

Small bowel obstruction by a terminal ileum carcinoid tumor: a case report

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Abstract

Carcinoid tumors are well differentiated neuroendocrine tumors with secretory components. These tumors are uncommon but the most common primary tumors of the distal small intestine. We present a rare terminal ileum carcinoid tumor presenting with a small bowel obstruction. A 65 years old man presented with intermittent, generalized, dull and colicky abdominal pain accompanied with intermittent nausea, fever and chills for 1 year and post prandial generalized colicky abdominal pain from 5 days prior to admission. He also complained of weight loss and frequent constipations during recent year. His abdomen was soft with mild tenderness in periumbilical, right lower quadrant and left lower quadrant without guarding, rebound tenderness and palpable mass. Laboratory findings indicated anemia, and barium enema showed right lower quadrant mass effect in small intestine. Narrowing of terminal ileum was noted in colonoscopy. Free fluid in lower abdomen and pelvis with 37*28*25 paravertebral hypoechoic pelvic mass, without peristalsis was seen in abdomen and pelvic sonography. After mass localization in abdominal CT scan, laparotomy and excisional biopsy was performed. The diagnosis of carcinoid tumor was confirmed by pathologic report. Carcinoid tumors are rare tumors of the Gastro intestinal tract, however, they are the most common primary tumors of the small intestines. Most of these tumors have a very indolent course and may present with non specific symptoms. In view of the poor prognosis associated with the late diagnosis, it is imperative to think of this differential diagnosis in patients presenting with non specific symptoms and in intermittent partial bowel obstruction.

Keywords: carcinoid tumors, small bowel obstruction, terminal ileum.

Introduction

The carcinoid tumors are well differentiated neuroendocrine tumors with secretory components [1]. These tumors are uncommon but the most common primary tumors of the distal small intestine [1,2]. They are also the most common tumor of the appendix [1]. The carcinoid tumors progress slow-

ly and remain clinically silent in most of cases [3]. They may present with pain, weight loss, abdominal mass, bowel obstruction, carcinoid syndrome or perforation [2]. It also can mimiek the symptom of IBS [1]. We present a rare terminal ileum carcinoid tumor presenting with a small bowel obstruction.

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Case presentation

A 65 years old man presented with intermittent, generalized, dull and colicky abdominal pain that was sometimes accompanied with nausea, fever and chills for 1 year prior to admission. He also complained of weight loss and constipation during recent year. He gave a history of 5 days admission in general surgery ward about 1 month prior to admission due to abdominal pain, and with impression mimicking chronic appendicitis receiving the IV antibiotic. His pain subsided and 5 days prior to admission developed severe post prandial generalized colicky abdominal pain.

On physical examination temperature was 36.6 (oral), BP: 140/60, pulse rate: 74 and respiratory rate: 18. His abdomen was soft with mild tenderness in periumbilical, right lower quadrant and left lower quadrant without guarding, rebound tenderness and palpable mass. Digital rectal examination showed empty rectum.

Laboratory findings indicated anemia (RBC:5.2 million, Hb:12.5, MCV: 95, MCH: 30.9), normal WBC count, platelet count, liver function tests, serum electrolytes, BUN, Cr, amylase and lipase. The urine analysis, and culture, stool examination and culture indices also did not show any abnormality.

The barium enema showed right lower quadrant mass effect in small intestine (Fig. 1). Narrowing of terminal ileum was noted in colonoscopy. Endoscopic examination revealed severe D1 and D2 dilatation and gastric, antrum, mucosal biopsy showed moderate chronic active gastritis with lymphoid



Fig. 1. Barium enema showing right lower quadrant mass effect in small intestines.

follicle formation. Free fluid in lower abdomen and pelvis with 37*28*25 paravertebral hypoechoic pelvic mass, without peristalsis was seen in abdomen and pelvic sonography. After mass localization in abdominal CT scan, (Fig 2,) laparotomy and excisional biopsy were performed (Fig 3). The diagnosis of carcinoid tumor was confirmed by pathologic report (Fig 4).

Discussion

The term carcinoid was first used to describe 'hormonally active' tumours in 1907 by Oberndorfer [4]. They are derived from stem cells in the gut wall and have been classified based on the location within the primitive gut from which they arise [5].



