

Management and conduct of vascular diseases of the portal system

Manejo e conduta das doenças vasculares do sistema porta

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Abstract

Aneurysms and thromboses of the portal vein are rare pathologies of the portal system that commonly follow an asymptomatic course. The vast majority of cases are diagnosed as incidental findings during imaging studies. Symptoms of aneurysms are the result of mass effects, while thrombosis symptoms are a function of the liver's ability to form a collateral circulation network in the thrombosis. The scant experience with such cases poses a dilemma for patient management and so the vast majority of authors choose an expectant approach with rigorous patient surveillance and only intervene in symptomatic patients. We report one case of an aneurysm of the portal vein and one case of portal vein thrombosis and discuss management and observation of these patients.

Keywords: thrombosis; aneurysm; portal system.

Resumo

O aneurisma e a trombose de veia porta são doenças raras do sistema porta, que comumente cursam sem sintomas. A grande maioria dos pacientes é diagnosticada com achados em exames de imagem. Os sintomas são atribuídos ao efeito de massa, no caso do aneurisma, e relativos à capacidade hepática de formar uma rede de circulação colateral, no caso da trombose. A escassa experiência nesses casos representa um dilema na abordagem desses pacientes e, portanto, a grande maioria dos autores opta por seguimento rigoroso e a intervenção é indicada apenas para os pacientes sintomáticos. Neste trabalho, relatamos um caso de aneurisma de veia porta e outro de trombose da veia porta, propondo o manejo e o acompanhamento desses pacientes.

Palavras-chave: trombose; aneurisma; sistema porta.

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INTRODUCTION

Vascular diseases of the portal system include aneurysms and thromboses. Unlike arterial aneurysms, venous aneurysms are rare. They are generally found in the popliteal, jugular and saphenous veins and rarely occur at other sites.¹ Aneurysms of the portal vein (APV) account for 3% of venous aneurysms.² They are most commonly seen in the extrahepatic region, at the confluence between the splenic and superior mesenteric veins, followed by the portal vein and its branches.³ Barzilay and Kleckner⁴ published the first report of a case of APV in 1956, describing aneurysm of the portal vein with thrombosis in its interior that had ruptured into the biliary system, found during an autopsy on a cirrhotic patient.⁴ Since then, around 200 cases of the condition have been described in the literature.⁵

The physiopathogenesis of aneurysm formation has not yet been established, but two theories exist: the first is a congenital condition, resulting from a failure of the distal vitelline vein to completely regress or from weakness of the internal vein wall, while the second is an acquired condition, secondary to chronic liver disease, portal hypertension, trauma, pancreatitis or surgery.³ It is believed that the increased intraluminal pressure observed in portal hypertension may cause dilation of the portal vein's relatively thin walls. However, some authors believe that there must be some other explanation in view of the low incidence of portal vein aneurysms among patients with portal hypertension.⁶

Thrombosis of the portal vein (TPV) describes complete or partial obstruction of blood flow through the portal vein caused by a thrombus within the vessel lumen.⁷ The first description of TPV was published in 1869 by Balfour and Stewart,⁸ and in the Western world the condition is considered the primary cause of extrahepatic portal hypertension among patients with normal livers.⁹ Although it is a rare event in the general population, its prevalence among cirrhotic patients ranges from 4.4 to 15%, and it is responsible for 5 to 10% of cases of portal hypertension.¹⁰

If the portal vein becomes obstructed, the liver loses two thirds of its blood supply. Notwithstanding, the condition is well-tolerated and patients are generally asymptomatic, in contrast with acute arterial obstruction, which always causes severe acute liver dysfunction and can be fatal.^{11,12} Obstruction of the portal vein triggers two compensatory mechanisms: vasodilation of the hepatic artery and development of collateral circulation to divert blood flow away from the obstruction.¹¹ This process leads to loss of hepatic tissues and, over the long term, a consequent loss of liver function.¹³

The etiology of TPV is generally multifactorial, and risk factors can be classified as localized or systemic:^{11,14}

- Localized risk factors: cancer; focal abdominal inflammatory lesions; lesions of the portal venous system (surgery, trauma and iatrogeny), and cirrhosis;
- Systemic: congenital thrombophilias (factor V Leiden, prothrombin gene mutations, hyperhomocysteinemia, C and S protein deficiencies, and antithrombin deficiency); acquired thrombophilias (antiphospholipid antibody syndrome and hyperhomocysteinemia); myeloproliferative diseases; paroxysmal nocturnal hemoglobinuria; oral contraceptive use, and puerperium.

