Case Report

# Subtotal Gastrectomy for Atypical Carcinoid of Stomach– A Rare Tumour

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## Abstract

The term carcinoid was first employed by Oberndorfer in 1907 to describe a group of tumours of the gastrointestinal tract that had a relatively indolent course and that were considered to be intermediate between adenoma and carcinoma in malignant potential.

Gastrointestinal carcinoid tumours are a type of cancer that form in the lining of the gastrointestinal tract originating from entero-chromaffin like (ECL) cells. Gastric carcinoid tumours are rare tumors that develop within the gastric mucosa. They can present as an isolated lesion or there can be multiple lesions. The tumours can invade locally into deeper structures of the gastrointestinal tract (GIT) wall. Solitary gastric carcinoids have a greater chance for the development of malignancy and metastases as compared to multiple gastric carcinoids due to hypergastrinemia.

A 60 years old man presented with abdominal pain, vomiting and weight loss and was found to have carcinoid tumour of stomach without classic carcinoid syndrome (CS). Despite advances in the understanding of patho-physiology of carcinoid tumour its complications remain enigmatic. Early, accurate diagnosis and aggressive treatment is recommended.

***Key-words:*** Carcinoid Syndrome (CS), Entero-chromaffin like (ECL) cells, Gastrointestinal tract (GIT), Gastric Carcinoids (GC).

## Introduction

Gastric carcinoids (GC) are rare human neuroendocrine tumours (NETs) and represent only 3%-4% of the alimentary tract carcinoids and 0.3% of the gastric neoplasm[1,2]. The original description of Carcinoids was made by Langhans in 1867. They are characterized by secretarygranules and ability to produce biogenic amines and polypeptide hormones. Gastric carcinoid may present with anaemia caused by bleeding from an ulcerative mass, abdominal pain or with carcinoid syndrome (CS). Typical carcinoid syndrome associated with specific set of symptoms including flushing, diarrhoea, weight loss, palpitations, congestive heart failure, asthma and Cushing's syndrome[2].

## Case History

A 60 years old male presented with history of localized persisting pain in the upper abdomen which was burning in nature aggravated by food and had no definite relieving factor. The intensity of pain increased day by day. The patient complained of loss of appetite, significant weight loss and also passage of black coloured stool several times within last 06 months. He also complained of occasional vomiting after food for last 02 months which contained partially digested food particles and there was no history of induced vomiting.

A firm to hard intra-abdominal mass was palpable in the epigastrium, globular in shape, mildly tender, 4cm x 3cm in dimension, consistency hard, margin irregular and mobile from side to side. The patient was evaluated thoroughly with relevant investigations which included: CBC: Hb-09 g/dl, ESR- 56 mm in 1st hour, Electrolytes: Na+ – 133mmol/l, K+ – 4.3mmol/l, Cl+ – 100mmol/l, LFT: Bilirubin-0.2 mg/dl, SGPT-33U/L, ALP- 51U/L, Serum total protein: 65gm/l, Serum Albumin: 37gm/l, CA 19-9: 12.0U/ml, CEA: 0.2ng/ml, Stool for OBT: Positive, Endoscopy and biopsy of stomach: There was an irregular ulcer in the antral region of the stomach, biopsy was taken and histopathology revealed poorly differentiated adenocarcinoma.

