International Surgery Journal
Vijay T et al. Int Surg J. 2023 Apr;10(4):791-794
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20231000

Case Report

Neuroendocrine carcinoma of stomach causing gastric outlet obstruction: a diagnostic dilemma

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Received: 17 February 2023
Accepted: 18 March 2023

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ABSTRACT

Gastric neuroendocrine tumour (GNET), also known as carcinoids, are a very rare cause of gastric outlet obstruction and arise from entero-chromaffin like cells of the mucosa. They account for 0.2/1000001 population and 8.7% of all Gastrointestinal carcinoids. They are four types based on the degree of differentiation, cell of origin and other pathologic features. Type 4 GNET are referred to as neuroendocrine carcinoma accounting for <1% of GNET. They are aggressive, poorly differentiated, locally invasive tumours with metastatic potential. Hence, they require an aggressive approach in terms of surgery and multimodality adjuvant treatment. Our patient is an elderly hypertensive who had complaints of projectile vomiting over ten days associated with weight loss over the last two years. A diagnosis of gastric outlet obstruction was made, and she underwent a contrast CT scan and endoscopy twice, both being suggestive of malignancy involving the antrum region however, the endoscopic biopsy showed unremarkable histology. Subsequently, she underwent emergency laparotomy in which there was a hard growth palpable in the antrum for which distal gastrectomy, D1 lymphadenectomy and gastrojejunostomy were done. Post-op HPE was suggestive of neuroendocrine carcinoma, thus, depicting our difficulty with diagnosis based on pre-op biopsy report given its rarity.

Keywords: Gastric outlet obstruction, Gastric carcinoid, Neuroendocrine carcinoma

INTRODUCTION

Gastric outlet obstruction (GOO) is a disorder that arises from any pathology causing a mechanical impediment to gastric emptying. It can either be benign or malignant.[4] The two commonest causes are gastric adenocarcinoma and pyloric stenosis.[1] Among the rare causes are gastric carcinoids accounting for <1% of all cases. Gastric neuroendocrine tumours (GNET) are mucosal tumours that arise from Enterochromaffin cells secreting histamine, bradykinin, 5-HT, gastrin and somatostatin. According to the WHO classification, these are subdivided into four distinct types based on their degree of differentiation, cell of origin and other clinical features. Type 1-type 3 arises from ECL cells whereas type 4 GNET referred to as neuroendocrine carcinoma arises from other types of endocrine cells and consists of poorly differentiated carcinomas. Type 4 tumours are not associated with hypergastrinemia, are ECL independent and are highly aggressive tumours with the majority of the patients presenting with metastasis at the time of presentation often behaving like adenocarcinomas and hence managed as aggressively with surgical resection followed by adjuvant multimodality therapy.[3]

We are reporting the case of an 80-year-old lady who presented with a classic history and examination findings of GOO with CT and endoscopy confirming the same, however, her endoscopic biopsy showed no remarkable histology. Given her age and symptoms, she was managed on the lines of malignant obstruction and despite her biopsy report, she was taken up for emergency laparotomy and distal gastrectomy with D1 lymphadenectomy and gastrojejunostomy was done. The postoperative biopsy

report concluded it to be a case of neuroendocrine carcinoma, hence confirming our suspicion of malignancy and aiding our diagnostic confusion based on the preoperative biopsy report.

CASE REPORT

An 80-year-old female presented to us with complaints of vomiting over the course of 10 days which was projectile, non-bilious, non-blood tinged, with multiple episodes, containing foul smelling, undigested particles after a few hours of consumption of solid food, being only able to consume small amounts of liquids, associated with weight loss over 2 years and burning retrosternal sensation with the use of antacids for 2 years. She is a known hypertensive on medication and known tobacco chewer for the past 50 years.

On initial examination, she had stable vitals, and her BMI was 19.2. Abdominal examination showed a distended stomach in the epigastrium with positive succussion splash and bowel sounds were present, 2-3/min. The rest of the examination was unremarkable. Blood investigation showed anaemia with hypoalbuminemia while arterial blood gas analysis showed the presence of hypokalemia, hypochloremia, and metabolic alkalosis in line with our suspicion.

