

Unusual Presentation of a Neuroendocrine Tumor of Stomach as an Ulcer

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Abstract

Background: Gastric neuroendocrine tumors (G-NETs) originate from the enterochromaffin like (ECL) cells of the gastric mucosa and form less than 1% of all gastric tumors. These present as single to multiple polyps on endoscopy. **Case Report:** A 52-year-old female patient found to have an ulcer on esophagogastroduodenoscopy (EGD) which was reported as chronic gastritis on biopsy. She underwent distal gastrectomy with Billroth II anastomosis due to suspicion of malignancy only to be diagnosed as a grade II gastric neuroendocrine tumor on final histopathological examination. **Conclusion:** Majority of G-NETs are detected incidentally on endoscopy or present with gastrointestinal bleeding, anemia, fatigue, epigastric pain and vomiting. One should be mindful of this unusual presentation of a G-NET as a gastric ulcer and further evaluation should be considered if there is suspicion of malignancy even after endoscopic biopsy.

Keywords: Gastric Ulcer, Neuroendocrine Tumors, Peptic Ulcer, Stomach Neoplasms, Surgical Anastomosis.

Introduction

Gastric neuroendocrine tumors (G-NETs) originate from the enterochromaffin like (ECL) cells of the gastric mucosa form less than 1% of all gastric tumors and about 6-8% of all neuroendocrine tumors (NETs) [1]. G-NETs have been classified into three subtypes. Type I arise in the presence of chronic atrophic gastritis and is the most common (70-80%). Type II is seen in association with gastrinomas and multiple endocrine neoplasia (MEN) syndrome (5-10%). Type III is sporadic in occurrence (10-15%) and has the highest malignant potential [2-4]. G-NETs are mostly asymptomatic or can present with gastrointestinal (GI) bleeding, anemia, fatigue, epigastric pain and vomiting [3,5]. They are seen as small to large, single to multiple polypoidal lesions on endoscopy.

This is an unusual case of G-NET presenting as an ulcer along the greater curvature of the stomach presenting with pain and misdiagnosed as chronic gastritis on endoscopic biopsy.

Case Report

A 52-year-old female presented to our hospital with complaints of epigastric pain for 1 year. She did not have any known co-morbidities. Her general and systemic physical examination was unremarkable. She underwent an esophagogastroduodenoscopy (EGD) which revealed chronic gastritis and an ulcer measuring around 2×2 cm on the greater curvature of the gastric body [Fig.1]. An adequate biopsy sample was obtained from the ulcer and histopathological examination of the same showed features of chronic gastritis. We did not have facilities for endoscopic ultrasound (EUS) and EUS guided biopsy at our centre. A contrast enhanced computerized tomographic (CECT) scan of the abdomen did not reveal any significant abnormalities. Based on the aggressive appearance of the ulcer and clinical history of patient, intractable ulcer to medical therapy for a long duration and with a high degree of suspicion for malignancy, decision was taken to perform gastric

resection. She underwent distal gastrectomy with Billroth II anastomosis.

Gross examination showed an ulcer of 1.3 cm in diameter along the greater curvature of stomach [Fig.2]. Histopathological examination showed strips of gastric mucosa with underlying tissue showing tumor cells, with scanty cytoplasm and round to oval nuclei with stippled chromatin, arranged in nests and organised pattern [Fig.3]. The mitotic index was 0-2/10 HPF. Immunohistochemical analysis revealed cells expressing synaptophysin and CD56 positivity and Ki-67 index up to 5%. These features were consistent with neuroendocrine tumor (WHO grade 2).

The post-operative recovery was uneventful and patient was referred to medical oncology for adjuvant chemotherapy. She was recurrence free and tolerating the treatment well at 6 months follow up.