Children can also develop TPV after an episode of omphalitis or after umbilical catheterization.¹⁴

Objective

To describe one case of portal vein aneurysm and one case of thrombosis of the portal vein and their clinical and surgical management.

CASE REPORTS

Case 1. A 27-year-old male patient with chronic pain in the right hypochondrium was admitted to the emergency room, where abdominal ultrasonography was conducted (Figure 1), showing chronic cholelithiasis



Figure 1. Ultrasonography showing the portal vein with a hyperechoic thrombus and echogenic foci, which cause a posterior acoustic shadow. There is no flow. There are also varices around the portal vein in the hepatic hilum. Spleen dimensions are increased.

and TPV. An abdominal computed tomography scan with intravenous contrast confirmed TPV (Figure 2). Laboratory tests did not reveal findings that could explain etiology and so the thrombosis was considered idiopathic. The decision was taken to employ clinical management with 100 mg of acetylsalicylic acid per day, which resulted in clinical improvement, and outpatients follow-up.

Case 2. A 60-year-old asymptomatic male patient was referred to the tertiary healthcare service because of a family history of chronic liver disease. Investigation with abdominal ultrasonography found

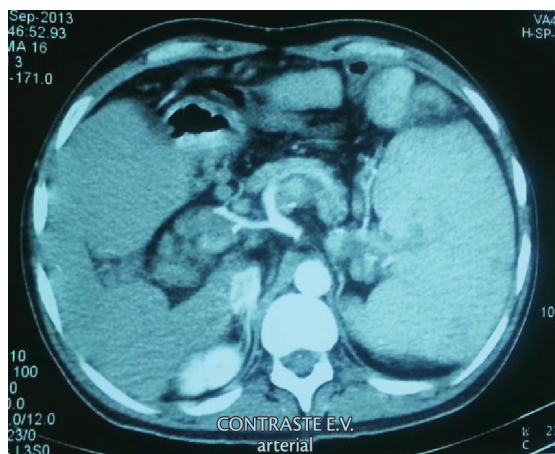


Figure 2. Portal vein without flow and with foci of calcification (chronic thrombosis). Exuberant collateral circulation in the hepatic hilum (cavernomatous transformation). Splenomegaly.

hepatic steatosis and an APV (Figure 3). Computed tomography showed an aneurysmal dilatation of the proximal portal vein with a maximum diameter of 5.5 cm and a 1.1 cm neck, with no signs of thrombosis (Figure 4). Expectant management was also chosen for this patient, with outpatients monitoring.

DISCUSSION

The clinical manifestations of vascular diseases of the portal system vary as a function of the degree to which the hepatic parenchyma is damaged and of the development of compensatory mechanisms.

The great majority of patients with APV are asymptomatic and diagnosis is generally the result of an incidental finding on an imaging exam requested for other reasons. When present, the symptoms are caused by mass effects: abdominal pains, due to compression of neighboring structures, and jaundice, caused by compression of bile ducts. Rupture of the biliary system can also occur, causing gastrointestinal bleeding.^{3,6}

In view of this, the great majority of physicians opt for conservative management, monitoring the patient with sequential imaging exams. In 2009, a systematic review was conducted of 93 reports published in the literature, describing a total of 176 patients with 198 visceral venous aneurysms. The authors concluded that careful observation is the most appropriate conduct, except when complications occur.¹⁵ Recently, a case was described in which a male patient diagnosed with asymptomatic APV was followed for 8 years

