

Question 37– Week of April 22

A 45 year old man presents with persistent epigastric pain. His bloodwork is remarkable for hypercalcemia and he has an upper endoscopy which reveals thickening of the gastric folds in fundus and corpus. He has never taken a PPI. The patient's gastroenterologist obtains a gastrin level which is 934 pg/ml. An CT is performed showing multiple small duodenal wall defects consistent with possible gastrinoma. Upon further questioning the patient notes that his mother and maternal grandmother also had gastrinomas and other tumors but he is unsure of the types. Which genetic syndrome does this patient most likely have?

- A. MEN 1a
- B. MEN 2a
- C. MEN 2b
- D. Lynch Syndrome

Answer: A

The patient's most likely diagnosis is Zollinger-Ellison Syndrome secondary to a gastrinoma. Gastrinomas are frequently found in the duodenum but might also be seen in the pancreas, lymph nodes and other organs. 60% of patients with MEN1 are found to have gastrinoma. Classically, MEN1 patients have hyperparathyroidism (elevated calcium levels), pituitary lesions (Prolactinomas are most common) and pancreaticoenteric lesions. The gastrinomas associated with MEN are frequently small in size, easily missed on EGD, and multifocal. ZES has a slight male predominance and is most commonly diagnosed in patients aged 40-50. The differential diagnosis for thickened gastric folds includes: Menetrier's disease, gastric neoplasm (lymphoma, carcinoma), granulomatous gastritides, gastric varices, infectious gastritis (particularly H. pylori and CMV), eosinophilic gastritis. The target of gastrin are the parietal cells of the stomach. Gastrin is trophic for oxyntic glands, thickens mucosal layer of the stomach, trophic for enterochromaffin like cells resulting in hyperplasia of the fundus and corpus (thickening the folds in that region). Gastrinoma's are most frequently seen in the Duodenum (~50% of the time) and less frequently in the pancreas (~20%).

Reference:

Chambers AJ, Pasiaka JL. Gastrinoma. Cancer Treatment and Research. 2010; 153: 213-233.