Original Research Paper



SMALL INTESTINAL CARCINOID TUMOR-A CASE REPORT

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ABSTRACT

Carcinoid tumors are neuroendocrine tumours, the most common tumour of the small bowel and its incidence is rising. Most carcinoid tumours grow slowly and mostly asymptomatic or may present with nonspecific abdominal pain or carcinoid syndrome. We had a 38 year old, female patient presented with generalized, intermittent, dull aching abdominal pain accompanied with distension and obstipation for 2 days with associated vomiting. She complained of weight loss and constipation during this period. On examination abdomen showed deep tenderness in periumbilical, right lower quadrant regions. No guarding, rigidity, mass or free fluid. On per-rectal examination, there was no tenderness and the rectum was empty. Contrast enhanced computed tomography scan showed enhancing polypoidal lesion noted in distal ileum causing proximal dilatation of small bowel with air fluid levels and circumferential wall thickening in dilated ileum proximal to polypoidal lesion, strongly enhancing nodal mass with specs of calcification, multiple mesentric lymph nodes and no liver metastasis. As the conservative management for 48 hours showed no improvement in symptoms, an emergency exploratory laparotomy was performed, and an intraluminal mass was identified from ileocecal junction causing obstruction with dilated loops of ileum and jejunum. Local resection of terminal ileum was performed and a side to side anastomoses was done using GI stapler. Histopathology showed diagnosis of neuroendocrine tumor.

KEYWORDS: Adult Intestinal obstruction, Ileal Carcinoid, Neuroendocrine tumour, Laparotomy

INTRODUCTION

Carcinoid tumors are classified as neuroendocrine tumours. Carcinoid tumours are a group of low-grade, slowgrowing, well-differentiated tumours originating from the endochromaffin cells of Kulchitsky, that is, neural crest cells situated at the base of the crypts of Lieberkuhn in the gastrointestinal tract (GIT) and can produce several biologically active substances (hormones) and hence are classified into 'functioning' and 'non-functioning' types.¹

It is the most common tumor of the small bowel and its incidence is rising. Most carcinoid tumors grow slowly and mostly asymptomatic or may present with nonspecific abdominal pain or carcinoid syndrome. After appendix small bowel (ileum) is the most common location and has metastatic potential. 2

Diagnosis of carcinoid tumours is made on the basis of clinical manifestations, peptide and amine secretion and radiological/nuclear imaging but confirmation is made by histological analysis. Medical treatment is mainly with somatostatin analogues, which are effective in controlling most symptoms. Surgery is the cornerstone of treatment for intestinal carcinoids.³

CASE REPORT

A 38-year-old, female patient presented with generalized, intermittent, dull aching abdominal pain accompanied with distension and obstipation for 2 days with associated vomiting. She complained of weight loss and constipation during this period. On physical examination patient was afebrile, blood pressure: 110/70 mm of Hg, pulse rate: 74/min and respiratory rate: 18/min. Her abdomen showed deep tenderness in periumbilical, right lower quadrant regions. No guarding, rigidity, mass or free fluid. On per-rectal examination, there was no tenderness and the rectum was empty. Blood investigations indicated anemia (Hemoglobin: 9.5 g%) while the other blood and biochemical tests were normal. An erect abdomen X-ray performed showed multiple air-fluid levels suggesting small bowel obstruction.

Contrast enhanced computed tomography scan with oral and

intravenous contrast showed enhancing polypoidal lesion 2.3 x 1.7cm noted in distal ileum causing proximal dilatation of small bowel with air fluid levels and circumferential wall thickening in dilated ileum proximal to polypoidal lesion of length > 10cm, minimal free fluid in pelvis, strongly enhancing nodal mass 4.5 x 2.7cm just anterior to aortic bifurcation with specs of calcification kinking of mesenteric vessels and adjacent fat stranding, multiple mesentric lymph nodes and no liver metastasis. As the conservative management for 48 hours showed no improvement in symptoms, an emergency exploratory laparotomy was performed, and an intraluminal mass of 3 x 2 cm was identified at 20 cm from ileocecal junction causing obstruction with dilated loops of ileum and jejunum (Figure 1). Bowel wall was thickened with narrowed lumen. An adjacent mesenteric nodal mass of 4 x 3 cm was noted and no liver metastasis. Local resection of terminal ileum was performed for a length of 50 cm and a side to side anastomoses was done using GI stapler. (Figure 2) Macroscopic examination showed typical yellowish nodular growth in the full thickness of the bowel wall with typical narrowing at the site of the growth (Figure 3). Specimen was sent for histopathology that showed diagnosis of neuroendocrine tumor Grade 2 with Chromogranin, Synaptophysin intense positivity and Ki67-3%. Postoperatively, the patient made an uneventful recovery and was started on orals on day 5.



Figure 1: Intraoperative picture



Figure 2: Stapled anastomosis after resection



Figure 3- Gross specimen

DISCUSSION

Carcinoid tumours were originally termed 'karzinoide' or 'cancer-like' by Oberndorfer in 1907 due to their slower growth in comparison with adenocarcinomas but the current nomenclature for tumours of epithelial origin occurring in the GIT with neuroendocrine differentiation is gastroenteropancreatic neuroendocrine tumours or 'GEP-NETs' and are specifically found in the ileum and appendix. They can also be found in the rectum, caecum, jejunum, pancreas, biliary duct, oesophagus, liver and lung. [3] Carcinoid tumours can produce several biologically active substances, such as serotonin, histamine, dopamine, gastrin, kinins, kallikrein, adrenocorticotropic hormone, growth hormone, substance P, calcitonin and prostaglandins. $^{[4.5]}$ Non-functioning carcinoids are usually asymptomatic or may show symptoms of mass effect, as in our patient. They are found incidentally during surgery, with their carcinoid nature being ascertained only following histological analysis. [6] Functioning carcinoids have the potential to cause carcinoid syndrome, resulting from excessive activation of bioactive mediators throughout systemic circulation and manifests with a plethora of symptoms including nausea, vomiting, watery diarrhoea, facial flushing, wheezing, weight loss, abdominal pain, GI blood loss and heart failure. This is most likely when the carcinoid tumour is situated in the small intestine (90%).[7,8] In some rare cases, tumour manipulation, anaesthetic induction or other invasive therapeutic procedures such as embolisation or radiofrequency ablation (RFA) may induce a sudden release of large amounts of vasoactive peptides into the systemic circulation resulting in carcinoid crisis, which consists of severe flushes, diarrhoea, dehydration, hypotension and arrhythmias.

Octreotide prophylaxis is recommended during invasive surgical procedures involving carcinoid tumours. [9] The majority of patients with small intestinal carcinoids present with abdominal pain, bowel obstruction and metastases. Metastases is most common to liver (60–80%) and lymph nodes. Another complication of small intestinal carcinoids is their tendency to cause mesenteric fibrosis, which could lead to kinking of the bowel, ischaemia, necrosis and perforation. [10]

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