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CASE REPORT

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Carcinoid tumor causing ileocecal intussusception in an adult patient

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ABSTRACT

Introduction: Little is known about adult intussusception, but current evidence suggests that malignancy, polyps, and diverticula are usual etiologies. We present a case of adult ileocecal intussusception secondary to carcinoid tumor.

Case Presentation: A 53-year-old African American male presented with hematochezia and non-radiating constant left upper quadrant pain accompanied by nausea and vomiting. CT of the pelvis demonstrated a pathognomic 'target' sign, consistent with ileocecal intussusception and early small bowel obstruction. Two years prior to this current presentation, the patient had experienced an episode of hematochezia for which he underwent colonoscopy and polypectomy, with subsequent pathology results negative for colon cancer. He denies diarrhea, constipation, weight loss, decreased appetite or skin flushing. Due to persistent symptoms of bowel obstruction, he underwent exploratory laparotomy. During the surgery a white-colored, chalky mass indicative of penetrating tumor was noted 13 cm proximal to the ileocecal valve. An extended right hemicolectomy followed the discovery of the mass. Pathology showed a well-differentiated neuroendocrine tumor consistent with carcinoid tumor. Evaluation for metastatic disease using 5-HIAA and chromogranin A was unremarkable, and the resection of the right colon carcinoid tumor was felt to be curative.

Conclusion: It is uncommon for adults to present with intussusception; in such cases, malignancy should be ruled out as an underlying cause. Carcinoid should be listed among the other secondary causes, which include inflammatory bowel disease, diverticulitis, polyps, scar tissue, adhesions, and lipomas.

Abbreviation: CT (Computer tomography), 5-HIAA (5-hydroxyindole acetic acid), NCCN (National Comprehensive Cancer Network)

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KEYWORDS
Intussusception; carcinoid tumor; hematochezia; small bowel obstruction

1. Introduction

Intussusception is defined as the telescoping of the proximal region of the intestine into the distal region. Intussusceptions are much more common benign entities in the pediatric population accounting for 90% of cases, whereas intussusception is a rare presentation in adults, comprising 5–10% of all intussusceptions [1]. Interestingly, only 1% of all adults' intussusceptions are found to cause small bowel obstruction, whereas the other 5% is of unknown etiology and could be due to secondary causes, most likely from adhesions [2]. One series identified the most common causes of adult intussusception as malignancy, polyps, or diverticula [3].

Intussusception presents clinically with intermittent, colicky abdominal pain, accompanied with nausea and vomiting [4]. Up to 63% cases of adult intussusception were tumor-related, none of which were carcinoid tumor [5]. Most of the malignant cases, 48%,were related to colo-colonic intussusception, with 17% enteric intussusception [6]. To date, there have been few reports of ileocolonic intussusception due to carcinoid tumor [7]. We present an interesting case of acute small bowel obstruction due to ileocolonic intussusception caused by a terminal ileal carcinoid tumor.

2. Presenting concerns

A 53-year old African American male presented to the emergency department with abdominal pain, nausea, vomiting, and bright red blood per rectum. The day prior to presenting to the ED, he had multiple episodes of bloody bowel movements increasing in magnitude accompanied by right lower quadrant pain that was constant and non-radiating.

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3. Clinical findings

The patient had experienced similar symptoms several years ago, but the source of bleeding was not identified. Upper endoscopy, colonoscopy and capsule endoscopy were non-diagnostic. The patient had an open appendectomy 34 years ago, and had polyps of unknown histology removed endoscopically four years ago from outside hospital.

Upon arrival to the emergency department, his vital signs were within normal limits. Hemoglobin was 14.7 g/dL. His physical examination was remarkable for abdominal distention and right lower quadrant abdominal tenderness with no rebound, guarding or rigidity.

4. Diagnostic assessments

Computed tomography (CT) abdomen/pelvis revealed ileocecal intussusception with an intraluminal hyperdensity in the intussusception concerning for a neoplasm (Figure 1). Segments of small bowel proximal to the intussusception were mildly dilated. There was no sign of metastases.

5. Therapeutic interventions

Gastroenterology was consulted for colonoscopy decompression of the intussusception. However, due to persistent nausea and vomiting and the inability to tolerate bowel prep, colonoscopy was deferred. Arrangements were made for laparotomy with reduction of the intussusception.

