

Anemia as the Only Clinical Presentation of Metastatic Jejunal Carcinoid Tumor: A Case Report with Radiological and Enteroscopic Findings

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Article Info	ABSTRACT
<p>Article type: Case Report</p>	<p>Background: Neuroendocrine tumors (NETs) are reported to be responsible for about 2% of all malignancies of the gastrointestinal tract. They consist of several different tumors, which are emanated from the cells of the diffuse NE cell system. Carcinoids, which are also called carcinoid tumors, constitute the largest group of NE tumors with an incidence rate of about 2.5 per 100,000 people. Carcinoid tumors are distinct tumor subtypes arising from the diffuse endocrine system outside the pancreas and thyroid. This neoplasm originates from different sites, including 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%). Primary jejunal carcinoid tumors are reported to be uncommon. Moreover, carcinoid tumors rarely presented with occult gastrointestinal bleeding.</p>
<p>Article History: Received: 17 November 2019 Revised: 22 December 2019 Accepted: 29 December 2019</p>	<p>Case Report: This report presents a case of metastatic jejunal carcinoid tumor in a 64-year-old male presented with anemia and positive guaiac test without any symptoms of carcinoid syndrome.</p>
<p>Keywords: Anemia Metastatic jejunal carcinoid Occult bleeding Radiological and enteroscopic findings</p>	<p>Conclusion: Primary jejunal carcinoid tumors are reported to be uncommon. Moreover, carcinoid tumors rarely presented with occult GI bleeding. Here, we report a case of jejunal carcinoid tumor with metastasis to the liver presented with anemia and positive guaiac test without any symptoms of carcinoid syndrome.</p>

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Introduction

Neuroendocrine tumors (NETs), which are defined as epithelial tumors with neuroendocrine distinctive features, are reported to be responsible for about 2% of all malignancies of the gastrointestinal (GI) tract (1). They usually develop as a slowly developing rare neoplasm, some characteristics of which include the organ specificity and some others are commonly observed in all forms. In most cases, this kind of tumor cannot be easily diagnosed due to various reasons. Differential diagnosis of these tumors is rather difficult since they constitute a rare form of the neoplasm. In addition, clinical manifestations of these tumors are nonspecific; therefore, nothing is indicative of this entity. Nonetheless, the annual incidence of NETs is quoted to increase by 40-50 cases per million population.

This sharp increase probably cannot be ascribed to a real increase in the incidence rate; however, it can be attributed to improved diagnostic tools that have recently become available (2).

Carcinoids, which are known as carcinoid tumors, constitute the largest group of NE tumors with the incidence rate of about 2.5 per 100,000 population and were traditionally divided into foregut, midgut, and hindgut tumors (3).

Nonetheless, according to the World Health Organization, carcinoid tumors are classified into five subtypes (4), including well-differentiated endocrine tumors, well-differentiated endocrine carcinoma, poorly differentiated endocrine carcinoma, mixed exocrine and endocrine carcinomas, and tumor-like lesions. This classification is not limited to carcinoids

and can be applied to all types of NETs. According to the recent findings, different molecular pathways may develop carcinoid tumors in the lung and GI tract. The biological behavior of GI tumors can be caused by the inactivation of tumor suppressor genes on chromosome 18. Although familial midgut carcinoids tumors are uncommon, bronchial carcinoids, endocrine pancreatic tumors, and gastric carcinoids may be indicative of multiple endocrine neoplasia type 1, which is a familial syndrome (5, 6).

Carcinoid tumors are commonly observed in the GI tract (66.9%) succeeded by the tracheobronchial system (24.5%). They may also occur in the liver, gallbladder, ovary, testis, and thymus in exceptional cases (7). This neoplasm originates from different sites, including 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%). Therefore, primary jejunal carcinoid tumors rarely occur. Small-bowel carcinoid tumors spread to the liver or regional lymph nodes in 58-64% of the patients when they are diagnosed with the disease (8).

Here, we report a case of jejunal carcinoid tumor with metastasis to the liver presented with anemia and positive guaiac test without any symptoms of carcinoid syndrome. The examinations included small-bowel barium follow-through, computed tomography (CT) scan, and single balloon enteroscopy.

Case Report

A 64-year-old male referred to our hospital with complaints of pallor and weakness within the preceding 6 months. He had no abdominal pain, flushing, or diarrhea, and physical examinations revealed only anemia. Laboratory analyses reported white blood cell: 7,900; seg/lym: 65/33%; hemoglobin: 9.6 g/dL; MCV: 68 fL; MCH: 26 pg; and platelet count: 267,000/cumm. Biochemistry analysis indicated normal blood urea nitrogen and creatinine, aspartate aminotransferase (35U/L), alanine aminotransferase (36 U/L), alkaline phosphate (545 U/L), total bilirubin (1.1 mg/dL), and Ferritin (10µg/L). In addition, the erythrocyte sedimentation rate was measured at 35 mm/hr and C-reactive protein was estimated at 0.9 mg/dL. Totally, two of three stool examinations revealed positive occult blood.

Colonoscopy and gastroscopy revealed no abnormalities. Small-bowel barium follow-through examination illustrated the thickening of mucosal folds and small bowel loops without obstruction (Figure 1). The CT scan of the abdomen indicated clear-cut hepatic mass lesions with necrotic areas (Figure 2), as well as a jejunal mass lesion (Figure 3).



Figure 1. Barium follow-through revealing the thickening of small bowel loops without obstruction

Subsequently, based on these radiological findings, the patient was initially diagnosed with a jejunal tumor and hepatic metastases. Figure 4 depicts the results of enteroscopy indicating the thickening of small bowel loops with ulceration. The CT-guided tru-cut biopsy was performed on hepatic mass lesions and mucosal biopsy samples of jejunum were evaluated as carcinoid tumor histopathologically.