Imaging findings

Ultrasound abdomen revealed concentric wall thickening in the pylorus of the stomach and proximal duodenum maximum of 15 mm. Contrast CT of chest and abdomen revealed circumferential homogenously enhancing wall thickening involving antropyloric region of the stomach causing luminal narrowing with few perigastric lymph nodes and ill-defined fat planes with pancreas likely suggestive of malignancy with few osteosclerotic bony lesions involving vertebral bodies, ribs, pelvis, clavicle likely metastatic. She underwent endoscopy two times. Her first endoscopy report showed the presence of ulcero-proliferative growth in the pre-pyloric region with reflux esophagitis and stale liquid matter in the stomach.

Our endoscopy showed altered mucosa in the stomach with distorted rugae and edematous thickened pylorus of the stomach likely malignant, with the presence of a polyp-like lesion in the pre-pyloric region/antrum. However, biopsies taken both times from the body of the stomach, pre-pyloric antrum and polyp yielded negative results for malignancy. She was planned for PETCT because of suspicious bony lesions, however, the patient refused to undergo the same opting for only surgical management further and hence the patient was taken up for surgery as an emergency given gastric outlet obstruction.

Intra-operative findings

Exploratory laparotomy with D1 distal gastrectomy and anterior ante-colic isoperistaltic loop gastrojejunostomy and subhepatic drain was inserted. The operative finding was a hard growth palpable at the antropyloric region of the stomach with no ascites or any other visible liver or peritoneal metastasis.

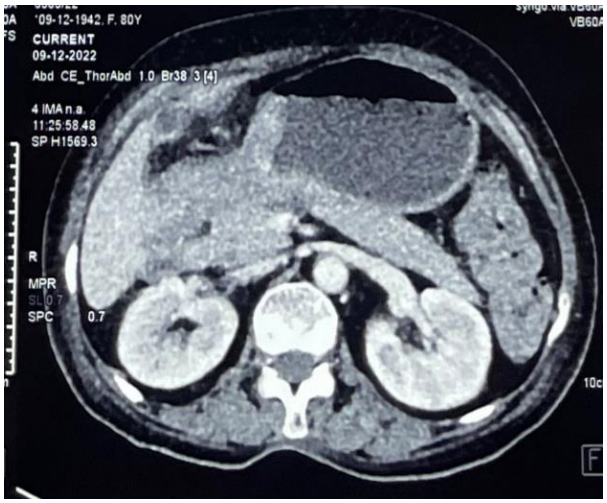

Figure 1: CT scan showing diffuse thickening in the antropyloric region.

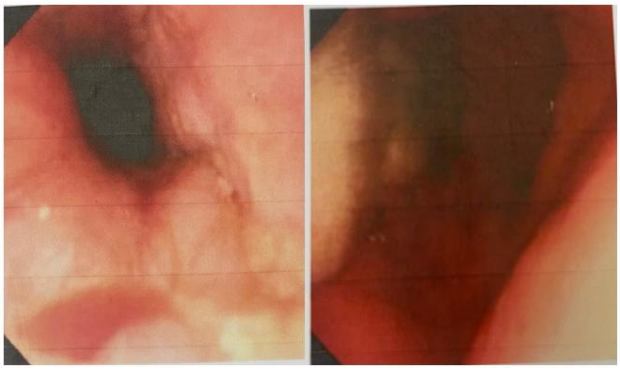

Figure 2: Endoscopy showing pyloric thickening with polyp.

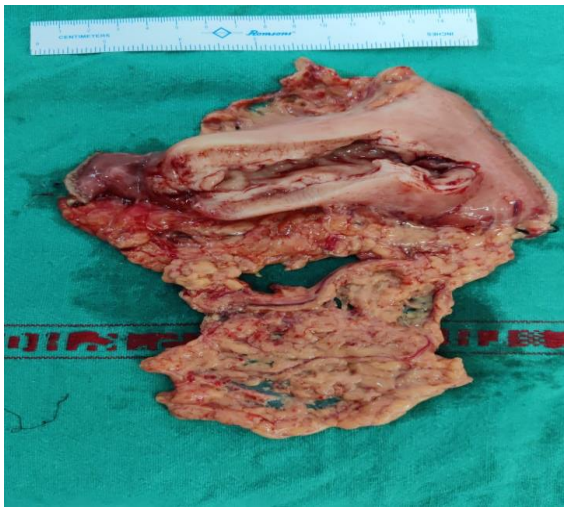

Figure 3: Intra-op specimen showing resected distal stomach with growth in the pre-pyloric region.

