Pancreatic carcinoma presenting as bleeding from segmental gastric varices: pitfalls in diagnosis

F.J. Mullan and S.T.D. McKelvey

Surgical Department, The Ulster Hospital, Dundonald, Belfast BT16 0RH, Northern Ireland.

Summary: Splenic vein occlusion leading to gastric variceal haemorrhage should be considered in cases of obscure upper gastrointestinal bleeding. We report an unusual case in which the underlying pathology was a resectable carcinoma of the pancreatic tail.

Introduction

Segmental gastric varices due to isolated splenic vein thrombosis is an unusual complication of pancreatic pathology. Early diagnosis is rarely made but may be vital where the underlying pathology is malignant. We describe a case of operable pancreatic carcinoma presenting with bleeding gastric varices which illustrates the diagnostic pitfalls and the need for a better awareness of the syndrome.

Case report

A 63 year old woman with maturity onset diabetes was admitted following a haematemesis and melena. Urgent barium meal examination suggested carcinoma of the cardia of the stomach. A further massive haematemesis prompted emergency surgery. At laparotomy there was no evidence of tumour. The gastric mucosa was friable and bled easily on contact. Several areas were undersewn. Truncal vagotomy and pyloroplasty were performed. The spleen was noted to be slightly enlarged. Six months after initial presentation the patient was readmitted with recurrent anaemia. Repeat barium meal again showed fundal mucosal distortion (Figure 1a) indistinguishable from malignant infiltration. Oesophagogastro-duodenoscopy failed to establish any site of mucosal destruction and the fundus appeared normally distensible.

At laparotomy the spleen was twice normal size. There was no infiltration or ulceration of the gastric fundus but the gastroepiploic and short gastric veins were markedly distented. The lesser sac was opened and after thorough exploration a small firm tumour was found in the pancreatic tail encroaching upon and occluding the splenic vein.

Splenectomy and distal pancreatectomy were performed. Histology confirmed a 3.5 x 2 cm adenocarcinoma of the pancreatic tail infiltrating around and compressing large blood vessels. Barium meal performed five months postoperatively showed complete resolution of the fundal varices (Figure 1b) and ultrasonic scanning confirmed patency of the portal vein.

Discussion

The splenic vein running along the postero-superior aspect of the pancreas is vulnerable to compression or infiltration by adjacent pancreatic lesions. This results in isolated splenic vein thrombosis (ISVT) and an unusual form of extrahepatic portal hypertension. An extensive collateral circulation develops involving the segment of stomach drained by the short gastric veins (Figure 2). Bleeding may occur from the resultant gastric varices.

Sutton¹ and Madsen² in comprehensive reviews of English language publications from 1900–69 and 1969–84, respectively, identified only 263 cases. Almost half the cases were secondary to pancreatitis or its sequelae, notably pseudocyst formation. Only 36 cases were attributable to adenocarcinoma of the pancreas, the vast majority of which were irresectable.

Diagnostic delay is common. In Madsen’s series only 49% of patients were correctly diagnosed at the time of first bleed, the remainder having a median diagnostic delay of 11 months. The correct diagnosis was missed at first operation in 22% of patients undergoing laparotomy for gastrointestinal bleeding. Only 45% of barium studies correctly diagnosed gastric varices³ which were completely missed in 29% of cases. Endoscopy was no more sensitive with localized gastric varices correctly diagnosed in less than 40% of cases.⁴ Experience of ISVT outside major referral centres is limited.

Correspondence: F.J. Mullan, F.R.C.S.
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What then gastroenterologist, radiologist, and surgeon to suspicion. Symptoms are prominent irregular as the gastric fundus.

Figure 1 (a) Barium meal demonstrating gastric varices as prominent irregular mucosal folds (arrows) limited to the gastric fundus. (b) Resolution of varices after splenectomy and resection of pancreatic tumour.

What then are the features which might alert the gastroenterologist, radiologist, endoscopist or surgeon to the possibility of this diagnosis? The key to early diagnosis is a high index of suspicion. Symptoms may be limited to those of the underlying disease. The two commonest clinical features, gastrointestinal blood loss and splenomegaly, are particularly suspicious when associated with a history of pancreatitis or recurrent upper abdominal pain. Bleeding is frequently severe enough to cause haematemesis but the source may have remained obscure following previous investigations. Splenomegaly is common but not invariable and may only be apparent at laparotomy. Clinical splenomegaly is apparent in about half of reported cases. Evidence of hypersplenism is diagnostically helpful but infrequently found. Liver function tests are invariably normal unless there is metastatic liver disease or alcohol abuse.

Gastric varices appear on barium studies as thick distorted, tortuous mucosal folds or filling defects over the greater curvature of the stomach extending towards the cardia. Confusion with gastric neoplasm is commonplace. Co-existing splenomegaly is more suggestive of varices. Isolated gastric varices must be included in the differential diagnosis of any polypoid lesion or hypertrophic rugae localized to the gastric fundus.

Ultrasoundography is a useful non-invasive method of confirming patency of the portal vein and with computed tomographic scanning may help to localize an underlying pancreatic mass. The position and patency of the splenic vein itself are more difficult to identify by ultrasonography.

ISVT may be confirmed pre-operatively by splenoportography or the venous phase of selective angiography. The latter procedure is now considered the method of choice being safer and more easily controlled than percutaneous splenoportography.

Endoscopic misinterpretation or hazardous, even lethal, biopsy may result unless ISVT is considered when a nodular submucosal mass or prominent fundal folds are found in the stomach. Suspicion of ISVT should be high when a local fundal abnormality identified radiologically is not confirmed endoscopically. Oesophageal varices are normally absent unless the short gastric collateral system is inadequate to decompress the splenic axis or the coronary vein inserts into the splenic vein rather than the portal vein.

Dilated gastroepiploic veins, particularly in the presence of a large spleen and a normal liver, are very suggestive of ISVT. These may be less obvious in a hypotensive patient when emergency laparotomy is undertaken for profound haematemesis. Gastric varices should be considered when laparotomy fails to reveal a convincing site of haemorrhage in a blood filled stomach.

Bleeding from gastric varices due to benign ISVT is almost always cured by splenectomy. Malignant ISVT usually implies extrapancreatic extension and a very poor prognosis.
creatic carcinomas causing ISVT have been re-
sected\textsuperscript{10,16} and Hurwitt\textsuperscript{16} reported a case surviving
19 months postoperatively. In view of the patho-
logic findings we could not claim a curative
operation in this case but excellent palliation has
been achieved in our patient, alive 20 months after
pancreatic resection.

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