During surgery, the patient was found to have an ileocecal intussusception with partial small bowel obstruction. A right hemi-colectomy was performed with primary anastomosis. The lead point of the intussusception originated in the ileum. Resection of the lead point led to the discovery of a whitish, chalky area with a mass underneath, consistent with a tumor invading the serosa (Figures 2 and 3). Pathology showed a 1.7 × 1.7 x 1.3 cm low grade well-differentiated neuroendocrine tumor (carcinoid tumor) that was found to invade subserosal tissues without involvement of visceral peritoneum (Figures 4 and 5). In addition, 3/15 lymph nodes were positive for tumor (T3N1).

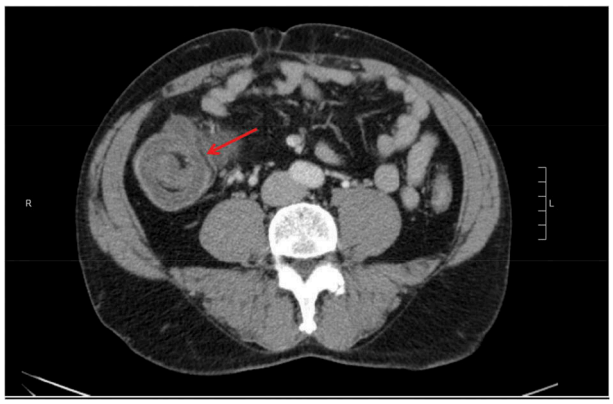

Figure 1. Axial CT of abdomen/pelvis. Appendiceal-colonic intussusception with the pathognomic 'target sign' as shown by red arrow.

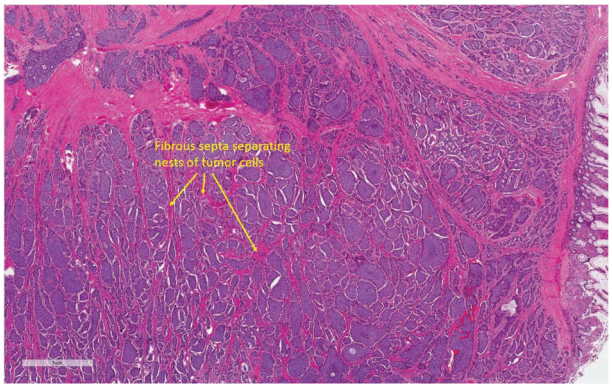

Figure 2. Pathology of carcinoid tumor. Fibrous septa are separated by nests of tumor cells as indicated by arrows.

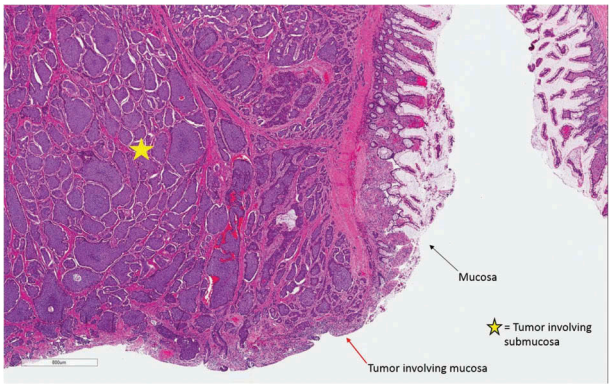

Figure 3. Pathology of carcinoid tumor. Tumor involving mucosa and submucosa layers.

6. Follow-up and outcomes

Further workup of 5-hydroxyindole acetic acid (5-HIAA) and chromogranin A showed no evidence of liver or metastatic involvement with follow up imaging. The post-operative course was unremarkable. He received outpatient follow up with oncology. Unfortunately, the patient was lost at follow-up.

7. Discussion

The patient was luckily found to have early stages of carcinoid tumor from aggressive surgical management, unfortunately since he did not follow up with outpatient oncologist his post-operative management and staging of the carcinoid tumor was inadequate in this case.

Adult ileocecal intussusception secondary to carcinoid tumor is a rare clinical presentation, as only eight cases have been reported to date [8]. The pathognomic 'target' sign lesion will typically be

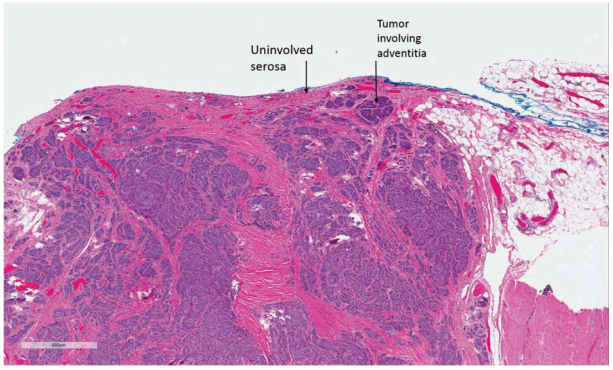

Figure 4. Pathology of carcinoid tumor. Tumor involving the adventitia layer.

