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A RARE CASE OF GASTRIC CARCINOID TUMOUR TYPE II

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ABSTRACT

To report a case of gastric carcinoid tumour type II.
To radiologically investigate a case of 31 year old female who presented with complaints of epigastric pain, vomiting, weight loss, flushing sensation and diarrhoea. Patient had history of repeated iron deficiency anemia and on clinical examination a palpable lump was found in epigastrium.
Abdomen ultrasonography, Barium meal study and Upper gastro-intestinal endoscopy were performed.
Contrast enhanced multislice multidetector computed tomography scan (MDCT) of the abdomen was performed on a 128 multislice machine.
Radiological evaluation revealed; marked diffuse polypoidal enhancing wall thickening involving the entire stomach causing significant luminal narrowing with multiple nodular lesions and multiple peri-gastric, peri-pancreatic, pre/para-aortic enlarged enhancing lymphnodes.
Gastrointestinal carcinoids are well-differentiated endocrine neoplasms that belong to a diverse group of tumors that arise from cells of the diffuse endocrine system. They may produce specific syndromes such as Zollinger-Ellison syndrome, or they may occur in association with inherited syndromes such as multiple endocrine neoplasia type-I or neurofibromatosis type-I.

Introduction:
Gastrointestinal carcinoids are well-differentiated endocrine neoplasms that belong to a diverse group of tumours that arise from cells of the diffuse endocrine system. Carcinoids of the stomach, duodenum, and colon are uncommon. The majority of gastric carcinoids are ECL-cell carcinoids that arise from oxyntic mucosa in the gastric body and fundus. Three distinct sub-types of ECL-cell carcinoids are recognized: type I, which is associated with autoimmune chronic atrophic gastritis; type II, which is associated with Zollinger-Ellison syndrome (ZES) in patients with multiple endocrine neoplasia type I (MEN-I); and type III, which are sporadic carcinoids not associated with atrophic gastritis or hypergastrinemia. We present a case of type II gastric carcinoid, in which the diagnosis was given on MDCT and confirmed by intra-operative excision biopsy.

Material and Method:
Abdomen ultrasonography, barium meal study, contrast enhanced multislice multidetector computed tomography scan (MDCT) of the abdomen on a 128 multislice machine, upper gastro-intestinal endoscopy and PET-CT scan were performed in order to obtain maximum diagnostic output and minimize the dose of radiation.

Case report:
- A 31 year old female presented presented with complaints of epigastric pain, vomiting, weight loss, flushing sensation and diarrhoea. Complete blood count results revealed severe anemia with a hemoglobin level of 4.0 mmol/L (normal range, 7.1-9.9mmol/L) and hematocrit level of 0.26 (normal range, 0.34–0.48). Results of an iron study performed at an outside institution revealed iron-deficiency anemia; the patient had no history of heavy menstrual periods. Thyroid function test revealed raised serum PTH level of 1202pg/ml. Serum gastrin level were also raised measuring >10000 pg/ml.

Imaging findings:
- On abdomen ultrasonography, large hyperechoic mass lesion with thin anechoic slit like structure within, was seen in epigastrium and left hypochondriac region (Figure 1). The lesion showed minimal internal vascularity. Enlarged retroperitoneal lymph nodes (metastatic) may be visualized.

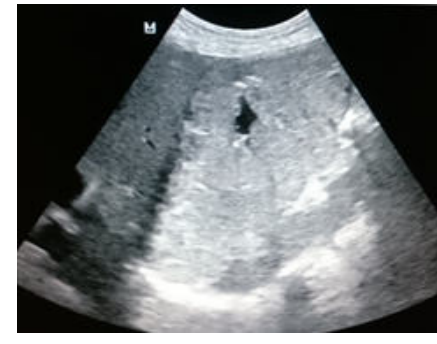

Figure 1: Abdomen ultrasound image shows large hyperechoic mass lesion (black arrows) with thin anechoic slit like structure within, in epigastrium and left hypochondriac region.

- Barium meal study was performed for further evaluation which showed narrow lumen of stomach seen as a thin streak of barium on single (air) contrast study and multiple polypoidal filling defects involving the lesser and greater curvatures of the stomach on double contrast study (Figure 2A and 2B). These filling defects were of variable size from 5mm to 50mm.

