associated with such tumors.^{4,5} Based on these reports, radiographic imaging may show intraluminal hydroaeric levels, hyperdense mobile filling defect, and a more distal fixed mural mass. None of these characteristics were observed in our patient.

Carcinoid tumors

Carcinoid tumors are slowly growing malignant neoplasms that usually follow an indolent clinical course. About 60% of such tumors are localized in the gastrointestinal tract, where they display late manifestation as a mass lesion, possibly with liver or regional lymph node metastasis, or a group of symptoms—encompassing flushing, diarrhea, abdominal pain, profuse sweating, telangiectasias, and heart disease—that result from the systemic effects of the tumor (referred to as the carcinoid syndrome).^{6,11–14}

Carcinoid tumors are not uncommon. There are 1.5 cases per 100,000 people in the United States per year or 2,500 new cases per year. They usually present in patients aged 50 to



Figure 1. An intact whole pepper shell entrapped in a jejunal narrowing due to a carcinoid tumor.

70 years and may secrete several neuroendocrine mediators, which results in the carcinoid syndrome. These tumor types are located in the appendix in 30% to 45% of cases, the ileum in 10% to 20% of cases, the duodenum in 3% of cases, and the stomach in 3% of cases. Extra-intestinal locations include the bronchus (which is involved in 20% to 30% of cases) and, on rare occasions, the ovary, kidney, or breast.^{11–14}

About one third of small bowel carcinoids are located near the ileocecal valve. These tumors commonly metastasize to the liver, cause cardiac and peritoneal fibrosis, and produce the classic carcinoid syndrome. When localized, these tumors have a 65% five-year survival rate, but when the tumor has metastasized, this rate drops to 35%. Jejunal carcinoid tumors are rare and have a relatively high rate of transmural invasion and aggressive clinical behavior.^{11–14}

Diagnosis involves analysis of urinary 5-hydroxyindoleacetic acid (5-HIAA) levels, which are usually higher than 100 mg/dL when a carcinoid tumor is present (reference range, 2 to 10 mg/dL). Two of every 10 patients with carcinoid tumors have normal urinary 5-HIAA levels, however. Markedly elevated fasting whole blood concentrations of serotonin also could indicate a carci-

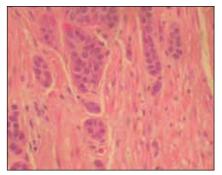


Figure 2. A classic carcinoid infiltrates the muscularis propria of the jejunal wall (hematoxylin eosin stain x 400).

noid tumor.^{11–14} In one study, patients without a carcinoid tumor exhibited serotonin concentrations of 71 to 310 ng/mL, while 10 patients with such a tumor exhibited concentrations between 790 and 4,500 ng/mL.¹⁴ Other diagnostic tests include epinephrine challenge and pentagastrin challenge. Localization can be achieved by CT of the abdomen, which may be performed to search for a tumor, liver metastasis, mesenteric stranding, and mesenteric lymph node enlargement. An indium 111 octreotide scan is equally effective for this purpose.^{11–14}

Treatment modalities include symptomatic regimens, surgery, interferon alfa, hepatic artery embolization, the somatostatin analogue octreotide, and 111-In pentetreotide radiotherapy. Carcinoid crisis—which encompasses the sudden onset of hypotension, flushing, and wheezing; is brought about by stress, palpation, or surgery; and can be exacerbated by vasopressors—is managed by administering octreotide and intravenous fluids to increase vascular volume.^{4,5,12,13}

IN SUMMARY

This case highlights an interesting association between a malignant carcinoid tumor, manifesting with acute obstructive signs and symptoms, and a pepper shell entrapped in the partially narrowed jejunum caused by the tumor. Although it is relatively uncommon for a carcinoid tumor to present with obstructive manifestation, in the absence of any obvious cause of a bowel obstruction, clinicians should suspect and evaluate for this entity.

Author disclosures

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