Post-operative course

The patient's post-operative period was uneventful, and she improved symptomatically, being able to take solids as well as liquids. On histopathological examination, the tumour site was the body and antrum of the stomach with poorly differentiated neuroendocrine carcinoma invading up to muscularis propria, perineural invasion and negative margins, 5/7 regional lymph nodes involved pT2N2Mx.

She underwent a PET scan after one month which showed DOTA noc avid sclerotic skeletal metastasis involving bilateral humeri, clavicles, scapulae, ribs, and pelvic bones, with right proximal femoral and multiple vertebral body lesions. The patient was referred to medical oncology for palliative chemotherapy with somatostatin analogues.

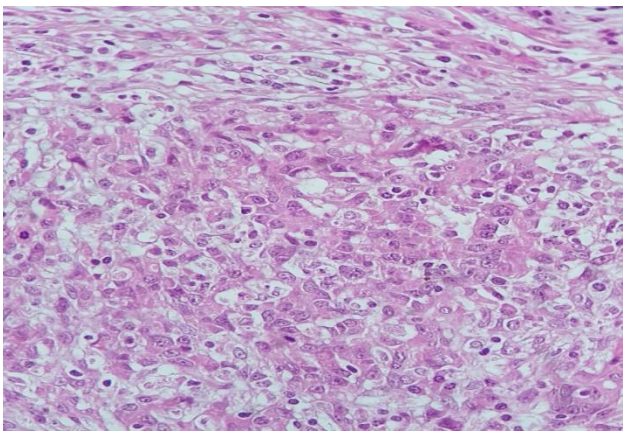

Figure 4: Post-op HPE showing neuroendocrine carcinoma.

DISCUSSION

GNET account for 2.4-8.7% of all gastrointestinal carcinoids. Their incidence however is on the rise in the past few decades because of the increasing use of endoscopy and increased awareness of this disease. The tumours arise from different types of endocrine cells that are situated in the stomach with the majority arising from Enterochromaffin cells in the mucosa that mainly secretes histamine. However, they can also arise from the D cells secreting somatostatin and G cells secreting gastrin (gastrinoma). They occur more commonly in females than males and in the older age group.[3] They have historically been classified into four subtypes based on their clinicopathologic features.[5]

Type1 GNET is well-differentiated tumours that account for 75% of cases. They are characterized by their association with chronic atrophic gastritis and hypergastrinemia.[3,5] They arise from ECL cells, are multiple, very rarely metastasize, have a low ki-67 index, are usually present in the body of the stomach, and hence are the most benign type with an excellent prognosis.[6,7]

Type 2 GNET is like type 1 GNET except that these are mostly seen in patients with MEN1 syndrome or Zollinger Ellison syndrome and account for only 5% of all cases. These tumours. Are also small, multiple, and well-differentiated but their prognosis is dependent on the presence of other tumours in MEN syndrome rather than the carcinoid itself.[3,5]

Type 3 GNET are different in that they are not gastrin dependent, often single and large with the presence of normal surrounding mucosa. Their cell of origin is also ECL cells and accounts for 20% of all cases. They may be well differentiated or poorly differentiated, associated with metastasis at the time of presentation and hence, have a poor prognosis.[8]