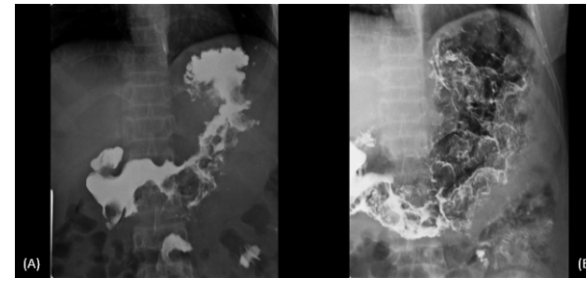

Figure 2: Barium meal study, (A) single contrast image shows narrowed lumen of stomach seen as a thin streak of barium and (B) double contrast image shows multiple

polypoidal filling defects involving the lesser and greater curvatures of stomach.

- Non enhanced CT and triple phase contrast enhanced CT scans of abdomen were also performed. The phases of imaging included unenhanced, arterial, and venous phases, with a 45-second delay from initiation of injection of the contrast material bolus for the arterial phase and a 65-second delay for the venous phase.
- NECT images showed markedly thickened gastric wall (Figure 3A and 3B). CECT arterial phase images showed multiple enhancing nodules lining the thickened rugal folds (Figure 3C and 3D). Portal venous phase images revealed that the folds are homogeneous in attenuation (Figure 3E and 3F). Multiple peri-gastric, peri-pancreatic, pre/para-aortic enlarged enhancing lymphnodes were also seen.

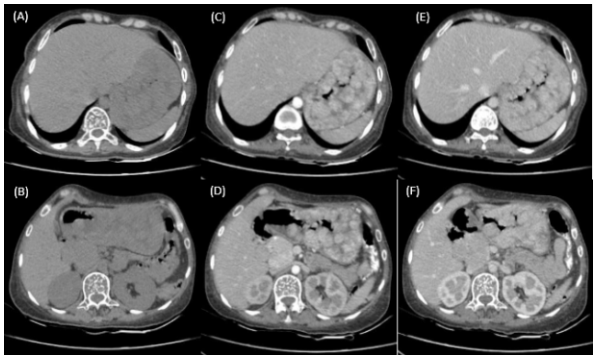

Figure 3: (A & B) NECT images show diffusely thickened gastric wall, (C & D) arterial phase images show multiple enhancing nodules lining the thickened rugal folds, and (E & F) portal venous phase images show that the folds are homogeneous in attenuation.

- Upper gastro-intestinal endoscopy revealed multiple 10-50 mm sized pedunculated and sessile polyps involving the lesser and greater curvatures of stomach with luminal narrowing. No site of active or recent bleeding was identified. (Figure 4)

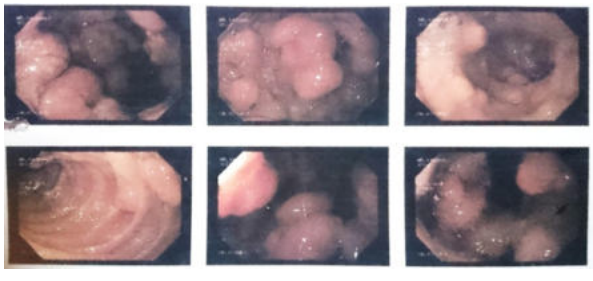

Figure 4: Upper gastro-intestinal endoscopy images show multiple 10-50 mm sized pedunculated and sessile polyps involving the lesser and greater curvatures of stomach with luminal narrowing. No site of active or recent bleeding was identified.

- Patient was undergone PET-CT scan, which showed increased FDG uptake as extensive enhancing polyps in stomach (Figure 5). PDG uptake was not seen at any other site in the body.

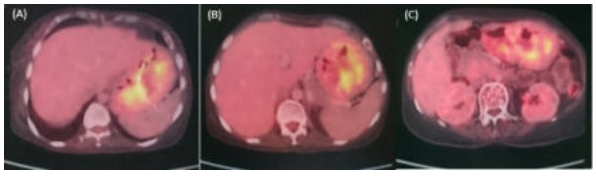

Figure 5 (A, B, C): PET-CT scan images show increased FDG uptake seen as extensive enhancing polyps in the stomach causing luminal narrowing.

- Gastro-duodenectomy and esophago-jejunostomy was performed followed by an excision biopsy and histologic analysis of the specimen revealed carcinoid tumor cells. (Figure 6)
- Patient was also diagnosed to have MEN – I syndrome.

