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Surgery for Gastrinoma: Short and Long-Term Results☆

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ABSTRACT

Introduction: Zollinger–Ellison syndrome (Z–E) is characterized by gastrin-secreting tumors, responsible for causing refractory and recurrent peptic ulcers in the gastrointestinal tract. The optimal approach and the extension of tumor resection remains the subject of debate.
Methods: During the period February 2005 and February 2014, 6 patients with Z–E underwent surgery, 4 men and 2 women with a median age 46.8 years (22–61). Two patients were affected with multiple endocrine neoplasia type-1 (MEN-1). Fasting gastrin levels greater than 200 pg/ml (NV: <100) was diagnostic. Radiologic imaging to localize the lesion included octreoscan 6/6, computer tomography (CT) 6/6, and endoscopic ultrasonography (EUS) 1/6.
Results: The octreoscan was positive in 5 patients. The CT localized the tumor in the pancreas in 2 patients, in the duodenum in 3 patients (1 confirmed by EUS) and between the common bile duct and vena cava in one patient. The laparoscopic approach was used in 4 patients, 2 patients converted to open surgery. The following surgical techniques were performed: 2 pylorus-preserving pancreatico-duodenectomy (PPPD), one spleen-preserving distal pancreatectomy, one duodenal nodular resection, 1 segmental duodenectomy and one extrapancreatic nodular resection. Pathological studies showed lymph nodes metástasis in 2 patients with pancreatic gastrinomas, and in one patient with duodenal gastrinoma. The median follow-up was 76.83 months (5–108) and all patients presented normal fasting gastrin levels.
Conclusions: Surgery may offer a cure in patients with Z–E. The laparoscopic approach remains limited to selected cases.

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Cirugía del gastrinoma: Resultados inmediatos y a largo plazo

Palabras clave:
Gastrinoma esporádico
Gastrinoma familiar
Laparoscopia
Gastrinoma linfático primario

RESUMEN

Introducción: El síndrome de Zollinger–Ellison (Z–E) está caracterizado por tumores productores de gastrina responsables de la aparición de úlceras recurrentes en el tracto gastrointestinal. El abordaje quirúrgico y la extensión de la resección tumoral son todavía controvertidos.

Métodos: De febrero de 2005 a febrero de 2014 se intervino a 6 pacientes con Z–E, 4 hombres y 2 mujeres, con una mediana de edad 46,8 años (22–61). Dos pacientes presentaban una neoplasia endocrina múltiple-1 (NEM-1). El diagnóstico se estableció por la determinación de gastrina basal en ayunas >200 pg/ml (VN <100). Para el diagnóstico de localización se utilizó el octreoscan (6/6), la tomografía axial computarizada (TAC) (6/6) y la ultrasonografía endoscópica (USE) (1/6).

Resultados: El octreoscan fue positivo en 5 pacientes. La TAC localizó el tumor en todos los pacientes: páncreas (2), duodeno (3, uno confirmado por USE), entre el conducto biliar y la vena cava (uno). El abordaje laparoscópico se utilizó en 4 pacientes, 2 pacientes fueron convertidos a cirugía abierta. Entre las técnicas quirúrgicas se realizaron: 2 duodenopancreatectomías cefálicas con preservación pilórica (DPCPP), una pancreatectomía distal con preservación esplénica, una resección nodular duodenal, una resección duodenal segmentaria y una resección nodular extrapancreática. La anatomía patológica demostró metástasis linfáticas en 2 pacientes con gastrinomas pancreáticos y en un paciente con gastrinoma duodenal. La estancia hospitalaria mediana fue 11,3 días (10–14). Durante el período de seguimiento clínico, con una mediana de 76,83 meses (5–108), todos los pacientes presentaron una gastrina en ayunas normal.

Conclusiones: La cirugía puede ofrecer la curación en pacientes con Z–E. El abordaje laparoscópico permanece limitado a casos seleccionados.

Introduction

Gastrinomas are uncommon endocrine tumors, with an incidence of 0.5–1/1 000 000 inhabitants/year, and the second in frequency, preceded by insulinomas. Their incidence is greater in men than in women, and mean age at presentation is between 45 and 50.[1] The clinical manifestations of these tumors are associated with gastrin hypersecretion, causing an elevation of gastric acid that leads to the appearance of Zollinger–Ellison syndrome. In turn, this produces gastroduodenal and jejunal ulcers and altered gastrointestinal motility, causing diarrhea in up to 70% of cases.[2] Gastrinomas can appear sporadically (70%) or as part of multiple endocrine neoplasia, type 1 (MEN-1) syndrome.[3] Although these tumors grow slowly, they are malignant in 60%–70% of cases, and 25% of cases progress quickly.[4]

The diagnosis and localization of gastrinomas has changed favorably in recent years with the use of computed tomography (CT), endoscopic ultrasound (EUS) and especially scintigraphy through the injection of octreotide, a somatostatin analog that binds with tumor somatostatin receptors.[5] This latter test has high diagnostic sensitivity when tumors are larger than 2 cm (96%), but it drops to 30% when tumors are smaller than 1 cm.[5] The diagnosis of gastrinomas requires demonstrating elevated fasting gastrin levels and, when in doubt, this elevation becomes more evident with the injection of secretin.[6,7]

The control of the disease involves the administration of proton pump inhibitors. However, it is surgery that provides a cure in up to 40% of patients with sporadic gastrinomas and, in cases of gastrinomas associated with MEN-1, the prevention of malignant transformation.[8] The choice of surgical technique is controversial with regards to either open or laparoscopic surgery and the extension of the surgery, which is either conservative or radical.

The aim of this study is to analyze the immediate and long-term results of surgery in patients with sporadic gastrinomas and in patients with gastrinomas associated with MEN-1.

Patients and Methods

From February 2005 to February 2014, 6 patients (4 men and 2 women) were diagnosed with Zollinger–Ellison syndrome and treated surgically. Mean age was 46.8 (22–61). Patients had experienced gastrointestinal discomfort, and multiple gastric or duodenal ulcers were detected by gastroscopy. Demographic data of the diagnosed patients are presented in Table 1. One patient (case 5) was treated surgically due to perforation of a duodenal ulcer, and another patient (case 6) had upper gastrointestinal bleeding due to erosive esophagitis. Two patients had a family history of MEN-1: the father of one patient (case 2) had died due to a malignant gastrinoma with hepatic metastasis; the other patient (case 3) had undergone subtotal parathyroidectomy due to primary