

Answer

Sporadic Duodenal Macrogastrioma

The diagnosis was confirmed by the measurement of the serum gastrin level, which was more than 10 times greater than the upper limit of normal. An antroduodenectomy associated with a prophylactic regional lymph node dissection was performed subsequently. Histological examination confirmed the diagnosis of malignant gastrinoma with 3 lymphatic metastases. The patient had an uneventful postoperative course and was discharged home on the eighth postoperative day.

Primary duodenal gastrinoma is a common etiology for patients with sporadic Zollinger-Ellison syndrome. Zollinger-Ellison syndrome is characterized by hypersecretion of gastrin resulting in a profound gastric acid hypersecretion and a spectrum of clinical presentations including symptoms of refractory peptic ulcer disease, severe diarrhea, or intestinal perforation. The most common differential diagnoses are idiopathic peptic ulcer disease, chronic idiopathic diarrhea, and gastroesophageal reflux disease. Other differential diagnoses include Crohn disease, irritable bowel syndrome, infectious diarrhea, celiac disease, and lactose intolerance. A gastrin serum level 10 times the upper limit of normal (the upper limit of normal is 100 pg/mL [to convert to picomoles per liter, multiply by 0.481]) accompanied by a gastric pH less than 5 is indicative of gastrinoma.¹ Gastrinomas exist both in hereditary and sporadic forms. Approximately 20% of gastrinomas occur as part of the inherited tumor syndrome multiple endocrine neoplasia type 1, characterized by the combined occurrence of primary hyperparathyroidism, duodenopancreatic endocrine neoplasms, and tumors of the anterior pituitary gland. Nevertheless, multiple endocrine neoplasia type 1 gene mutations are also identified in 33% of sporadic gastrinomas.²

Some 80% to 90% of all gastrinomas are located in the so-called gastrinoma triangle, which includes the duodenum, pancreatic head, and hepatoduodenal ligament.³ At least 50% of sporadic gastrinomas are found in the duodenum and the majority of them are situated in the first and second part of the duodenum.⁴ Duodenal gastrinomas tend to be much smaller than pancreatic gastrinomas and are often occult (<1 cm), making them almost impossible to locate on preoperative conventional images. Somatostatin receptor scintigraphy is the most sensitive method for imaging either primary or metastatic gas-

trinomas. However, the best method of diagnosis remains surgical exploration associated with external palpation, transillumination, or intraoperative ultrasonography.⁵ A duodenotomy, therefore, should be routinely performed on all patients with Zollinger-Ellison syndrome.

Although most gastrinomas grow slowly, 60% to 90% are malignant at the time of initial diagnosis and are associated with lymph node metastases in more than 50% of individuals on operation.⁶ If left untreated, patients with gastrinoma may develop liver metastases, which are associated with a significantly shorter survival rate. Currently, surgical management is directed toward identification and resection of the primary tumor and regional metastases to lymph nodes or liver to prevent malignant progression of disease.

Accepted for Publication: October 1, 2009.

Correspondence: Julien Jarry, MD, Hôpital Haut Leveque, Avenue de Magellan, 33604 Pessac, France (julienjarry@hotmail.com).

Author Contributions: *Study concept and design:* Jarry, Peycru, and Collet. *Acquisition of data:* Sa Cunha. *Analysis and interpretation of data:* Rault. *Drafting of the manuscript:* Rault. *Critical revision of the manuscript for important intellectual content:* Jarry, Peycru, Sa Cunha, and Collet. *Study supervision:* Jarry, Rault, Peycru, Sa Cunha, and Collet.

Financial Disclosure: None reported.

REFERENCES

1. Berna MJ, Hoffmann KM, Serrano J, Gibril F, Jensen RT. Serum gastrin in Zollinger-Ellison syndrome, I: prospective study of fasting serum gastrin in 309 patients from the National Institutes of Health and comparison with 2229 cases from the literature. *Medicine (Baltimore)*. 2006;85(6):295-330.
2. Zhuang Z, Vortmeyer AO, Pack S, et al. Somatic mutations of the MEN1 tumor suppressor gene in sporadic gastrinomas and insulinomas. *Cancer Res*. 1997; 57(21):4682-4686.
3. Stabile BE, Morrow DJ, Passaro E Jr. The gastrinoma triangle: operative implications. *Am J Surg*. 1984;147(1):25-31.
4. Zogakis TG, Gibril F, Libutti SK, et al. Management and outcome of patients with sporadic gastrinoma arising in the duodenum. *Ann Surg*. 2003;238(1):42-48.
5. Sugg SL, Norton JA, Fraker DL, et al. A prospective study of intraoperative methods to diagnose and resect duodenal gastrinomas. *Ann Surg*. 1993;218(2):138-144.
6. Delcore R Jr, Cheung LY, Friesen SR. Characteristics of duodenal wall gastrinomas. *Am J Surg*. 1990;160(6):621-624.