Pancreatic neuroendocrine tumour presented as isolated gastric varices

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Isolated gastric varix (IGV) is an uncommon condition. It is frequently seen in patients with liver cirrhosis. However, in patients without cirrhosis, IGV may signify the presence of segmental portal hypertension (PH). In contrast to generalized PH, segmental PH is usually caused by splenic vein obstruction. The raised left side has portal venous pressure and thus gives rise to gastric varices. The following is a report of a patient with a pancreatic neuroendocrine tumour that presented as IGV.

Case report

A 54-year-old man presented with epigastric discomfort for 6 months. Physical examination was unremarkable. Blood tests were unremarkable except for a low platelet count (78×10^9/L). The liver function test was normal, and the hepatitis B and C serology tests were negative. Abdominal ultrasound showed gallstones and splenomegaly. The liver was normal in size, contour and echogenicity. Oesophagogastroduodenoscopy was performed, and IGVs were found at fundus. Subsequent CT scan revealed a 3-cm hypodense lesion at the pancreatic tail near the splenic hilum. The splenic vein was compressed by the lesion and terminated abruptly just distal to it (Fig. 1). Also, there were multiple varices in the perigastric region, as shown on the CT scan (Fig. 2).

The patient underwent surgical exploration for the pancreatic tail lesion. A 3-cm indurated mass at the tail of the pancreas was found. It was located at the splenic hilum and compressed onto the splenic vein, causing splenomegaly and engorgement of short gastric veins. Distal pancreatectomy with splenectomy was performed (Fig. 3). Microscopic examination of the specimen revealed an infiltrative pancreatic tumour, which invaded the spleen and encased the splenic vein at the splenic hilum. Metastatic tumours were also noted in 2 of the 3 peripancreatic lymph nodes. All the resection margins were clear. Immunohistochemical studies showed that the tumour cells were positive for chromogranin and synaptophysin and were negative for insulin, gastrin, somatostatin and pancreatic polypeptide. The diagnosis was nonfunctioning malig-

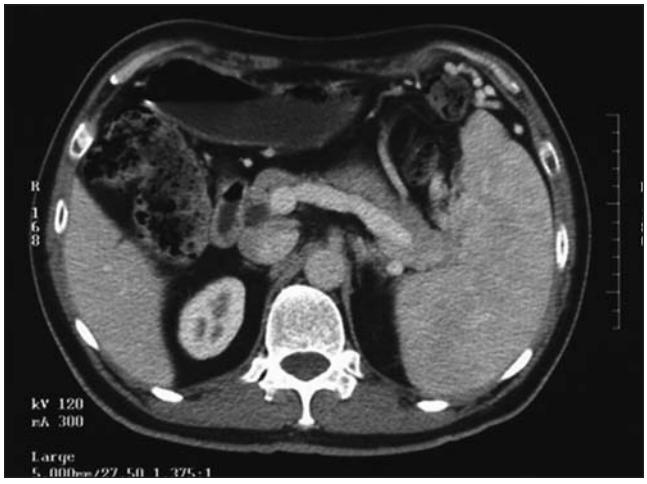

FIG. 1. Axial CT scan showing a 3-cm pancreatic tail lesion compressing the splenic vein.

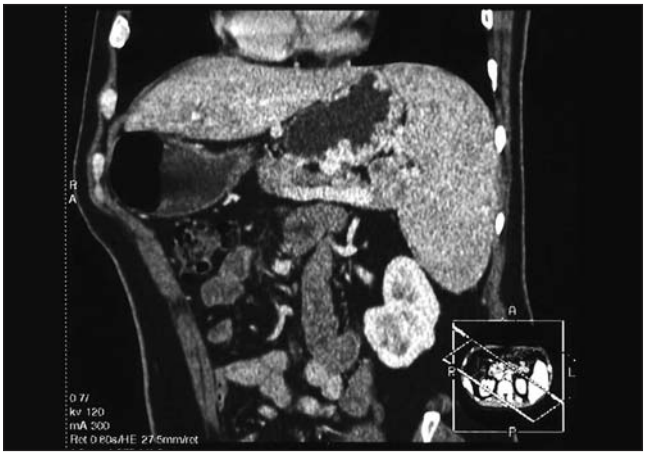

FIG. 2. Coronal reconstruction of the CT scan, showing multiple perigastric varices with abrupt termination of the splenic vein distal to the pancreatic tumour.