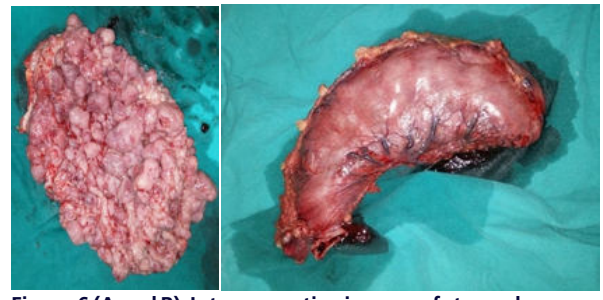

Figure 6 (A and B): Intra-operative images of stomach

Discussion:

- Gastric carcinoid tumors are rare, constituting less than 1% of gastric neoplasms (1) and approximately 8.7% of gastrointestinal carcinoids (2). The majority of gastric carcinoids are ECL-cell (enterochromaffin like cell) carcinoids that arise from oxyntic mucosa in the gastric body and fundus. Gastric carcinoids may be divided into three subtypes based on pathogenesis and histologic characteristics (3-5).

- TYPE – I : Most common and are associated with entero-chromaffin like cellular hyperplasia, hypergastrinemia, and chronic atrophic gastritis, with or without pernicious anemia (4).
- TYPE – II : Least common and account for 5%–10% of all gastric carcinoids. These tumors are caused by hypergastrinemia from a gastrin-producing tumor of the pancreas or small intestine and are seen in patients with MEN-I. Thus, patients with type II gastric carcinoids also have MEN-I or Zollinger-Ellison syndrome (4-5). ***Our case was type – II gastric carcinoid tumour with MEN –I.***
- TYPE – III : Not associated with hypergastrinemia or atrophic gastritis and represent about 13% of gastric carcinoids (4,6).

- Very rarely, non–ECL-cell carcinoids may occur within the stomach (eg, G-cell [gastrin cell] carcinoids, adrenocorticotrophic hormone–producing carcinoids, or EC-cell [enterochromaffin cell] serotonin-producing carcinoids).
- The differential diagnosis of multiple gastric carcinoid tumors includes hyperplastic polyps, adenomatous polyps, Peutz-Jeghers syndrome, and multiple hamartoma syndrome (Cowden disease).
- Type I ECL-cell carcinoids are the most common type in the stomach, representing 74% of gastric endocrine tumours. They occur most frequently in women (female-to-male ratio, 2.5:1) at a mean age of 63 years (10). Patients with type I ECL-cell carcinoids usually do not have clinical symptoms directly related to the tumors. The lesions are usually encountered during endoscopy performed for dyspepsia, anemia that may be due to chronic atrophic gastritis, or other reasons. Achlorhydria and, less commonly, pernicious anemia may be present. Hypergastrinemia or evidence of antral G-cell hyperplasia is usually observed (9).
- Type II gastric ECL-cell carcinoids account for 6% of gastric endocrine neoplasms, occur in patients at a mean age of 50 years, and have no gender predilection (10). Hypergastrinemia from a gastrin-producing endocrine neoplasm of the pancreas or the small intestine produces ZES in patients with MEN-1. Elevated serum levels of gastrin stimulate ECL-cell hyperplasia. In the setting of MEN-1, multiple carcinoids develop in ECL-cell hyperplasia. Because gastric carcinoids are rarely seen in patients with ZES alone, it is surmised that the genetic abnormalities related to altered tumor suppression in MEN-1 contribute to the development of carcinoids from ECL-cell hyperplasia. Clinically, elevated gastrin levels produce signs and symptoms of a hypertrophic, hypersecretory gastritis: abdominal pain or bleeding from multiple or recurrent peptic

ulcers, diarrhea, and elevated serum levels of gastrin.

- Type III gastric ECL-cell carcinoids represent 13% of gastric endocrine tumors and occur most commonly in men (male-to-female ratio, 2.8:1) at a mean age of 55 years (10). There is no associated hypergastrinemia or chronic atrophic gastritis. These patients typically present with signs and symptoms related to a solitary, aggressive mass and have no endocrine manifestations. Therefore, the clinical manifestations in these patients are similar to those in patients with other gastric neoplasms: bleeding, abdominal pain, anorexia, and weight loss.

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