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Synchronous Medullary Carcinomas of the Small Bowel Presenting as Recurrent Small Bowel Obstruction

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Received: 30 August 2019; **Accepted:** 17 September 2019; **Published:** 19 September 2019

Abstract

Small bowel neoplasms of any type are rare, and fewer than 2% are malignant. There are numerous risk factors associated with small bowel carcinomas, but symptoms are vague making diagnosis difficult. Medullary carcinoma of the small bowel is even more unusual with few published articles on its presentations and treatment. Given the rarity of small bowel medullary carcinoma, the treating physician may not be familiar with the options for workup and treatment for this problem.

We describe the case of a 77 year old female with celiac disease who presented to the Emergency Department with small bowel obstruction. A small bowel radiologic examination demonstrated dilated loops of proximal small bowel with contrast traversing to decompressed distal small bowel. She underwent exploratory laparotomy where a strictured portion of the jejunum with an adjacent mass was identified. A second mass was noted distally, in the small bowel. Resection of both masses was performed, and pathology demonstrated these were separate primary medullary carcinomas.

Small bowel medullary carcinoma is a rare neoplasm and therefore treatment is not standardized. Resection appears to be the mainstay of treatment, especially for symptomatic patients. Awareness of this pathology and a high index of suspicion in patients with major risk factors, such as celiac disease presenting with obstructive symptoms, is required for diagnosis. Further studies to understand the molecular basis and natural progression of the disease is required to optimize management strategies.

Keywords: Medullary adenocarcinoma; Celiac; Small bowel; Synchronous

Abbreviations

FAP: Familial Adenomatous Polyposis; HNCC: Hereditary Non-polyposis Colorectal Cancer; LFT: Liver Function Test; CT: Computed Tomography; NPO: Nil Per Os; SBFT: Small Bowel Follow Through; POD: Post-operative Day

Introduction

Small bowel neoplasms are a rare subset of gastrointestinal neoplasms, of which fewer than 2% are malignant [1]. Thirty to fifty percent of malignant small bowel neoplasms are adenocarcinoma, making it the most common type of small bowel malignancy, followed by neuroendocrine tumors, lymphoma, and stromal tumors [2]. Many risk factors are associated with small bowel carcinomas, including familial adenomatous polyposis (FAP), hereditary nonpolyposis colorectal cancer (HNPCC), Crohn's disease, celiac sprue and Peutz-Jeghers syndrome [2]. The Kras mutation is commonly found in small bowel cancers and microsatellite instability is noted in tumors associated with celiac disease.

Symptoms of small bowel tumors are vague and non-specific and may be ascribed to a patient's previously known intestinal disease making diagnosis difficult. Benign masses are more likely to present with obstructive signs due to intussusception, whereas malignant tumors are more likely to cause pain and weight loss. Obstruction can be a presenting symptom in malignant tumors but it is more likely due to tumor infiltration or adhesions [1].

Medullary carcinoma of the small bowel is an exceedingly rare diagnosis with few published reports [3]. The diagnosis of medullary carcinoma is based on pathology and, given its rarity, there is no consensus on a standard care for treatment [3]. Surgical resection of the tumor appears to be the most appropriate and common initial treatment especially when symptomatic; however, the role for surveillance only or adjuvant therapies are unclear.

We describe the case of a patient admitted to our hospital with obstructive symptoms and celiac disease found to have two separate medullary carcinomas of the jejunum in close proximity.

Case Report

The patient is a 77-year old female with a history of Celiac disease, diverticulosis, hypothyroidism, Raynaud's disease, and surgical history of tonsillectomy and adenoidectomy, who presented for two days of an intermittent sharp abdominal pain in the periumbilical region. The pain was associated with poor appetite but without nausea or vomiting. She was passing flatus but had not had a bowel movement for two days. She had been seen for the same symptoms two months prior, when a computed tomography (CT) scan of the abdomen showed thickening of the distal duodenum and dilatation of the jejunum. At that time her symptoms resolved and she was discharged to home.

Upon further questioning it was noted that the patient had been seen in multiple emergency departments over the past two years with similar symptoms CT scans suggesting small bowel obstruction; however, she was discharged each time as she had return of bowel function and resolution of symptoms. Her laboratory tests were unremarkable, including liver function tests in the normal ranges. Her diagnosis of celiac disease was made by endoscopic biopsy many years prior. Upper endoscopy and colonoscopies since the onset of these symptoms were unremarkable.

At our hospital, she underwent a CT scan of the abdomen and pelvis, which revealed moderate distention of the proximal and mid small bowel with diameters measuring up to 5.3 cm. Transition in caliber in the right posterior pelvis at the level of the mid sacrum was seen, and the colon was decompressed. A small bowel follow through showed dilated loops of small bowel in the left upper and lower quadrants, with water-soluble contrast traversing these segments into decompressed distal small bowel within 1 hour 35 minutes. Given her symptoms and no previous abdominal operations, she was made *Nil Per Os* with plans for an exploratory laparotomy.

Intra-operatively, there was a strictured area and palpable intraluminal mass in the jejunum roughly 45 cm from the ligament of Treitz (Figure 1). This strictured area was resected. A second intra-luminal mass was palpated distal to the area of resection. This was resected as well, and the remaining small bowel was anastomosed. No other gross areas of abnormalities were noted in the small bowel, stomach or colon, nor was there any evidence of distant metastasis. Post-operatively, the patient did well and was discharged on the fourth postoperative day, with return of bowel function and tolerating a normal diet.

