Left-sided portal hypertension due to retroperitoneal fibrosis treated with an oesophagus preserving, modified Sugiura procedure

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ABSTRACT

Left-sided portal hypertension is a very uncommon condition and retroperitoneal fibrosis has rarely been reported as a cause. We present the case of a 77-year-old man with retroperitoneal fibrosis obstructing the splenic vein and causing recurrent episodes of upper gastrointestinal bleeding. Computed tomography showed a retroperitoneal mass as being responsible for the obstruction of the splenic vein, splenomegaly, and diffuse varices around the gastrosplenic and gastrohepatic ligaments. An oesophagus preserving, modified Sugiura procedure was performed with disconnection of the gastric vessels on the lesser curve of the stomach, preserving the pylorus branches of the nerves of Latarjet.

KEYWORDS

Portal hypertension – Oesophageal and gastric varices – Retroperitoneal fibrosis – Modified Sugiura procedure

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Introduction

Left-sided portal hypertension (LSPH) is a very uncommon condition that occurs mainly as a result of splenic vein obstruction, causing gastrointestinal (GI) bleeding. LSPH has been associated with pancreatic disorders such as pancreatitis, pancreatic pseudocysts and pancreatic carcinomas. However, retroperitoneal fibrosis (RF) has rarely been reported as a cause.¹ We present the case of a patient with LSPH due to RF treated with an oesophagus preserving, modified Sugiura procedure, followed by a review of the literature highlighting the clinical management of this condition.

Case history

Our patient was a 77-year-old man with a background of hypertension, myelodysplastic syndromes, a cerebrovascular accident in 2012 and RF diagnosed in 2008. The RF was diagnosed in an external centre following severe back pain, with computed tomography (CT) showing a retroperitoneal mass encasing the left ureter and hydronephrosis. He started oral steroids and was planned for surgical debridement of the left ureter. Careful lysis was performed of adhesions between the ureter and the retroperitoneal mass. A biopsy of the retroperitoneal mass was taken, confirming the diagnosis of RF, with no

evidence of any tumour. Steroids were gradually suspended and the patient started treatment with mycophenolate.

In 2017 the patient presented to the emergency department because of upper gastrointestinal bleeding with a haemoglobin level of 6.8g/dl, requiring repeated transfusions of red cell units. A gastroscopy showed diffuse fundal gastric varices but no oesophageal varices (Fig 1). CT demonstrated progression of the retroperitoneal mass (which was obstructing the splenic vein), splenomegaly, and diffuse varices around the gastrosplenic and gastrohepatic ligament (Fig 2).

With the diagnosis of LSPH due to RF, the patient was referred to our clinic, giving consent for an oesophagus preserving, modified Sugiura procedure. Intraoperative examination showed diffuse varices around the splenic hilum, the gastrosplenic ligament and the gastrohepatic ligament. A biopsy was taken from the retroperitoneal tissue, confirming the diagnosis of RF, without any evidence of malignancy. A splenectomy was performed with careful disconnection of varices from the greater curve of the stomach and retroperitoneal tissue (Fig 3A). Attention was then given to the lesser curve and the varices arising from the gastrohepatic ligament (Fig 3B). Ligation of these varices was carried out, preserving the pylorus branches of the nerves of Latarjet (Fig 3C). Postoperative recovery was uneventful and the patient was discharged on day 4. After

18 months of follow-up review, he has not suffered any further episodes of bleeding and his haemoglobin levels have remained stable.

Discussion

LSPH is a very uncommon condition. Its reported incidence has increased over the past few decades owing to advances in diagnostic approaches and the wide diffusion of abdominal imaging tests. However, owing to its asymptomatic nature, the exact incidence is still unknown, probably accounting for less than 5% of all patients with portal hypertension.¹

The vast majority of published series includes a limited number of patients and they are usually retrospective. The main cause of LSPH is splenic vein thrombosis, followed by compression of the splenic vein mainly related to pancreatic disorders such as chronic pancreatitis, pancreatic pseudocysts, and pancreatic carcinomas. RF has rarely been noted as a cause, with its first report in 1962 by Eiseman and Yeoh.²

Splenic vein occlusion or thrombosis can result in LSPH, with the venous drainage of the spleen redirected through the short gastric vessels to the gastric and oesophageal vessels. Eventual decompression into the portal system occurs through the coronary and epiploic veins but this situation can also cause a dilation of the gastro-oesophageal submucosal venous reticulum into varices, which can lead to gastric or oesophageal bleeding. The presence of a coronary vein draining distally to the obstruction in the splenic vein can exacerbate this phenomenon, resulting in chronic dilation of intraperitoneal gastro-oesophageal veins.