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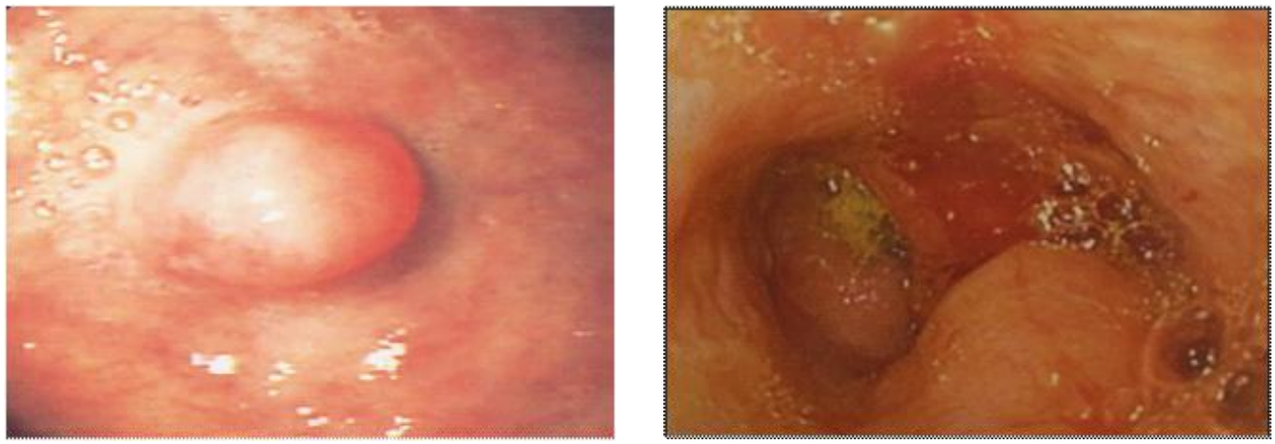

**Fig-1:** Endoscopic image of poorly differentiated gastric carcinoid tumours showing large, irregular friable mass

CT scan of abdomen: Circumferential wall thickening is noted in the antral part of stomach with luminal narrowing, leading to proximal dilatation of stomach. Post contrast scan revealed enhancement of the lesion.

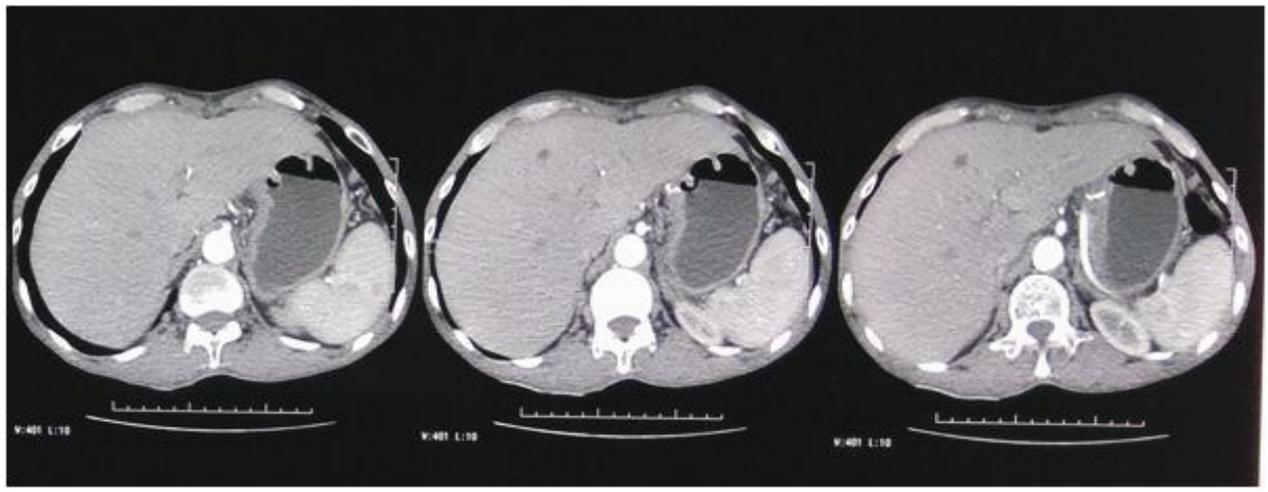

**Fig-2:** Computed tomography scan of abdomen with oral contrast demonstrates evidence of enhancing irregular thickening in the antrum

The patient underwent subtotal gastrectomy and gastrojejunostomy. Histopathological examination of gastric mass was suggestive of atypical carcinoid tumour of the stomach. He received three cycle chemotherapy post operatively. He had been followed up at six months interval.