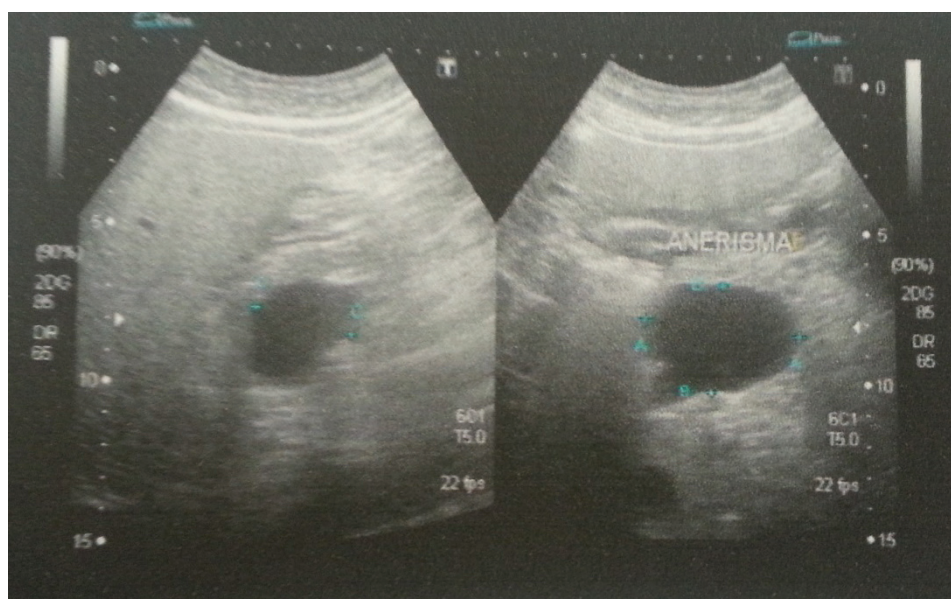


Figure 3. Ultrasonography showing aneurysm of the portal vein.

and enjoyed full regression of the aneurysm, with no explanation.¹⁶

In 2012, Ma et al.⁵ proposed an interesting algorithm for management of patients with APV (Figure 5).⁵

With regard to TPV, the appearance and extent of the thrombus are determinant factors of the resulting clinical manifestations, which in turn can be divided into acute and chronic, and differentiated by emergence of cavernoma (a network of collateral vessels connecting proximal and distal portions of the thrombus, which defines the chronicity of the process).^{10,17} Acute manifestations include intestinal congestion and ischemia, which can cause pain and/

or abdominal distension; diarrhea; rectal bleeding; nausea; vomiting; anorexia; fever; lactic acidosis; splenomegaly, and sepsis. If the obstruction is not resolved rapidly, intestinal perforation, peritonitis, shock and death may follow. On the other hand, chronic TPV can be asymptomatic, except for hypersplenism and consequent pancytopenia, esophageal varices, abdominal collateral circulation and ascites. This fact means that it is necessary to conduct an upper digestive endoscopy on all patients with TPV.¹⁰ Figure 6 illustrates an adaptation of a diagram originally proposed by Chinese researchers that relates the course of TPV to progression of symptoms.¹⁷

Since patients with TPV in the chronic phase are very often asymptomatic, diagnosis is commonly an incidental imaging exam finding. Once clinical suspicion has been aroused, the first examination requested is usually abdominal ultrasonography, which may show solid hyperechoic material in the portal vein; distension of the portal vein and/or tributaries, and a network of collateral vessels or cavernoma. Next, abdominal ultrasonography with Doppler, computed tomography or magnetic resonance imaging may be used to confirm diagnosis.¹⁰

Although resolution of TPV in the acute phase has been described, specific therapeutic management is mandatory to resolve portal obstruction and avoid serious complications, primarily to impede progression to the chronic phase of the process.¹⁸ The objective

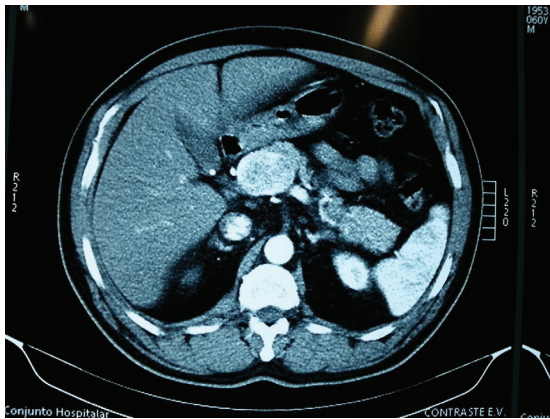


Figure 4. View of aneurysm on abdominal computed tomography with contrast.