LSPH is generally asymptomatic and its first clinical manifestation is usually acute upper GI bleeding from ruptured gastric or oesophageal varices. Patients may also present with chronic anaemia due to portal hypertensive gastropathy. Splenomegaly is a hallmark of chronic LSPH. Gastro-oesophageal and upper GI varices can be seen both radiologically and endoscopically. In addition, contrast enhanced CT, magnetic resonance imaging and endoscopic ultrasonography can be used to assess portal vasculature and clarify the diagnosis, identifying the primary cause of LSPH.

Conservative management can be indicated in patients with uncomplicated LSPH whereas episodes of upper GI bleeding are an indication for splenectomy. Management of LSPH involves surgical correction of the underlying causes, combined with splenectomy. The rationale for splenectomy is to interrupt the arterial supply feeding the gastro-oesophageal varices. A laparoscopic approach has traditionally been considered contraindicated because of the increased risk of intraoperative haemorrhage related to portal hypertension. However, recent series have shown favourable outcomes with laparoscopic splenectomy in patients suffering from cirrhosis related portal hypertension, and in 2014 Patrono *et al* reported successful management of a case of LSPH with preoperative splenic artery embolisation followed by laparoscopic splenectomy.³

There is controversy regarding the ideal management of gastric or oesophageal varices so as to reduce the risk of further episodes of upper GI bleeding. The role of surgery in portal hypertension has decreased since the advent of alternative endoscopic and percutaneous therapies. The Sugiura procedure used for the treatment of portosystemic shunt secondary to portal hypertension included extensive para-oesophagogastric devascularisation, oesophageal transection, splenectomy, vagotomy and pyloroplasty. Several modifications of the original procedure with gastro-oesophageal devascularisation without oesophageal

Figure 1 Gastroscopy showing diffuse fundal gastric varices

Figure 2 Computed tomography showing a retroperitoneal mass (asterisk) obstructing the splenic vein with diffuse varices in the fundus of the stomach (white arrow), and around the gastrohepatic and gastrosplenic ligament (black arrows)

Figure 3 Oesophagus preserving, modified Sugiura procedure: splenectomy performed with disconnection and ligation of varices from the gastrosplenic ligament and retroperitoneum (white asterisks) (A); varices on the lesser curve (black asterisk) before their ligation (B); and ligation of varices from the gastrohepatic ligament, preserving the pylorus branches of the nerves of Latarjet (white arrow) (C)

transection have shown similar results and less morbidity compared with devascularisation with oesophageal transection.⁴ Nowadays, these surgical procedures represent an effective rescue therapy for patients with bleeding oesophageal varices when other, more conservative treatments fail or are not indicated.

For LSPH caused by benign pancreatic disease such as RF, there is a lack of evidence on the optimal management of gastric and oesophageal varices. Gastric disconnection in addition to splenectomy can represent an excellent alternative to reduce the risk of further bleeding, adding only a very low risk of comorbidity to the procedure, especially in patients with benign disease and a coronary vein draining distally to the obstruction in the splenic vein. Our technique included isolated disconnection of gastric vessels on the lesser curve, preserving the pylorus branches of the nerves of Latarjet, as described by Ginsberg *et al* in their modified Sugiura procedure.⁵

Our patient had an uneventful recovery and after 18 months of follow-up review, he has suffered no further episodes of upper GI bleeding or anaemia. The modified Sugiura procedure can therefore represent a reproducible, effective and safe option in treating LSPH due to RF or other benign pancreatic diseases, preventing recurrent episodes of bleeding. Nevertheless, a prospective trial comparing the modified Sugiura procedure with isolated splenectomy will determine its eventual role in the management of bleeding from LSPH.

Conclusions

RF is an extremely rare cause of LSPH. An oesophagus preserving, modified Sugiura procedure is a reproducible, effective and safe alternative to treat LSPH, and prevent further episodes of bleeding.

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