Discussion

More than 95% of all carcinoids arise in the GI tract comprising 1.5% of all GI tumors. These

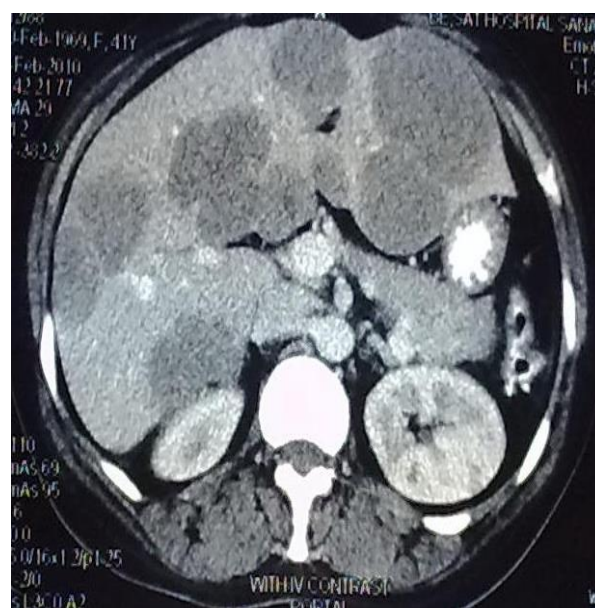


Figure 2. Computed tomography scan of the abdomen demonstrating clear-cut hepatic mass lesions with necrotic areas

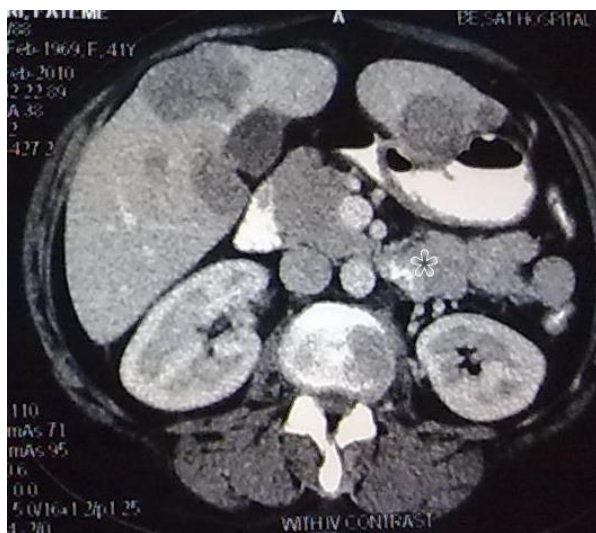


Figure 3. Computed tomography scan of the abdomen indicating a jejunal mass lesion (*) and the thickening of small bowel loops

tumors originate from the enterochromaffin cells derived from neural crest cells, which are located at the base of crypts of Lieberkuhn (9). This tumor has a trend towards male predominance with the sex prediction of M/F: 2/1. Although the majority of carcinoids tumors develop in patients older than 50 years of age, appendicular carcinoid is more prevalent in young patients in the second to fourth decades of their lives. The majority of the patients are asymptomatic; nonetheless, symptoms can vary from pain to intestinal obstruction (19%), weight loss (16%), palpable mass (14%), intussusceptions, perforation, or GI bleeding (rare) (10). Our patient was a 64-year-old male presented with symptoms of anemia and occult GI bleeding, which is quite uncommon in the cases of carcinoid tumors.

The primary carcinoids of jejunum and ileum vary in their biologic behavior and ability to metastasize as the other carcinoids do. In a broad sense, carcinoids of the small intestine are likely to act in a malignant form leading to liver and lymph node metastases. In addition, carcinoid tumors secrete serotonin and other histamine-like substances that can cause carcinoid syndrome, which is characterized by such symptoms as abdominal cramps, diarrhea, and flushing (11). Nonetheless, no symptom of carcinoid syndrome was detected in our case with liver metastasis.

The primary tumor is hardly detectable on radiology since it is small and slow-growing. However, intramural or intraluminal filling defects may be observed on barium examination. This tumor can also be manifested in other ways, including stricture (narrowing of the lumen), Kerckring folds/valves, and an increase in the inter-bowel loop distance due to wall thickening. The distinguishing



Figure 4. Enteroscopy revealing a jejunal mass lesion and the thickening of small bowel loops with ulceration

feature of carcinoid is the looping of the small bowel which is demonstrated on barium examinations.

The CT scan can reveal a soft density mass with spiculated borders and radiating strands with or without calcification in 80% of the cases (12, 13). In our patient, the CT scan was clearly indicative of jejunal and hepatic masses. Metastases frequently occur in midgut primary tumors, whereas appendiceal carcinoid rarely metastasizes. The odds of metastases also depend on the size of the primary tumor, and tumors smaller than 1 cm and within 1-2 cm metastasize in 2% and 50% of the cases, respectively. Moreover, the tumors larger than 2cm are clinically silent; however, they may lead to carcinoid syndrome. Carcinoid tumors rarely metastasize to the bones, and the majority of them are observed in carcinoid of the rectum and stomach (13).

Conclusion

Primary jejunal carcinoid tumors are reported to be uncommon. Moreover, carcinoid tumors rarely presented with occult GI bleeding. Here, we report a case of jejunal carcinoid tumor with metastasis to the liver presented with anemia and positive guaiac test without any symptoms of carcinoid syndrome.

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Conflict of interest

The authors of the present article did not use

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