

# Answer

## Wermer Syndrome and Zollinger-Ellison Syndrome

**Z**ollinger-Ellison Syndrome (ZES) is characterized by ulcer disease and dyspepsia due to inappropriate secretion of gastrin by neuroendocrine tumors called gastrinomas. Gastrinomas represent only a small fraction of pancreatic and duodenal tumors. They may occur sporadically (75%) or in association with genetic endocrine disorders such as MEN I (25%), also known as Wermer syndrome.<sup>1</sup> Differences in the pathogenesis of gastrinomas in these 2 subtypes have implications for presentation, treatment, and survival.

Patients with MEN I often present with ZES at a younger age, typically in the fourth decade of life.<sup>1,2</sup> The diagnosis is based on elevated gastrin levels in the presence of gastric hyperacidity. Secretin stimulation of gastrin levels may be required to diagnose ZES. The severity of symptoms and degree of gastrin level elevation does not differ by tumor location or by the subtype of disease.

In patients with ZES and proved or suspected MEN I, serum calcium and parathyroid hormone levels often may be elevated. If hyperparathyroidism is present, it should be treated initially or concurrently because calcium potentiates gastrin secretion. Initial treatment of ZES is often pharmacologic, consisting of proton pump inhibitors. Although these may control symptoms and prevent sequelae of ulcer disease, they do not address tumor behavior or the malignant potential. If surgical resection is possible, it is often recommended to prevent tumor spread and to provide a long-term cure. Application of this principle in patients with MEN I is more contentious because localization is more difficult and multifocal disease is common. Preoperative tumor localization often includes somatostatin receptor scintigraphy and CT to evaluate the pancreas, peripancreatic lymph nodes, and potential liver metastases.<sup>2,3</sup> Endoscopic ultrasonography is used in some centers to identify occult primary lesions. Imaging of small duodenal gastrinomas with any of these techniques is exceptionally rare.

Unlike those with sporadic ZES, patients with MEN I frequently have gastrinomas that are small, extrapancreatic (80%), and multifocal (43%).<sup>2</sup> Multiple small duodenal gastrinomas are common, which makes selection of the proper operation in patients with MEN I difficult if cure is defined by durable postoperative eugastrinemia. Compared with sporadic cases, patients with MEN I are less likely to present with metastatic disease. When present, metastatic disease involves nodal spread more commonly than it does hepatic sites.<sup>4</sup> This likely explains the improved survival seen in patients who have MEN I and ZES compared with those who have sporadic disease because liver metastases are a significant predictor of poor long-term outcome.<sup>2</sup>

We present a novel technique that uses a low-density, barium-based oral contrast agent during the arterial phase of intravenous contrast-enhanced spiral CT of the abdomen. The ability of the oral agent to distend the duodenum without obscuring small arterially enhancing tumors allowed preoperative confirmation of multiple duodenal tumors, which influenced surgical planning. Although surgical management of ZES in patients with MEN I remains controversial, improvements in radiology and operative technique may better influence surgical planning for these patients.

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