Myoclonic Jerks and Chronic Diarrhea; The Challenges of Zollinger-Ellison Syndrome

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INTRODUCTION

Zollinger-Ellison Syndrome (ZES) is a rare disorder caused by hypersecretion of gastrin, leading to peptic ulcer disease (PUD) and chronic diarrhea. In most cases, inappropriate gastrin secretion is from duodenal or pancreatic neuroendocrine tumors (gastrinomas). Diagnosis of gastrinoma can be delayed as the disease has nonspecific symptoms and can be misdiagnosed as other GI disorders. [1]

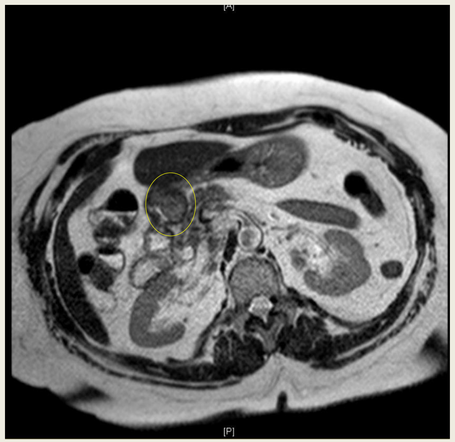

Figure 1 2.5 x 2 x 2.5 cm mass-like thickening in the distal gastric antrum near the first portion of the duodenum

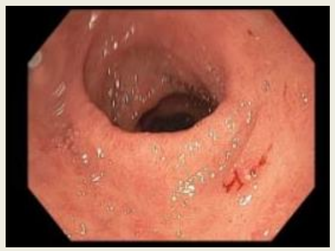

Figure 2 Ulcers in the bulb of duodenum

CASE PRESENTATION

A 79 year-old female with a history of type 2 diabetes mellitus, hypertension, esophagitis, and peptic ulcer disease was admitted due to myoclonic jerk movements in the whole body and watery diarrhea. She was found to have severe hypomagnesemia and acute kidney injury (AKI). Notably, the patient had a history of chronic diarrhea for several months with several admissions due to hypomagnesemia, hypocalcemia, and AKI. Previous chronic diarrhea workup including celiac serology, Vasoactive intestinal peptide test (VIP) level, fecal calprotectin, and colonoscopy were inconclusive. She had been started on Lomotil with a possible diagnosis of Irritable Bowel Syndrome (IBS) with partial response.

On this admission, the stool osmolar gap was less than 50, chromogranin A level was increased to 8224 ng/mL and gastrin was 596 pg/mL. Due to AKI abdominal CT with contrast could not be performed. She underwent MRI/MRCP which showed a 2.5 cm mass-like thickening with restricted diffusion in the distal gastric antrum. Esophagogastroduodenoscopy (EGD), showed gastritis and non-bleeding duodenal ulcers. EUS was performed for her, but FNA was inconclusive. She underwent an Octreotide scan which was compatible with a somatostatin receptor-bearing neuroendocrine tumor (NET) in the distal stomach. The patient underwent laparoscopic distal gastrectomy, with reconstruction and peri-pancreatic LN resection for neuroendocrine tumor after the hospitalization. Histopathology confirmed a well-differentiated neuroendocrine tumor with metastasis to the lymph node. Her diarrhea resolved and chromogranin A and gastrin normalized, 90 ng/mL and 43 pg/mL respectively.

DISCUSSION

Chronic diarrhea is one of the main manifestations of gastrinoma. Still, the diagnosis can be challenging as many different disorders can cause development of diarrhea and also due to the rarity of this syndrome.[2] Many patients are diagnosed and treated for more common conditions such as IBS and GERD. An initial evaluation in suspected cases should include Gastrin level. Gastrin level can be falsely elevated in the patients with chronic PPI use and makes the assessment difficult as patients with PUD need to be off the medication for two weeks.

Most gastrin-producing tumors arise from duodenum (75%) and pancreas (20%).[3] Less than 5% of the patients have a tumor in other locations such as stomach, peri-pancreatic lymph nodes, liver, bile duct, or ovary. EGD can show prominent gastric folds, and pathology can be nonspecific. In patients with chronic diarrhea and suspected ZES, chest and abdominal imaging is warranted to identify intra- or extra-abdominal lesions. Octreotide scan is a good tool for localizing the tumor's primary location and also metastasis to the lymph nodes. ZES diagnosis consideration in patients with chronic diarrhea and timely evaluation and treatment can prevent the complications of the syndrome such as electrolyte abnormality, PUD, and its complications as well as the risk of metastasis.

REFERENCES

1 Gibril F, Jensen RT. Zollinger-Ellison syndrome revisited: diagnosis, biologic markers, associated inherited disorders, and acid hypersecretion. Current Gastroenterology Reports. 2004 Dec;6(6):454-63.
2 Arasaradnam RP, Brown S, Forbes A, Fox MR, Hungin P, Kelman L, Major G, O'Connor M, Sanders DS, Sinha R, Smith SC. Guidelines for the investigation of chronic diarrhoea in adults: British Society of Gastroenterology. Gut. 2018 Aug 1;67(8):1380-99.
3 Rossi RE, Elvevi A, Citterio D, Coppa J, Invernizzi P, Mazzaferro V, Massironi S. Gastrinoma and Zollinger Ellison syndrome: A roadmap for the management between new and old therapies. World Journal of Gastroenterology. 2021 Sep 9;27(35):5890.