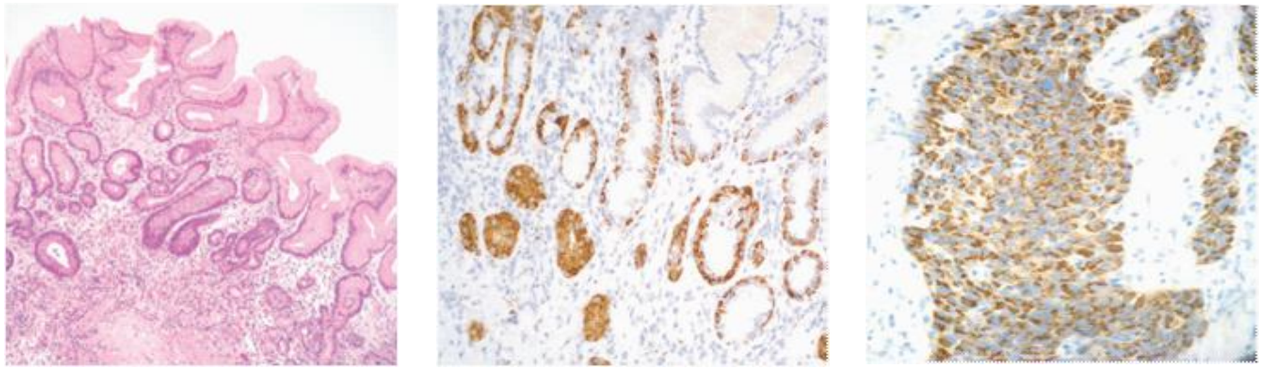

**Fig-3:** Histology slides photomicrographs of a poorly differentiated gastric carcinoid tumour stained with H & E chromogranin A and synaptophysin

## Discussion

Gastrointestinal carcinoids arise from neoplastic proliferation of enterochromaffin cells or Kulchitsky cell[3]. Incidence of carcinoid is estimated at 0.8-2.1 new cases per 100,000 people per year and majority are sporadic in nature[4]. In 2000, World Health Organization revised the classification of GEP-NET, abandoning the term carcinoid in favour of NET[5].

The age of presentation ranges from 4th to 8th decade[6]. The exact aetiology of carcinoid tumourogenesis is not known. Only a few risk factors for gastrointestinal (GI) carcinoid tumours are known e.g. genetic syndromes, Tuberous sclerosis, Von Hippel-Lindau disease, Multiple endocrine neoplasia type-I, Neurofibromatosis type-I. Carcinoid tumours are more common among African Americans than whites slightly, more common in women than men. Carcinoid tumours are classified, depending on the site of origin, as foregut (lung, thymus, stomach, duodenum) or midgut (distal ileum, proximal colon) or hindgut (distal colon, rectum)[7].

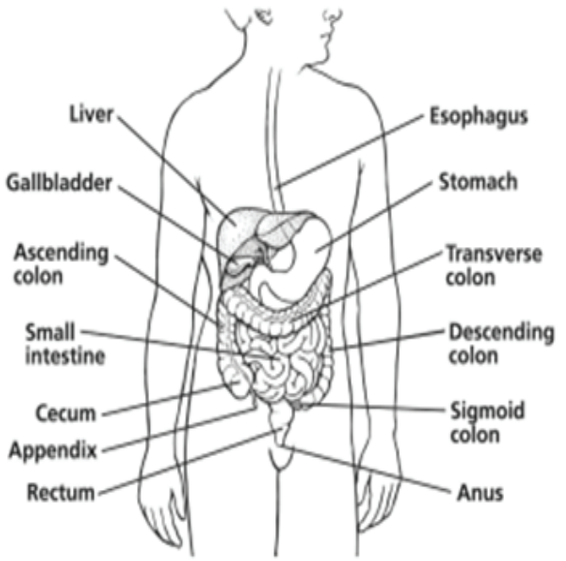


**Fig-4:** Sites of carcinoid tumour

In 28.5% cases it occurs in small intestine, 28% in bronchial system of the lungs,14% in rectum, 5-7% in colon, 5% in appendix, 4% in stomach, 1% in pancreas, 1% in liver and 8% in others. World Health Organisation histological classification of Gastrointestinal endocrine tumours[7,10].