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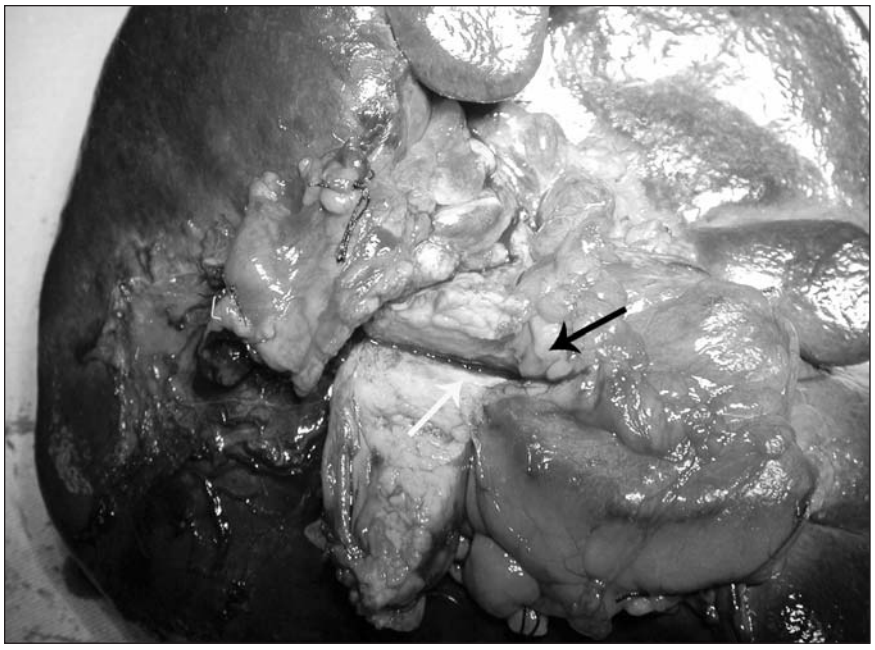

FIG. 3. The cut surface of the surgical specimen (distal pancreas with spleen) showing the distal pancreatic tumour (black arrow) compressing the splenic vein (white arrow).

nant pancreatic neuro-endocrine tumour.

The patient recovered from the operation uneventfully and was discharged 9 days after the operation. The platelet count returned to normal (173×10^9/L) and the patient remained asymptomatic, with no evidence of recurrence 18 months after surgery.

Discussion

Gastric varices in the absence of esophageal varices are termed isolated IGVs. Type 1 IGV (fundal IGV) is located at the gastric fundus, whereas type 2 IGV (ectopic IGV) may present anywhere in the stomach, such as in the antrum or pylorus.[1] IGV is predominantly secondary in origin, developing after esophageal variceal obliteration in patients with cihrrhosis.[2] For primary IGV, the most common cause is still liver cirrhosis, with resulting portal hypertension.[3,4] In patients without liver disease, segmental PH is an important cause of IGV and is usually secondary to splenic vein obstruction. Madsen and colleagues reviewed 209 patients with isolated splenic vein obstruction.[5] Sixty-three percent of the cases were caused by pancreatitis, whereas pancreatic neoplasms contributed to 18% of the cases. However, extrinsic compression of the splenic vein by a pancreatic neuroendocrine tumour is extremely rare.

Segmental PH commonly presents as bleeding gastric varices and abdominal pain.[3] The characteristic physical finding in segmental PH is IGV and splenomegaly.[3] In these cases, gastric varices usually originate from the short gastric and gastroepiploic veins.[1] It appears as fundal varices (type 1 IGV) on endoscopy.[1] Endoscopy and splenoportovenography are useful in diagnosing IGV, whereas CT scan is helpful in detecting the underlying pancreatic pathologies.[1,4] The definitive treatment of segmental PH depends on the underlying causes. Resectable pancreatic tumour should be resected. Other causes of segmental PH can be treated by splenectomy to minimize the pressure and flow in the varices and other collaterals.[5]

In summary, segmental PH should be suspected in patients with IGV without signs of liver disease. Underlying pancreatic tumours must be looked for, because they may be the only presenting feature of a malignant tumour, as illustrated in this case.

Competing interests: None declared.

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