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EDITORIAL

Left-Sided Portal Hypertension: A Clinical Challenge

Hipertensão Portal Esquerda: Um Desafio Clínico

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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.[1] 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.[2,3] 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.[1] 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.[4] In particular, the presence of pseudocyst in patients with chronic pancreatitis might be associated with significantly higher incidence of splenic vein thrombosis.[5] These account for about 18% of the LSPH cases include benign neoplasms, adenocarcinoma and functioning and non-functioning neuroendocrine tumors.[6,7] 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.[1]

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,[2] 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.[8] Up to 71% of patients have splenomegaly, few patients suffer from splenic pain and develop leukopenia or thrombocytopenia.[2] As the

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majority of patients does not present with cirrhosis, development of ascites is rare unless they develop acute dilutional hypoalbuminemia for any reason.[1]

While diagnosis is mainly clinical and often made by exclusion of systemic portal hypertension, diagnostic imaging plays an important role in confirming the diagnosis in the majority of cases.[9] Angiography of splenic vein remains the gold standard in diagnosing sinistral portal hypertension, although it is less used today by the availability of less invasive methods. Transabdominal ultrasonography (US) with Doppler is often the initial imaging modality utilized, but it is more helpful in excluding presence of systemic portal hypertension and its primary etiologies such as liver cirrhosis. The accuracy of trans-abdominal US is limited in detecting splenic or superior mesenteric veins thrombosis, which are smaller and more subtle than those of portal veins.[10] Recently, endoscopic ultrasound (EUS) has been used to assess the portal vasculature. This method appears to be a more accurate test than transabdominal US for evaluating patency of the splenic vein.[11] EUS should be considered when other diagnostic methods have failed to confirm splenic vein thrombosis as a cause of bleeding gastric or gastroesophageal varices, especially in patients with previous history of pancreatitis.[1] It should also be considered in cases occurring without previous history of chronic pancreatitis, so that pancreatic carcinoma can be investigated as a potential cause. In recent years contrast enhanced CT scan and magnetic resonance angiography are gaining popularity due to faster acquisition of images with low invasiveness.[9] However, the sensitivity and specificity for detection of minor alterations of splenic vasculature are still modest.

Management of this condition traditionally involves removal of the primary cause if possible, which can be combined with splenectomy. Symptomatic patients with variceal bleeding may be severe and life threatening. Current guidelines recommend endoscopic therapy as first-line treatment for bleeding gastric varices. Endoscopic therapeutic options for gastric variceal bleeding include band ligation, cyanoacrylate (CYA) injection, and thrombin. The greatest evidence exists for CYA injection, which is recommended as first-line endoscopic therapies in both the American Association for the Study of Liver Diseases guidelines and by the Baveno V consensus.[12,13] CYA injection is also indicated as the first-line endoscopic treatment of ectopic small bowel varices.[14] Main complications include pulmonary or portal vein embolism. Recently, Bhat et al reported the six-year experience of EUS-guided treatment of gastric fundal varices with combined injection of coils and CYA in 152 patients.[15] Treatment was technically successful in the majority (99%), with re-bleeding in only 3% during long-term follow-up. The injection of CYA with coils under EUS guidance appears to increase the efficacy, and may reduce the risk of CYA embolization.

A patient with active bleeding unresponsive to conservative management should be submitted to an emergent splenectomy, which decreases the venous outflow through the collateral circulation and decompresses the associated varices.[1] Embolization of the splenic artery by selective catheterization has been tried with varying success and should be an option in patients who are not medically fit for a splenectomy procedure, depending of local expertise.[16] 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 acceptable course of management in asymptomatic individuals.[9]

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[17] report the clinical presentation, etiologies and outcome in a cohort of 22 patients with LSH. 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 LSH 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 splenoportal 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[18] Splenoportal venous obstruction was found in 35 patients (13.2%) 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 confirmation of gastrointestinal varices in all of them. Moreover, 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 determinate 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

1. Köklü S, Coban S, Yüksel O, Arhan M. Left-sided portal hypertension. Dig Dis Sci. 2007;52:1141–9.
2. Madsen MS, Petersen TH, Sommer H. Segmental portal hypertension. Ann Surg. 1986;204:72–7.

3. Sutton JP, Yarborough DY, Richard JT. Isolated splenic vein occlusion. Review of literature and report of an additional case. Arch Surg. 1970;100:623–6.
4. Wang L, Liu GJ, Chen YX, Dong HP, Wang LX. Sinistral portal hypertension: clinical features and surgical treatment of chronic splenic vein occlusion. Med Princ Pract. 2012;21:20–3.
5. Agarwal AK, Raj Kumar K, Agarwal S, Singh S. Significance of splenic vein thrombosis in chronic pancreatitis. Am J Surg. 2008;196:149–54.
6. Thompson RJ, Taylor MA, McKie LD, Diamond T. Sinistral portal hypertension. Ulster Med J. 2006;75:175–7.
7. Bradley EL. The natural history of splenic vein thrombosis due to chronic pancreatitis: indications for surgery. Int J Pancreatol. 1987;2:87–92.
8. Kokabi N, Lee E, Echevarria C, Loh C, Kee S. Sinistral portal hypertension: presentation, radiological findings, and treatment options – a case report. J Radiol Case Rep. 2010;4:14–20.
9. Weber SM, Rikkers LF. Splenic vein thrombosis and gastrointestinal bleeding in chronic pancreatitis. World J Surg. 2003;27:1271–4.
10. Lewis JD, Faigel DO, Morris JB, Siegelman ES, Kochman ML. Splenic vein thrombosis secondary to focal pancreatitis diagnosed by endoscopic sonography. J Clin Gastroenterol. 1998;26:54–6.
11. Garcia-Tsao G, Sanyal AJ, Grace ND, et al. Prevention and management of gastroesophageal varices and variceal hemorrhage in cirrhosis. Hepatology. 2007;46:922–38.
12. de Franchis R, Baveno V. Faculty Revising consensus in portal hypertension: report of the Baveno V consensus workshop on methodology of diagnosis and therapy in portal hypertension. J Hepatol. 2010;53:762–8.
13. Sarin SK, Jain AK, Jain M, Gupta R. A randomized controlled trial of cyanoacrylate versus alcohol injection in patients with isolated fundic varices. Am J Gastroenterol. 2002;97: 1010–5.
14. Bhat YM, Weilert FF, Fredrick RT, et al. EUS-guided treatment of gastric fundal varices with combined injection of coils and cyanoacrylate glue: a large U.S experience over 6 years (with video). Gastrointest Endosc. 2015. pii:S0016-5107(15)02955-7.
15. Sarin SK, Kumar CKN. Ectopic varices. Clin Liver Dis. 2012;1:167–72.
16. Adams DB, Mauterer DJ, Vujic IJ, Anderson MC. Preoperative control of splenic artery inflow in patients with splenic venous occlusion. South Med J. 1990;83:1021–4.
17. Carmo J, Marques S, Bispo M, Barreiro P, Gonçalves B, Guerra A, et al. Massive gastrointestinal bleeding from an ectopic varix in a non-cirrhotic patient: angiographic approach following endoscopic treatment failure. GE Port J Gastrenterol. 2015.
18. Bernardes P, Baetz A, Lévy P, Belghiti J, et al. Splenic and portal venous obstruction in chronic pancreatitis. A prospective longitudinal study of a medical-surgical series of 266 patients. Dig Dis Sci. 1992;37:340–6.