



BMH Med. J. 2019;6(4):135-139. **Case Report**

Ileal Carcinoid With Hepatic And Peritoneal Metastasis Presenting As Carcinoid Syndrome

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Abstract

Gastrointestinal carcinoid tumours are uncommon neuroendocrine tumours. They can either present as a non-functioning tumour mass, or as a "functioning" tumour secondary to the production of several biopeptides resulting in carcinoid syndrome. The incidence of carcinoid syndrome is less, and usually occurs in carcinoids of small bowel origin with hepatic metastasis. The patient being reported is a case of ileal carcinoid with hepatic and peritoneal metastasis presenting as carcinoid syndrome.

Keywords: Carcinoid tumour, carcinoid syndrome, ileum, octreotide

Case presentation

An elderly person presented with 4 days history of diffuse abdominal pain and 3 to 4 episodes of loose watery stools. There was no fever or vomiting. He also noticed episodic flushing of his face and upper chest for the past 2 months. On presentation, his vitals and systemic examinations were normal. His face and upper chest were red.

His complete blood counts, electrolytes, renal and liver functions were normal. Serum albumin and corrected calcium were 3.0 g/dL and 8.5 g/dL respectively. Amylase and lipase levels were normal. Ultrasound of the abdomen showed large heterogenous mass lesions in the liver and a few lesions in the mesentery and anterior abdominal wall with mild ascites. Alpha-feto protein and CA19-9 levels were normal. Contrast enhanced computed tomography of the abdomen revealed an enhancing endo-exenteric mass of about 35 x 31 x 29 mm in the distal ileal loop (**Figure 1**), a mass lesion (41 x 33 mm) in the mesentery at the level of L4 vertebrae encasing the trunk of the superior mesenteric artery, multiple variable sized enhancing nodules throughout the peritoneal cavity and multiple variable sized enhancing nodules in both lobes of the liver.

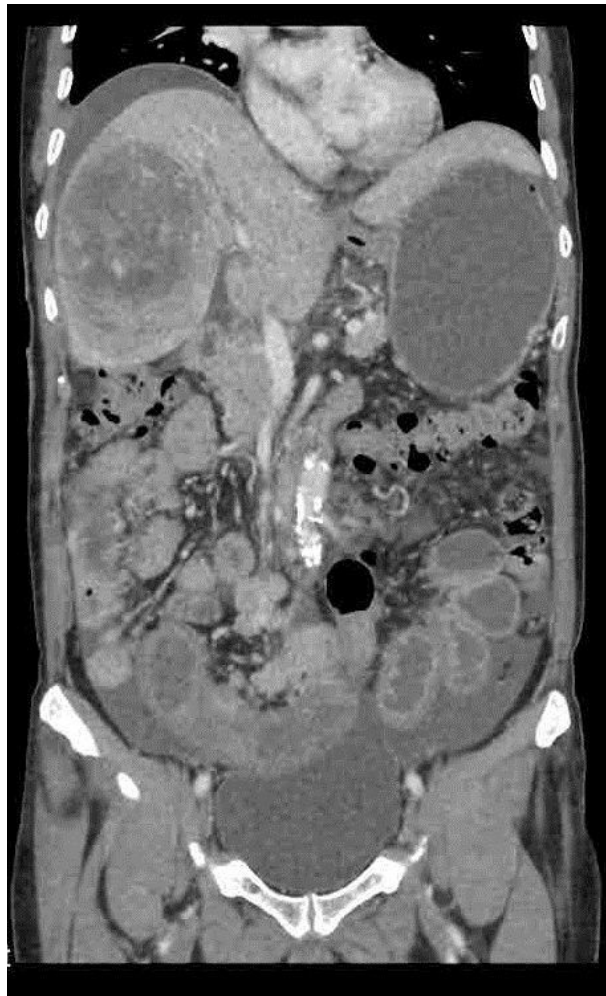


Figure 1: Contrast enhanced CT abdomen showing ileal carcinoid with liver metastasis

Histological examination of biopsy of liver mass showed neoplasm composed of organoid nests and lobules of cells having small uniform nuclei and fine chromatin and moderate cytoplasm (**Figure 2**), with the cells being positive for cytokeratin, chromogranin A and synaptophysin, suggestive of carcinoid tumour.