Fig. 2. CT Scan images reveal a soft density mass with speculated borders.

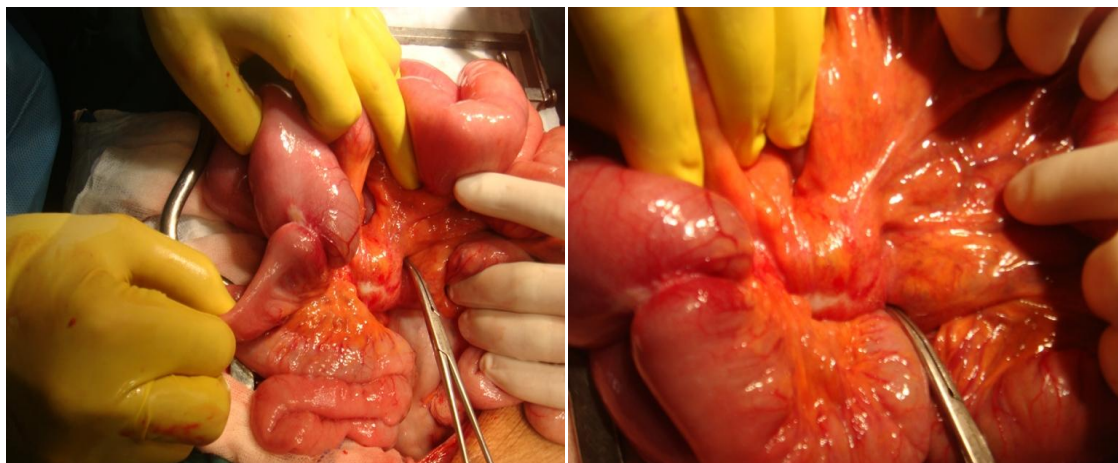


Fig. 3. Laparotomy and excisional biopsy of the primary lesion.

They are the most common primary tumor of the small bowel and mesentery and account for more than 95% of all carcinoids and 1.5% of all gastrointestinal tumors. The gastrointestinal carcinoid arises from the endochromaffin cells of Kulchitsky i.e. neural crest cells situated at the base of crypts of Lieberkuhn. Various sites of origin of this neoplasm are appendix- 30-45%, small bowel - 25-35% (duodenum 2%, jejunum 7%, ileum 91%, multiple sites 15-35%), rectum 10-15%, caecum - 5% and stomach - 0.5% carcinoids can also rarely occur in pancreas, biliary tract, esophagus and liver [6,7].

The sex prediction for the tumor is M:F

Most carcinoids occur in patients older than 50 years, however, appendiceal carcinoids occur in young patients in their second to fourth decade.

Clinical presentation can be with non-hormonal symptoms secondary to tumor bulk and local reaction, or with symptoms of a functioning tumor, described as 'carcinoid syndrome'. 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 and are thus locally advanced at presentation