Type 4 GNETs are the rarest of all the GNETs and account for<1% of all gastric carcinoids. The tumours are gastrin independent and arise from other endocrine cells that secrete 5-HT, gastrin, and ACTH and are poorly differentiated carcinomas being single, large, and highly malignant. They are often associated with a state of parietal cell hyperplasia and can be located anywhere in the stomach. These tumours have a very high ki-67 proliferation index, high mitotic count, and high grade and are associated with angioinvasion and local invasion with the highest incidence of metastasis at the time of presentation.[3,5] Thus, this portends it the worst prognosis of all these tumours with a mean survival reported to be 6-14 months.[8] The presentation of these patients cannot be differentiated from gastric adenocarcinoma with weight loss, postprandial epigastric pain, and non-bilious projectile vomiting related to gastric outlet obstruction.[3] A patient may also present with features of carcinoid syndrome in the event of hepatic metastasis consisting of four cardinal signs: bronchospasm, flushing, sweating and restrictive cardiomyopathy.[1] Examination findings may reveal the presence of a distended stomach suggested with succussion splash, visible gastric peristalsis and auscultopercussion supplemented by the saline load test.[4] There may be the presence of a palpable liver in the presence of metastasis. Diagnosis is usually established by endoscopic biopsy of the largest polyp and taking 6 samples from the fundus and the antrum along with a biopsy of the normal mucosa to look for the presence of atrophy. Staging of the carcinomas requires Contrast CT to look for the presence of distant metastasis.[3]

While type1 and type 2 GNETs behave as benign carcinoids and usually require surgical resection, type 3 and 4 GNETs are usually aggressive and hence, require a multidisciplinary approach consisting of aggressive surgical resection with negative margins, formal lymphadenectomy, and assessment of metastasis. A patient who has only liver metastasis can be managed with surgical resection of primary with hepatectomy/ adjuvant treatment but those who have a widely disseminated metastasis should undergo resection of primary only for symptomatic relief.[3,5]

Our patient presented with a typical picture of gastric adenocarcinoma in terms of history, examination, blood analysis and radiographic findings. Endoscopy was done twice, revealing a growth-like thickening in the pyloric region of the stomach and a polyp in the pre-pyloric region/antrum. Although multiple biopsies were taken both times she underwent this procedure, they yielded negative results for malignancy. We decided to go ahead with the surgery because of gastric outlet obstruction and our clinical suspicion of malignancy given the advanced age of the patient. Post-operative HPE confirmed our suspicion but turned out to be an extremely rare neuroendocrine carcinoma infiltrating into muscularis propria, poorly differentiated with perineural invasions, negative margins, 5/7 lymph nodes+, and tumour immunohistochemistry for chromogranin, synaptophysin, CK7 + and ki-67= 60-70%, PT2N2M1.

CONCLUSION

Neuroendocrine carcinoma is a very rare cancer that accounts for less than <1% of gastric carcinoids. Our patient represents one of the rare cases of GNET and highlights the importance of clinical and radiologic correlation with preoperative biopsy reports for appropriate management.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Stechman MJ, Scott-Coombes DM. The adrenal glands and other abdominal endocrine disorders. Bailey and Love's Short Practice of Surgery. In: Connell PR, McCaskie AW, Sayers RD, eds. 28th ed. Boca Raton: CRC Press; 2023.
2. Modlin IM, Lye KD, Kidd M. A 5-decade analysis of 13,715 carcinoid tumors. Cancer. 2003;97(4):934-59.
3. Linda B, McFadden DW. Gastrointestinal Carcinoid Tumors. Shackelford's surgery of the alimentary tract. In: Yeo CJ, eds. 8th ed. Philadelphia, PA: Elsevier; 2019.
4. Kumar A, Annamaraju P. Gastric Outlet Obstruction. Statpearls; 2022.
5. Li TT, Qiu F, Qian ZR, Wan J, Qi XK, Wu BY. Classification, clinicopathologic features and treatment of gastric neuroendocrine tumors. World J Gastroenterol. 2014;20(1):118-25.
6. Ahlman H, Kölby L, Lundell L, Olbe L, Wängberg B, Granérus G, et al. Clinical management of gastric carcinoid tumors. Digestion. 1994;55(3):77-85.
7. Kaizaki Y, Fujii T, Kawai T, Saito K, Kurihara K, Fukayama M. Gastric neuroendocrine carcinoma associated with chronic atrophic gastritis type A. J Gastroenterol. 1997;32(5):643-9.
8. Hosoya Y, Nagai H, Koinuma K, Yasuda Y, Kaneko Y, Saito K. A case of aggressive neuroendocrine carcinoma of the stomach. Gastric Cancer. 2003;6(1):55-9.

Cite this article as: Vijay T, Saurabh G, Singh R. Neuroendocrine carcinoma of stomach causing gastric outlet obstruction: a diagnostic dilemma. Int Surg J 2023;10:791-4.