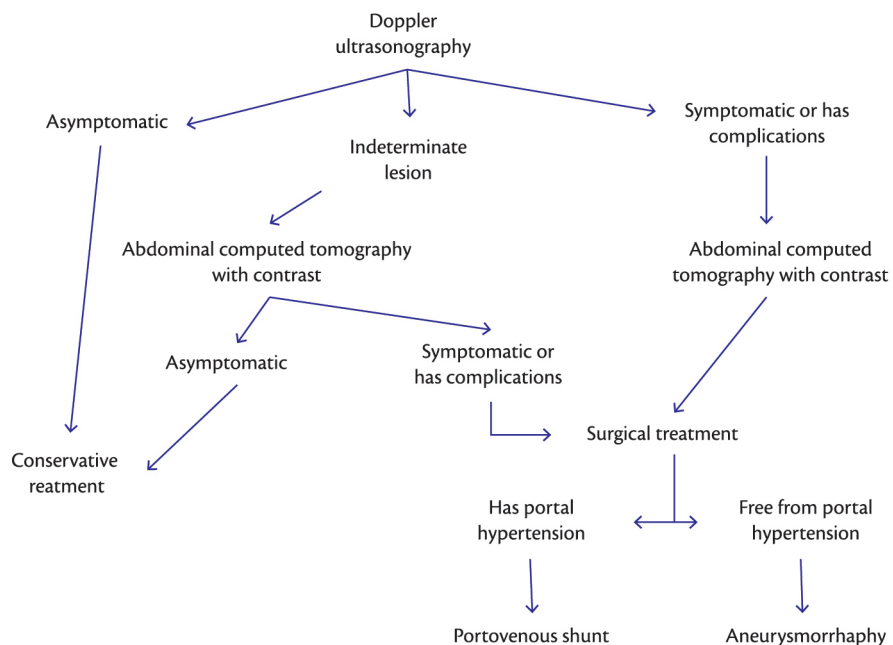


Figure 5. Adaptation of the algorithm proposed by Ma et al.⁵

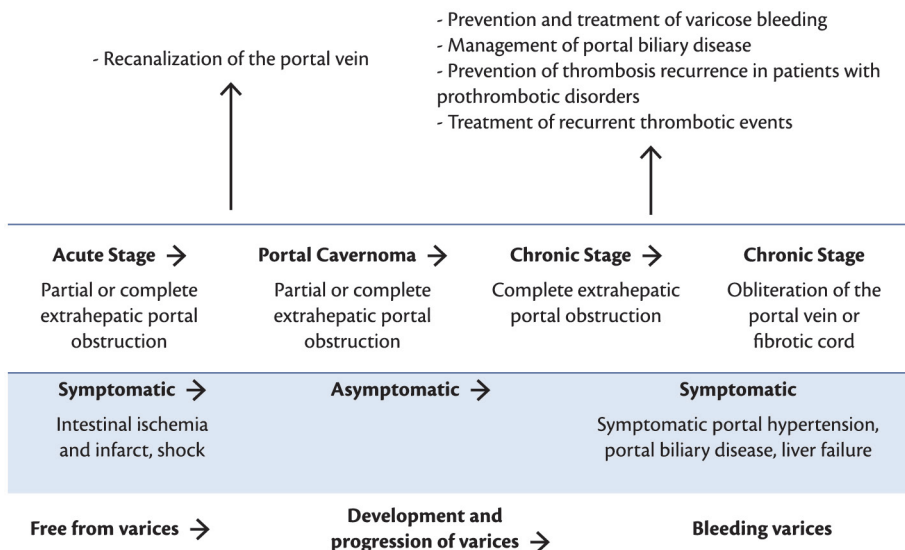


Figure 6. Progression of thrombosis of the portal vein and treatment strategy for patients free from cancer and cirrhosis. Adapted from Qi et al.¹⁷

of treatment is correction of risk factors, prevention of thrombosis growth and achieving patency of the portal vein.¹⁰ The great majority of authors recommend anticoagulant treatment in the acute phase of the disease; but this becomes controversial with respect to the chronic phase, because it could encourage unfavorable progression to esophageal varices.^{11,19} Other authors recommend that anticoagulant treatment should only be prescribed for patients with confirmed thrombotic disorders or a family history of venous thrombosis.²⁰ Other treatments that do not enjoy consensus among authors include: thrombolysis, transjugular intrahepatic portosystemic shunts (TIPS) and surgical shunts (distal splenorenal).^{10,11,19}

Prognosis is more related to the presence of comorbidities, rather than to presence of bleeding esophageal varices.²¹ A study conducted in the city of Bologna conducted a prospective investigation of patients with hepatocellular carcinoma who were candidates for liver transplantation and concluded that TPV that are not caused by tumoral invasion should not be considered as an absolute contraindication to transplantation in patients with hepatocellular carcinoma and cirrhosis of the liver.²² Another study, conducted at the University of Michigan, concluded that presence of TPV may be associated with reduced short-term survival after transplantation, which should be taken into account when taking decisions on indications for transplantation and on the best time to attempt transplantation in these patients.²³

We conclude that vascular diseases of the portal system are rare entities, the great majority of which

can be managed by watchful waiting. Rigorous surveillance is necessary to enable assessment of any emerging need for intervention in cases in which the disease progresses.

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Critical revision of the article: RAB

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