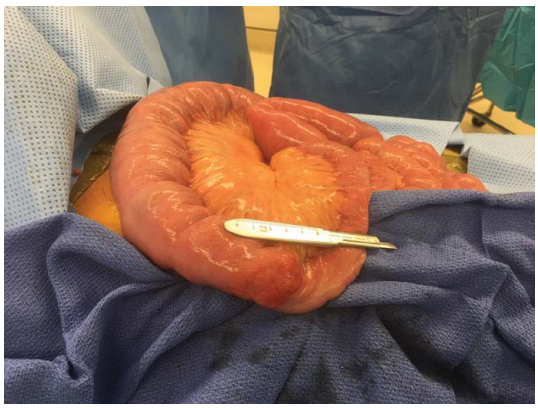

Figure 1: An intra-operative image of the palpable mass noted at the area of transition causing partial obstruction. Scalpel handle is in place to provide size reference.

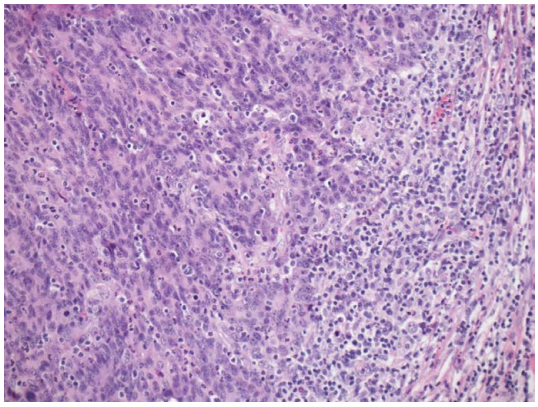

Figure 2: Pathology revealing a malignant infiltrate of cells which appears circumscribed with a pushing border, surrounded by abundant lymphocytes. Numerous tumor infiltrating lymphocytes are also noted.

The first resection showed a 3.7 cm poorly differentiated carcinoma with medullary features (Figure 2). The carcinoma was noted to be invading through muscularis propria with thirteen nodes negative for carcinoma. Pathological staging was pT3N0M0. The second resection showed a 2.2 cm poorly differentiated carcinoma with medullary features as well as adjacent tubular adenoma with intramucosal adenocarcinoma. This was invading into submucosa and 4 lymph nodes were negative for carcinoma. Pathological grouping was pT1bN0M0. Based on the histology, the tumors were consistent with 2 separate primary carcinomas. Immunohistochemical staining showed loss of MLH1 (Figure 3) and PMS2. Additional immunostains including synaptophysin, chromogranin, CDX2, and cytokeratin 20 were all negative making other tumor types such as neuroendocrine carcinoma and adenocarcinoma unlikely.

On follow up, the patient had no recurrence of symptoms. She was seen by the gastroenterology service and underwent Capsule Endoscopy which showed no other masses. She was evaluated by the oncology service and no further treatment was recommended.

Discussion

Medullary small bowel carcinoma is a rare malignant neoplasm of the small bowel with very little published literature [3]. One case series by Brcic et al. included three patients found to have small bowel medullary carcinoma. Interestingly, two of these patients were known to have celiac disease [4]. Celiac disease is known to be a risk factor for small bowel malignancies, however is typically associated with lymphoma and adenocarcinoma [5].

We found no literature describing a patient with two synchronous primary medullary carcinomas. A case report from 1989 by R.J Straker et. al discusses a patient found to have two synchronous adenocarcinomas of the jejunum within 8.5 cm of each other [5]. This leads one to question whether celiac patients are at increased risk for synchronous small bowel cancers compared to the general population. No information could be found to suggest that medullary carcinoma is associated with higher rate of synchronous malignancies.

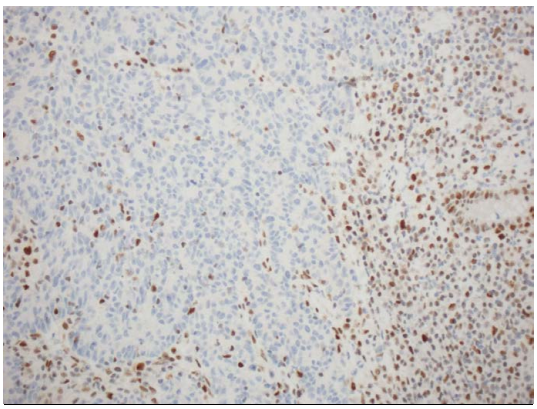

Figure 3: Pathology slide as in Fig 2 with MLH1 staining showing loss of expression.

The treatment for medullary carcinoma of the small bowel is not currently established. For symptomatic lesions, the mainstay in an eligible candidate should be surgical resection. Given its rarity, it is unclear what further evaluation should be done for a patient with medullary carcinoma of the small bowel especially with respect to further imaging and any adjuvant therapy. Our patient's post-operative workup showed no signs of metastatic disease, and no further treatment was administered. Further studies are indicated to determine the relationship of medullary carcinoma of the small bowel and celiac disease.

Conclusions

In patients with medullary small bowel carcinoma, it is imperative to perform a complete history, physical, and a thorough diagnostic evaluation. Presenting symptoms may be vague, and in patients with other established diagnoses such as celiac disease, they may be easy to overlook. While small bowel neoplasm, and medullary carcinoma of the small bowel in particular, is uncommon, it must remain part of the differential diagnosis with small bowel obstruction in the setting of celiac disease. Surgical resection of the tumor appears to be the most appropriate and common initial treatment especially when symptomatic; however, the role for surveillance only or adjuvant therapies are unclear.

Contributors

Scott Kurtzman MD. FACS., David Aughton MD., and Priyanka Shetty MD.

Special thanks for Miss Tiffany Zhang's manuscript proof reading.

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