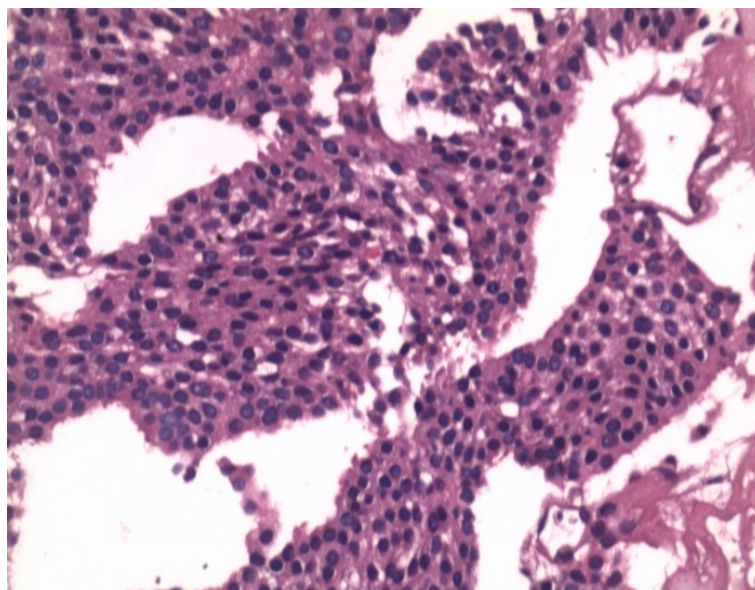


Figure 2: Liver biopsy showing carcinoid tumour

Urine spot 5-hydroxy indole acetic acid (5-HIAA) and plasma chromogranin A levels were elevated,

114 mg/g creatinine and 2220 ng/mL respectively. Endoscopic evaluation revealed antral gastritis and colonic erosions with colonic and ileal diverticuli. GA68 DOTATOC PET whole body scan showed somatostatin receptor expressing primary neuroendocrine tumour involving distal ileal loop and metastatic lesions in mesentery, omentum, peritoneum and liver. Chest X-ray, ECG and echocardiography were normal.

He was started on subcutaneous octreotide (150 mcg thrice daily), following which he became asymptomatic. His facial flushing disappeared after 1 week of octreotide (**Figure 4 a and b**). He was not willing for any surgical intervention. Prior to discharge, he was started on once monthly long acting octreotide (20 mg intramuscular). On 6 months follow-up, patient continued to be asymptomatic. His repeat urine spot 5-HIAA and plasma chromogranin A levels got reduced to 84 mg/g creatinine and 520 ng/mL respectively He was primed about the need for intestinal resection in case of any obstruction. He was also informed about embolization of liver metastasis.



Figure 3: a) Redness of upper chest on presentation b) Disappearance of redness after starting octreotide

Discussion

Though carcinoid tumours are relatively rare, the incidence is more in gastrointestinal tract (67.5%), followed by the bronchopulmonary system (25.3%), and the remaining being found in the liver, pancreas, thymus, ovaries, prostate and kidneys [1]. The tumour arises from the endochromaffin cells of Kulchitsky. The incidence is more among males and age above 50 years. These tumours also have the tendency to metastasize to liver, lymph nodes and rarely bone [2,3].

Carcinoid syndrome is caused by the tumour secretion of hormonal mediators like serotonin, histamine, kinins and prostaglandins. It manifests with facial flushing, abdominal cramps, diarrhoea, bronchospasms, edema of head and neck, telangiectases and valvular lesions like pulmonary and tricuspid stenosis or regurgitation. The cardiac lesions occur due to the formation of serotonin mediated fibrotic plaques and can result in heart failure. Patient may also present with asthma-like symptoms and pellagra-like skin lesions. Retroperitoneal involvement can lead to fibrosis and subsequent urethral obstruction, intra-abdominal fibrosis and mesenteric arterial or venous occlusion. Atypical carcinoid syndrome is characterized by Cushing syndrome and acromegaly due to production of adrenocorticotrophic hormone and growth-hormone-releasing hormone [4].