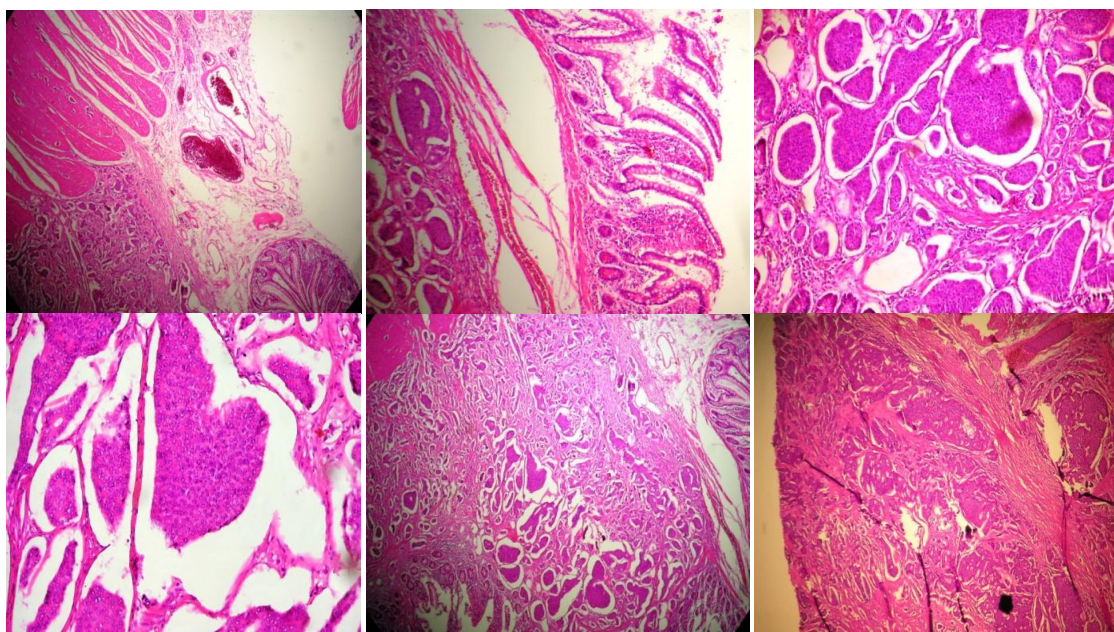


Fig. 4. Histopathological slides confirming the diagnosis of Carcinoid tumor.

[8,9].

Non-hormonal symptoms are most commonly secondary to partial mechanical obstruction of the small bowel with patients complaining of intermittent vague abdominal symptoms such as pain and distention. These obstructive features may be due to peritumoral fibrosis or invasion, causing direct luminal strictures, or secondary to desmoplastic reaction leading to ischemic changes [10]. Other non-hormonal symptoms include anorexia, weight loss, fatigue and occasionally a palpable abdominal mass is noted. The term 'carcinoid syndrome' is used to describe the hormonal manifestations of carcinoid tumors and occurs secondary to the secretion of serotonin, tachykinins, bradykinins and prostaglandins. Midgut carcinoid tumors are typically seen to secrete serotonin [11]. Recent consensus is that functionally active tumors account for around 20–30% in this group, higher than previously thought [12]. The syndrome occurs when vasoactive substances from tumor secretion enter the systemic circulation. In the case of small bowel tumors this can only occur in the presence of multiple hepatic metastases, as substances such as serotonin usually undergo hepatic degradation to non-active substrates.

The primary tumor is small, slow growing and is rarely demonstrated radiologically. The diagnosis is made when the tumor has spread through the bowel wall into the peritoneum and mesentery and is achieved by several complementary imaging techniques [13]. Plain abdominal skiagram is the initial screening modality and it may show curvilinear calcification. However, it is highly non-specific [6]. Barium studies (enteroclysis, follow through examination) may show intramural or intraluminal filling defects in the distended ileum. There can also be narrowing of the lumen with stricture formation, thickening of the valvulae conniventes and increase in the interbowel loop distance due to wall thickening. Sonography of the bowel can depict pseudokidney sign due to wall thickening, associated lymphadenopathy and liver metastasis. However, the

findings are too nonspecific to offer a confident diagnosis [6]. Diagnosis can be made fairly confidently on computed tomography. It reveals a soft density mass with spiculated borders and radiating strands with or without calcification in 80% of cases [7]. However, similar mesenteric masses can be seen in lymphoma, leukemia, metastatic carcinoma and sarcoma (especially ovary and leiomyosarcoma, retractile mesenteritis, kaposi's sarcoma and peritoneal mesothelioma. Other findings on CT can be lymphadenopathy (retroperitoneal, retrocrural) and metastasis commonly to the liver. Metastases are frequent from a midgut primary tumor and rare from appendiceal carcinoid. They also depend on size of the primary tumor: tumors smaller than one cm metastasize in two per cent of cases, tumor 1-2 cms metastasize in 50% and >2 cms are clinically silent but they may produce carcinoid syndrome. Bone metastases are rare and most osseous metastases have been reported from carcinoid of the stomach and rectum. Mostly they are osteoblastic but rarely, lytic or mixed lesions occur [6].

Carcinoid tumors may also be associated with a high incidence of other primary malignant neoplasms e.g. in the colon, breast, pancreas, kidney, lip, vulva, cervix, larynx, rectosigmoid and histiocytic lymphoma [14]. Somatostatin receptor scintigraphy with Indium III Octreotide, Indium III pentetrotide and Iodine 123 meta iodobenzylguanidine is a sensitive and non-invasive technique for imaging carcinoid and its metastatic spread.