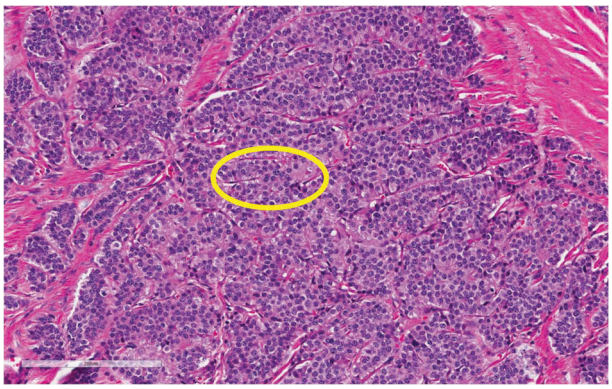

Figure 5. Pathology of carcinoid tumor. Tumor cytology with spindle cells.

demonstrated on CT scan [9]. Carcinoid tumor is a neuroendocrine tumor arising from enterochromaffin cells (Kulchitsky cells). It is the most common tumor of the small intestine, accounting for approximately 20–50% of tumors within the small intestine [7]. Primary neuroendocrine tumor arise in GI tract are not usually associated with carcinoid syndrome unless extensive metastasis or metastasize to the liver. Common symptoms include flushing, diarrhea, abdominal cramping, right-sided cardiac valvular involvement, and bronchial constriction.

Intussusception may be the only manifestation of carcinoid tumor; therefore, the cause of intussusception should always be assiduously uncovered. As carcinoid tumors may secrete 5-hydroxyindole acetic acid (5-HIAA) and chromogranin A, serial measurement of 24-hour urinary 5-HIAA can aid in evaluating for possible metastatic involvement or recurrence. Octreotide scan may be warranted during follow up to further evaluate for any metastatsis. Other possible treatment options include somatastatin, which works by decreasing the levels of serotonin secretion and also decreasing the breakdown of serotonin.

The National Comprehensive Cancer Network (NCCN) guidelines for the management of ileal carcinoid tumor with loco-regional disease as demonstrated in this case is bowel resections with regional lymphadenectomy with serial biomarker such as Chromogranin A and urinary 5′- HIAA surveillance every 3–6 months within the first year [10–19]. If the tumor recurs, further management may include different treatment options, such as chemotherapy, radiation therapy, and surgery [20].

Though ileoceccal intussusception in the adult population is rare, approximately 80–90% of adult intussusception is commonly found within the small bowel [21]. The most common malignancy in small bowel intussusception is metastatic disease to the small bowel, particularly melanoma, in addition to colon, lung, or kidney cancer. Other etiologies include lymphoma and primary adenocarcinoma [5].

In reference to our patient, the prior history of abdominal surgery which include appendectomy predisposes the patient at a higher risk to develop adhesions, which is one of the leading causes of acquiring a small bowel obstruction. Because our patient had carcinoid tumor, his prognosis is better than compared to carcinoid syndrome as that is indicative of metastasis.

8. Patient perspective

Unfortunately, the patient was lost to follow up as outpatient, his perspective could not be obtained after wards. However, during the hospitalization he was very pleased that he came in early for his hematochezia and have ‘cured’ his cancer.

9. Conclusion

Adults who present with ileoceccal intussusception should undergo a careful investigation to uncover the cause, as this can be initial presentation of a malignancy. On very rare occasions, carcinoid tumor in the small or large bowel may cause intussusception, and it may not cause the carcinoid syndrome. In an adult patient with intussusception, carcinoid tumor should be considered in the differential diagnosis.

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Availability of data and material

Data and supporting materials available upon request.

Authors’ contributions

JZ and DHT has full access to the data in the case presentation and takes full responsibility for the integrity of the data and the accuracy of the data; JZ, DHT, ACV and RDC took part in patient care; JZ and DHT equally drafted

and contributed to the manuscript and all authors have provided critical revision for important intellectual content. All authors have read and approved the final manuscript.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Disclosure statement

No potential conflict of interest was reported by the authors.

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Ethics approval and consent to participate

Not applicable.

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