The Zollinger-Ellison syndrome and ectopic gastric mucosa

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Summary

Ectopic gastric mucosa in an intramesenteric ileal diverticulum caused fatal bleeding after total gastrectomy in a patient with the Zollinger-Ellison syndrome (ZES).

Introduction

The Zollinger-Ellison syndrome may be treated by removal of the tumour(s), total gastrectomy (Zollinger and Ellison, 1955), an H₂ inhibitor, such as cimetidine (McCarthy, 1976) or a combination of these three methods. Our approach (Stadil and Stage, 1978; Welbourn and Galland, 1982) is to control the ulceration with cimetidine and then, if possible, to localize and remove the tumour. If this is not possible, we either continue with cimetidine or undertake a total gastrectomy, the choice depending upon factors peculiar to each patient.

The aim of total gastrectomy is to remove the 'target organ' for the action of gastrin. The presence of a second, undetected, target organ may lead to unexpected postoperative problems.

Case report

An Indian man aged 29 presented at Hammersmith Hospital in December 1979 with a 2 year history of intermittent epigastric pain and vomiting. Four previous barium meals and 2 endoscopies had shown duodenal ulceration. Two courses of cimetidine (1 g/day), each lasting for 6 weeks, had caused remissions. On each occasion the ulceration recurred after about 6 months. He had been hypertensive since 1975. His father had had diabetes, hypertension, hyperparathyroidism and a neuroblastoma, and his mother had suffered from a Wilms tumour. There was no family history of ulcer disease. Previous studies had shown basal acid outputs of 21 mmol/hr in 1977 and 41 mmol in 1979 (normal 0-5), with maximum acid outputs of 52 mmol/hr (normal 5-40) on each occasion.

On admission there were no abnormal signs and his basal acid output was 63.4 mmol/h. The fasting plasma gastrin and pancreatic polypeptide were raised, being 400 and 660 pmol/l respectively (upper limits of normal being 30 and 200). By this time 2 g per day of cimetidine were required to control the ulceration. These findings suggested a diagnosis of Zollinger-Ellison syndrome, but selective pancreatic angiography failed to localize a tumour.

The patient was not able to travel from India for regular supervision of his cimetidine therapy, and after discussion it was agreed with him that, unless a resectable gastrinoma was found at operation, total gastrectomy would be undertaken. At laparotomy in February 1980, multiple small (1 cm) tumours were found throughout the pancreas and two superficial ones were removed. Examination of frozen sections showed them to be endocrine tumours with cystic degeneration. There was no sign of spread of tumour or of other disease within the abdomen. However, obesity obscured the ileal mesentery. Since no single resectable gastrinoma was found, total gastrectomy was performed and thoracotomy was required for completion of a Roux-en-Y oesophago-jejunal anastomosis.

Postoperatively he developed tachycardia and pyrexia, thought to be due to right lower lobe
atelectasis, and he was treated with physiotherapy and antibiotics. On the second postoperative day he became hypotensive following severe loss of dark red blood from the rectum. After resuscitation immediate laparotomy and simultaneous endoscopy were undertaken. The bleeding had apparently stopped and no cause was found for it.

After this operation he required ventilation and cardiac support for one day. However, soon after extubation his breathing became laboured again, his blood gases deteriorated and it was decided to reintubate and ventilate him. During this process he suffered sudden deterioration and ventilate him. A small amount of altered blood was passed rectally at this time.

A coroner's post-mortem showed that the anastomoses were intact but that there was a diverticulum, 2.5 cm deep and 1.5 cm in diameter, on the mesenteric border of the distal ileum, invisible until the bowel had been opened. There was an eroded artery near its mouth (Fig. 1).

Fig. 1. Drawing of the intramesenteric diverticulum containing the eroded artery

Histology of the pancreatic lesions confirmed that they were endocrine cell tumours. Specific immunocytochemistry revealed pancreatic polypeptide in many cells and glucagon in some. No reactivity to gastrin was found. The gastric antrum contained normal numbers and distribution of cells containing gastrin and somatostatin. A small endocrine type tumour was present in the fundus of the stomach, but no specific immunoreactivity was found in its cells. The ileal diverticulum was lined mainly by gastric type mucosa and contained many parietal cells. It had undergone some autolysis, particularly in an area at the mouth, where the mucosa was absent. It contained an eroded artery 2 mm in diameter.

Discussion

A solitary ileal diverticulum lined by gastric mucosa situated between the layers of the mesentry is rare. It probably represents one of the 'duplication cysts' described by Bremer (1944), which occasionally contain gastric mucosa (Webster, 1955).

Many diverticula of the small intestine are readily seen at operation. In this case it was obscured by fat and was not visible until the bowel was opened at post-mortem. Such a lesion in a patient with the Zollinger-Ellison syndrome does not appear to have been described previously.

The source of gastrin production in this patient was not found. The material obtained fresh at operation (two small pancreatic tumours and the whole stomach) did not contain any recognisable gastrin-producing tissue and the remainder of the pancreas, which was obtained at autopsy, was unsuitable for study. However, it seems likely that one or more of the remaining pancreatic tumours was a gastrinoma.

Despite the rarity of this association, a diverticulum of the bowel, or other source of ectopic gastric mucosa, should be borne in mind as a possible cause of bleeding after total gastrectomy for the Zollinger-Ellison syndrome.

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