Left-sided portal hypertension (LSPH), also known as segmental, regional, localized, compartmental, lineal, splenoportal, or sinistral hypertension is a rare, but life threatening cause of upper gastrointestinal bleeding. It usually occurs as a result of isolated obstruction of the splenic vein. The incidence of LSPH has increased over the past three decades due to increased awareness of the entity and advances in diagnostic approaches. Since most patients are asymptomatic and experience no complications, its exact incidence is unknown. However, it accounts for less than 5% of all patients with portal hypertension. To date, less than 500 cases of LSPH have been reported in all. Most of the studies in the literature comprise a limited number of patients and are usually retrospective. Due to its low incidence, it is likely that most cases of sinistral hypertension are initially misdiagnosed as a generalized portal hypertension.

Blood flow through the splenic vein may be blocked secondary to either thrombosis formation or neighboring mass effect. Following obstruction, splenic blood typically drains through the short gastric veins to the stomach. In the gastric wall veins of the fundus, blood flow and pressure increase and submucosal structures consequently dilate, producing gastric varices. Fortunately, due to several anatomic variations, obstruction of the splenic vein may not always result in portal hypertension or formation of varices. It is believed that similar pathophysiological mechanisms may occur in cases of superior mesenteric vein obstruction, although in this case the risk of ectopic varices involving the proximal small bowel might be more common.

A recent study confirmed the past presumptive knowledge that most common pathologies resulting in splenic vein thrombosis or obstruction and leading to LSPH are chronic pancreatitis, pancreatic pseudocysts and pancreatic neoplasms. In particular, the presence of pseudocyst in patients with chronic pancreatitis might be associated with significantly higher incidence of splenic vein thrombosis. These account for about 18% of the LSPH cases include benign neoplasms, adenocarcinoma and functioning and non-functioning neuroendocrine tumors. Iatrogenic splenic vein injury after liver transplantation, infiltration by colonic tumor, spontaneous splenic vein thrombosis and perirenal abscess have also been linked with LSPH, but these etiological factors are rare.

The diagnosis of sinistral portal hypertension should be considered in all those with upper gastrointestinal bleeding associated with splenomegaly and normal liver function tests. Most commonly, however, LSPH is asymptomatic and is found incidentally on investigation. In up to 73% of the symptomatic cases, the first clinical manifestation of LSPH, is generally acute (often massive) or chronic upper gastrointestinal bleeding from ruptured esophageal or gastric varices, and rarely from colonic varices. Up to 71% of patients have splenomegaly, few patients suffer from splenic pain and develop leukopenia or thrombocytopenia. As the
Management of asymptomatic patients is more controversial than the symptomatic ones: splenectomy has been suggested as a prophylactic measure by some while others have not shown any significant benefit of this procedure in survival. However, more evidence suggests that watchful waiting as an acceptable course of management in asymptomatic individuals.

Excluding deaths from gastrointestinal bleeding, the prognosis of LSPH mainly depends on the underlying disease, with those with pancreatic malignancies having the worst outcomes. As such, the evidence LSPH, although benign in most cases, can sometimes be the first signal of a sinister entity, like the pancreatic cancer.

In this issue of GE Portuguese Journal of Gastroenterology, Fernandes et al. report the clinical presentation, etiologies and outcome in a cohort of 22 patients with LSPH. This retrospective analysis highlights some particular issues regarding this difficult to diagnose and manage entity. Similarly to reported series, most patients (n = 14) experience no symptoms or presented non-specific abdominal pain, and LSPH was found incidentally on investigation. This raises the question about the value of screening asymptomatic patients for the presence of LSPH. Bernardes et al. search for splenorenal venous obstruction in a medical-surgical prospective series of 266 patients with chronic pancreatitis who were followed up a mean time of 8.2 years but was symptomatic in only two. Development of varices in LSPH occurs only when formation of adequate low-pressure collateral do not develop, so diagnostic test should evaluate not only splenic vein obstruction but also the presence of varices. In this context, EUS may be the ideal diagnostic modality since it can be used to assess portal vasculature, patency of splenic vein and to diagnose gastric varices. In this cohort of patients EUS was used in nine patients with LSPH. The authors find that the principal etiology of LSPH was pancreatic pathology (chronic pancreatitis, acute pancreatitis and pancreatic tumours), situations in which EUS has demonstrated superior accuracy to transabdominal US or abdominal CT. Another controversial issue raised by this study is the management of LSPH, namely the treatment of varices and the need to initiate anticoagulation therapy. In this regard, we agree with the authors that anticoagulation therapy should be instituted after treatment of varices, since it’s difficult to determine the potential risk of bleeding from gastric varices. CYA obliteration is the recommend approach to control upper gastrointestinal bleeding, however treatment of asymptomatic patients with gastric varices is not consensual since this treatment is associated with risk of embolic events.

This pertinent study brings to the forefront a condition not as uncommon as previously thought, but whose approach remains based on small cases series and clinical cases and whose consequences can be catastrophic.

References