(a) Well-differentiated endocrine tumour, Carcinoid: An epithelial tumour of usually monomorphous endocrine cells showing mild or no atypia and growing in the form of solid nests, trabeculae or pseudoglandulae, restricted to the mucosa or submucosa.

(b) Well-differentiated endocrine carcinoma, Malignant carcinoid: A malignant epithelial tumour of endocrine cells showing moderate atypia and growing in the form of solid nests, trabeculae or larger less well-defined cellular aggregates, which deeply invades the gut wall (muscularis propria or beyond) or shows metastases to regional lymph nodes or liver.

(c) Poorly differentiated endocrine carcinoma, Small cell carcinoma: A malignant epithelial tumour composed of highly atypical, small to intermediate-sized tumour cells growing in the form of large, ill-defined aggregates, often with necrosis and prominent angioinvasion and/or perineural invasion.

The biological and clinical characteristics of the tumours vary considerably. Most of the gastric carcinoids show no evidence of endocrine function[3]. They may synthesize and secret serotonin (5-HT) which is metabolized to 5-hydroxyindoleacetic acid (5-HIAA). Other hormones secreted are ACTH, histamine, dopamine, substance P, prostaglandins, kallikrein and tachykinins[8].

This patient presentation was associated with antral carcinoma stomach and histopathologically revealed atypical carcinoid tumour. He did not have any feature of carcinoid syndrome. The differential diagnoses of the syndrome include inflammatory bowel disease, gut neoplasia, diverticular disease and irritable bowel syndrome. Endoscopy and endosonography are common diagnostic tools. CT and MRI are required to assess the presence of metastases and plan of surgery[8]. Measurement of urinary 5-HIAA levels (73% sensitivity and 100% specificity) is used for confirming the diagnosis and monitoring patients with metastatic disease[8]. Other markers help in diagnosis are chromogranin -A, neuron-specific enolase, synaptophysin and CDX-2. Somatostatin receptor Scintigraphy, Gallium-68 receptor PET are newer modalities for diagnosis.

Surgery is the only curative therapy for NETs if performed before metastasis[2,9]. Surgical options are endoscopic mucosal resection, partial gastrectomy and total gastrectomy.

Non-surgical therapeutic options include:
(a) Chemotherapy: Drugs used Capecitabine, 5-fluorouracil (5-FU), Doxorubicin, Etoposide, Dacarbazine,Streptozocin, Temozolomide, Cisplatin. (b) Radiation therapy: It includes External beam radiation therapy (EBRT), Radioembolization. (c) Ablation: It may be cryotherapy where injecting liquid nitrogen through the needle to kill the carcinoid cells by freezing, Radiofrequency ablation uses high-energy radio waves released from the end of the needle which destroy the cancer cells with heat and percutaneous ethanol injection. (d) Drugs for treating carcinoid tumours e.g. Somatostatin analogue, Octreotide, Lanreotide, Interferons, Cyproheptadine.

The prognosis of the gastric carcinoids is significantly better than that of adenocarcinoma even in the presence of regional or hepatic metastasis[9]. The case represents poorly differentiated endocrine carcinoma which was associated with gastric carcinoid.

## Conclusion

Gastric carcinoids are increasingly being recognized, particularly because of the increasing use of diagnostic gastroscopy. There are abundant treatments for carcinoid tumours and syndrome though choice of treatment and their applications can be quite complex. Even though this is a rare disease, there are experts available who are interested and willing to help. A great deal of research is in progress which promises additional effective therapy in the foreseeable future.

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