Discussion

Neuroendocrine tumors are epidermal tumors characterized by both endocrine and neural differentiation and these can arise from any part of the body. Gastric NETs (G-NETs) are known to originate from the enterochromaffin like (ECL) cells of the gastric mucosa [1]. G-NETs tend to be asymptomatic in many cases and are detected incidentally on endoscopic evaluation. They can present with GI bleeding, anemia, fatigue, epigastric pain and vomiting. Rarely they can present with carcinoid syndrome due to histamine release [3,5]. They are seen as small to large, single to multiple polypoidal lesions on endoscopy. Work-up for diagnosis and staging of G-NETs include EGD, endoscopic ultrasound and CECT scan to accurately determine the extent and depth of the lesion and lymph nodal involvement. Newer modality of imaging includes somatostatin receptor scintigraphy (SRS) with labelled octreotide that can help detecting primary as well as metastasis [3,4].



Fig.1: Ulcer measuring around 2×2 cm on the greater curvature of the gastric body.



Fig.2: Resected surgical specimen showing gastric body with ulcer (black arrow).

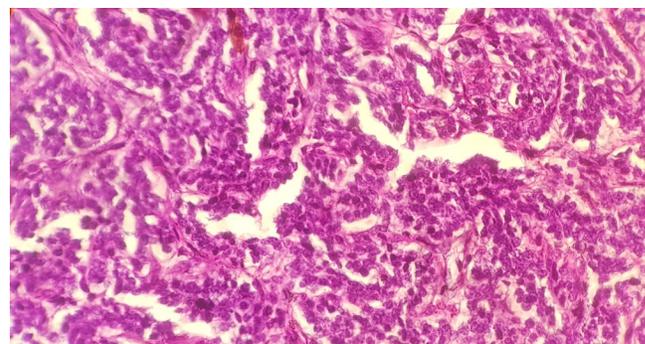


Fig.3: Histopathological examination (40× magnification) showing polygonal tumor cells with scanty cytoplasm and round to oval nuclei with stippled chromatin, arranged in nests and organised pattern.

Chromogranin A is the most useful plasma marker of NET used for follow up [3].

On histopathology, G-NETs appear as small round to polygonal cells with central nuclei and fine chromatin, small nucleoli and varying amount of cytoplasm with few mitosis [3]. Synaptophysin and chromogranin A staining is used for neuroendocrine phenotype confirmation. Ki-67 is recommended as a proliferative index marker for prognostication and management [5,6]. Factors such as perineural and angioinvasion, Ki-67 index of >3%, high mitotic count and cellular atypia are features of a malignant variety of NET [3,7]. WHO classifies G-NETs into low (G1), intermediate (G2) and high grade (G3) based on mitotic count and Ki-67 index [7,8].

The European Neuroendocrine Tumor Society (ENETS) guidelines recommend endoscopic resection of superficial type I G-NETs <2 cm [6,9]. Type II tumors tend to be more aggressive and management consists of endoscopic resection for early tumors smaller than 2 cm and polypectomy or gastric resection for larger and multiple tumors. Type III are treated with total or partial gastrectomy [3,4]. Gastric resection is the gold standard of treatment for resectable G-NETs and has been shown to have survival benefit even among patients with metastatic disease [1]. Metastatic tumors presenting with poorly differentiated histology are treated with platinum based chemotherapy [4]. Recent literature also describes type IV G-NETs which are large in size, poorly differentiated on histology and have a very high malignant potential with poorest prognosis of all. These are treated with radical surgery and chemotherapy [5,10].

In our case, typical endoscopic findings of NETs such as polypoidal growth with or without ulceration were not observed. Even endoscopic biopsy did not yield the correct diagnosis. We were limited by absence of EUS guided FNAC facilities at our centre and the patient underwent

surgery based on clinical judgement. This case highlights an atypical presentation of G-NET with an ulcerated lesion which is rarely seen.

Conclusion

Gastric neuroendocrine tumors are rare and mostly present with polypoidal lesions. Here we report a case of an ulcerated G-NET which was managed with gastric resection with favourable outcome. This presentation should be considered with high index of suspicion when evaluating a gastric ulcer clinically suggestive of neoplastic etiology.

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