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GASTRINOMA: IMPORTANCE OF LOCALIZATION. ANATOMICAL, SURGICAL AND DIAGNOSTIC PERSPECTIVE

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ABSTRACT

Gastrinoma is also commonly called as Zollinger-Ellison syndrome. It accounts for about 15-20% of Pancreatic Neuroendocrine Tumors. In These tumors There is excessive and overproduction of the hormone gastrin, leading to excessive secretion of gastric acid. The patients suffer from multiple and recurrent peptic ulcers that are in most cases resistant to medical treatment or that are in uncommon locations. In most cases these tumors are small, having a distribution that is diverse and difficult to detect complicated with the fact that patients present with varied clinical signs and not detected or diagnosed at early stages .As such a high suscpicion about Gastrinoma should always be there once a patient fails to respond to proton pump therapy in case of refractory peptic ulcer disease . Despite extensive preoperative localization studies and meticulous surgical exploration, some patients with gastrinoma have no tumor demonstrable at laparotomy.

INTRODUCTION

Gastrinomas are rare tumors in patients with intractable peptic ulcer disease, recurrent ulceration, ulceration at atypical sites and failure to respond to medical therapy. The article presents and anatomical, surgical and diagnostic perspective of these rare tumors which can undergo a malignant transformation

DISCUSSION

Anatomical and Surgical Perspective

Following the first report of an islet cell tumor of the pancreas associated with peptic ulcer disease by Sailer and Zinninger in 1946, Zollinger and Ellison in 1955 described two patients with florid peptic ulcer disease and pancreatic islet cell tumors. The diagnostic triad proposed for this syndrome at that time included

- the presence of primary peptic ulcerations in unusual locations
- gastric hypersecretion of gigantic proportions that persists despite adequate therapy, and
- the identification of an islet cell tumor of the pancreas. In the intervening decades, much information has accumulated regarding these gastrin-secreting tumors and the associated Zollinger-Ellison syndrome.

The most common symptoms of ZES are epigastric pain, GERD, and diarrhea. More than 90% of patients with gastrinoma have peptic ulcer. Most ulcers are in the typical location in proximal duodenum, but atypical ulcer location such as distal duodenum, jejunum, or multiple ulcers should prompt an evaluation for gastrinoma. Gastrinoma also should be considered in the differential diagnosis of recurrent or refractory peptic ulcer, secretory diarrhea, gastric rugal hypertrophy, esophagitis with stricture, bleeding or perforated ulcer, familial ulcer, peptic ulcer with hypercalcemia, and gastric carcinoid. The Gastrinoma triangle is an anatomical area in the the abdomen where most of the gastrinomas are throught to arise from. The boundaries are defined as the confluence of the cystic and common bile ducts superiorly, the second and third portions of the duodenum inferiorly, and the neck and body of the pancreas medially (Horton *et al*., ?; Stabile *et al*., 1984; Yu *et al*., 2004). Gastrinomas found in this triangle include those in the head of the pancreas, duodenal gastrinoma, rare isolated nodal gastrinoma, and rare antral tumors. The Gastrinoma syndrome can exist in multiple forms,

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including benign sporadic, malignant metastatic, and as part of the MEN-I syndrome. Most of the gastrinomas are sporadic (Kulke *et al*., 2012) and occur primarily in the gastrinoma triangle. Although they can occur in the pancreas, the duodenum has been shown to be the most common site of gastrinomas, based on the pioneering work of Debas and colleagues (Rehfeld *et al*., 2011) Thompson and colleagues (Strosberg *et al*., 2011) and others. Duodenal wall gastrinomas have been identified in more than half of patients (Norton *et al*., 2011; Grobmyer *et al*., 2009). Sporadic tumors occurring in the pancreas tend to be solitary. Primary tumors may also occur in a variety of ectopic sites, including the body of the stomach, jejunum, peripancreatic lymph nodes, splenic hilum, root of the mesentery, omentum, liver, gallbladder, common bile duct, and the ovary (Goudet *et al*., 2010; Lopez *et al*., 2012; Dickson *et al*., 2011; Glazer *et al*., 2010). About 30-35% of gastrinomas are associated with the MEN-I syndrome (Turner *et al*., 2010; Raymond *et al*., 2011; Yao *et al*., 2010; Yao *et al*., 2011). The tumors are usually multiple, and often small and undetectable. Most cases are discovered at a younger age than in sporadic cases. The likelihood of surgical cure in MEN-I patients is considerably less than in sporadic patients, unless a specific site or sites responsible for the hypergastrinemia has been identified preoperatively.

Radiological Perspective

Eighty percent of primary tumors are found in the gastrinoma triangle and many tumors are small (<1 cm), making preoperative localization difficult. Transabdominal ultrasound is quite specific, but not very sensitive. CT will detect most lesions >2 cm in size and MRI is comparable. EUS is more sensitive than these other noninvasive imaging tests, but it still misses many of the smaller lesions, and may confuse normal lymph nodes for gastrinomas. Currently, the preoperative imaging study of choice for gastrinoma is somatostatin receptor scintigraphy When the pretest probabilitof gastrinoma is high, the sensitivity and specificity of this modality approach 100%. Gastrinoma cells contain type 2 somatostatin receptors that bind the indium-labeled somatostatin analogue (octreotide) with high affinity, making imaging with a gamma camera possible. Currently, angiographic localization studies are infrequently performed for gastrinoma. Both diagnostic angiography and transhepatic selective venous sampling of the portal system have been supplanted by selective secretin infusion, which helps to localize the tumor as inside or outside the gastrinoma triangle.

Conclusion

At present, the cause of death in the majority of gastrinoma patients is tumor growth and dissemination. Chemotherapy, hormonal therapy with octreotide, hepatic transplantation, hepatic embolization, and interferon have all been used to treat patients with unresectable or metastatic gastrinoma. As such the localization of Gastrinonas is very important. A thorough intraoperative exploration of the gastrinoma triangle and pancreas is essential, but other sites (i.e., liver, stomach, small bowel, mesentery, and pelvis) should be evaluated as part of a thorough intra-abdominal evaluation to find the primary tumor, which is usually solitary. The duodenum and pancreas should be extensively mobilized and intraoperative ultrasound should be used. Intraoperative EGD with transillumination should be considered. If the tumor cannot be located, generous longitudinal duodenotomy with inspection and palpation of the duodenal wall should be considered. Lymph nodes from the portal, peripancreatic, and celiac drainage basins should be sampled. Ablation or resection of hepatic metastases should be considered.

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