The estimation of neuroendocrine markers, chromogranin A in serum and 5-HIAA in urine, may be useful for the diagnosis. 5-HIAA has a sensitivity of 73% and a specificity of 100% for detecting carcinoid; however, these levels have no clear correlation with symptoms. Serum chromogranin A levels are elevated in 56-100% of carcinoid tumours, and they correspond to the tumour bulk. Other tests include platelet 5-hydroxytryptamine, urinary 5-HT and urinary tryptophan. Oesophagogastroduodenoscopy is useful in diagnosing duodenal carcinoids. Contrast enhanced CT and MRI mainly indicates liver and/or mesenteric metastases, and do not usually identify small primary tumours. The tumour size, level of wall invasion and the presence of regional lymphatic metastases can be determined by endoscopic ultrasonography [5,6]. Indium111 or metaiodobenzylguanidine scan can be used to identify carcinoid tumours.

Intestinal carcinoid is a common malignant tumour of the distal small intestine. Even though its prevalence is relatively high, the occurrence of symptoms or carcinoid syndrome is very rare. The incidence is similar for both sexes, and the average age of presentation is 66 [7]. On clinical presentation, about 12% of patients have distant metastases, whereas carcinoid syndrome occurs only in one out of 300,000 patients with intestinal carcinoid tumour [8]. The clinical presentation can be secondary to the tumour or carcinoid syndrome. About 90% of acute presentation is due to intestinal obstruction which occurs secondary to the desmoplastic reaction in the intestine and the mesentery. This may also cause acute mesenteric ischaemia in 30% of patients. Mild bleeding is another acute intestinal manifestation. Chronic intestinal symptoms include pain and change in bowel habits [9,10].

The main objective of treatment is aimed at controlling the symptoms, improving the quality of life and increasing the survival period. Wide surgical resection, including the mesentery and lymph nodes, is required. Cytoreductive surgery should be performed, if complete resection is not possible due to retroperitoneal spread or spread to the large vessels. The options in cytoreductive therapy include ablation with surgical metastasectomy, radiofrequency and cryotherapy. Arterial embolism can be performed in cases where the metastases are unresectable, in order to reduce the tumour size and symptoms of carcinoid syndrome. The response to chemotherapy rarely exceeds 30%. Drugs like 5-fluorouracil and streptozocin can be used palliatively. The use of radiotherapy is limited to metastases of the central nervous system and painful bone metastases [11]. Somatostatin analogues are used for symptomatic relief and to reduce hormone levels. They may be considered in asymptomatic patients with progressive disease. They are also administered before, during, and after surgical procedures and embolization in order to prevent or treat carcinoid crises. Interferon alfa is the main medical treatment for low-proliferating gastroenteropancreatic tumours. They inhibit protein and hormone synthesis in the tumour cells, inhibits angiogenesis, and stimulate the immune system. It can be used in combination with somatostatin analogues. Loperamide or diphenoxylate can be used to control diarrhoea. H1 or H2 blockers can be administered for histamine-secreting tumours [12].

The prognosis and follow-up of intestinal carcinoid patients is based on their clinical condition at the time of presentation and the therapy they have received. Serum markers like chromogranin A and 5-HIAA are monitored every 3-4 months in the first year to establish the pace of the disease, along with conventional imaging. A once yearly Octrescan is recommended if the baseline scan was positive. Annual follow-ups are required for those who have undergone complete macroscopic resection [12].

Conclusion

Carcinoid tumours are rare and have a wide spectrum of symptoms. Majority of the tumours are slow-growing with non-specific symptoms. Of these, only a few present with carcinoid syndrome. Therefore, an early diagnosis and appropriate treatment can prove beneficial to these patients.

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