Given that the majority of these tumours are well-differentiated, following an indolent course, prognosis is surprisingly unfavorable. This can mainly be attributed to the fact that the majority of cases present so late by which time spread has invariably occurred. The principal management approach in these cases is surgical resection of the primary lesion and is the only curative option. With smaller lesions (<1cm) local resection is usually adequate. However, with lesions over 1.5 cm there is a high risk of recurrence and thus segmental resection is required with extensive clearance of the associated mesen-

teric lymph nodes [1]. Surgery has been shown to be of benefit even in patients with metastatic disease, both to gain symptomatic relief and improve survival [12]. Other possibilities for symptomatic management of patients with hormonal symptoms include biotherapy using drugs such as somatostatin analogs. These have been shown not only to provide symptomatic improvement in 70–80% but also to stabilize tumor growth [12]. This therapy is therefore used as first-line treatment in inoperable functioning tumors and to avoid a carcinoid crisis in those undergoing surgery.

Conclusion

Carcinoid tumors are rare tumors of the Gastro intestinal tract, however, are the most common primary tumors of the small intestine. Most of these tumors have a very indolent course and may present with non specific symptoms. Prognosis in this condition can be dramatically improved with early diagnosis, and surgical management at this stage is often curative. In view of the poor prognosis associated with the late diagnosis, it is imperative to think of this differential diagnosis in patients presenting with non specific symptoms and in intermittent partial bowel obstruction.

References

1. Singhal H: Carcinoid tumor, Intestinal. Available online at: <http://www.emedicine.com>. Accessed 22 Jul 2010.
2. Burke AP, Thomas RM, Elsayed AM, Sobin LH: Carcinoids of the jejunum and ileum: an immunohistochemical and clinicopathologic study of 167 cases. *Cancer* 1997; 79:1086-1093.
3. Marshall JB, Bodnarchuk G: Carcinoid tumors of the gut. Our experience over three decades and review of the literature. *J Clin Gastroenterol* 1993; 16(2):123-9.
4. Oberndorfer S: Karzinoide tumoren des dünn-darms. *Frankfurt Zietschrif fur Pathologie* 1987; 1:426-429.
5. Broaddus RR, Herzog CE, Hicks MJ: Neuroendocrine tumors (carcinoid and neuroendocrine carcinoma) presenting at extra-appendiceal sites in childhood and adolescence. *Arch Pathol Lab Med* 2003; 127(9):1200-3.
6. Khan AN, Carcinoid. *Gastrointestinal. emedicine*, July, 2005. Available online at: <http://www.emedicine.com>. Accessed 25 Jul 2010.
7. Gould M, Johnson RJ. Computed Tomography of abdominal carcinoid tumor. *British J. of Radiol.* 1986; 59, 881-885.
8. Yamaguchi T, Manabe N, Tanaka S, et al. Multiple carcinoid tumors of the ileum preoperatively diagnosed by enteroscopy with the double-balloon technique. *Gastrointest Endosc* 2005; 62:315-8.
9. Strodel WE, Vinik AI, Thompson NW, et al.: Small bowel carcinoid tumors and the carcinoid syndrome. In *Endocrine Surgery Update* Edited by: Thompson NW, Vinik AI. New York, NY: Grune and Stratton; 1983:277-291.
10. Levy AD, Sobin LH: Gastrointestinal Carcinoids: Imaging features with clinicopathologic comparison. *Radiographics* 2007; 27(1):237-257.
11. Basson MD, Ahlman H, Wangberg B, et al.: Biology and management of the midgut carcinoid. *Am J Surgery* 1993, 165:288-297.
12. Eriksson B, Klopel G, Krenning E, et al.: Consensus guidelines for the management of patients with digestive neuroendocrine tumors – well-differentiated jejuno-ileal tumor/carcinoma. *Neuroendocrinology* 2008; 87:8-19.
13. Piais D, Glazer HS, Levitt RG, Husband JE. Computed tomography of abdominal carcinoid tumors. *Am. J. of Roentgenology*, 1985; 143: 581-584.
14. Barclay THC, Schapira DV. Malignant Tumors of the small intestine. *Cancer* 1983; 51: 878-887.