Digestive and Liver Disease 49 (2017) 113–120

Contents lists available at ScienceDirect

Digestive and Liver Disease

journal homepage: www.elsevier.com/locate/dld

Review Article

Portal vein thrombosis: The role of imaging in the clinical setting

Cristina Margini, Annalisa Berzigotti*

Swiss Liver Center, Hepatology, University Clinic for Visceral Surgery and Medicine, Inselspital, University of Bern, Switzerland

ARTICLE INFO

Article history:
Received 4 November 2016
Received in revised form
21 November 2016
Accepted 24 November 2016
Available online 1 December 2016

Keywords:
Cavernoma
Cirrhosis
Neoplastic invasion
Thrombophilia
Ultrasound

ABSTRACT

Portal vein thrombosis is an infrequent condition occurring in several different clinical scenarios. In the last years it has been increasingly recognised due to the broad use of radiological methods. In this review we underline the central role of imaging in diagnosing portal vein thrombosis, in clarifying its etiology, choosing the best therapeutic approach and screening possible complications. Special attention is given to the role of imaging to differentiate portal vein thrombosis from neoplastic invasion of the portal vein, and to new diagnostic methods available for clinical practice in this field.

1. A short clinical introduction to PVT

Portal vein thrombosis (PVT) is the second most common cause of portal hypertension (PH) in the Western world and it is classically defined as the partial or complete obstruction of the portal vein lumen by a clot, or as the finding of its substitution by neo-formed tortuous vessels with hepatopetal flow termed "cavernoma". Thrombosis in the portal venous system is increasingly recognised as the result of predisposing factors acting on one or more elements of the Virchow's triad, namely hypercoagulability, endothelial dysfunction and stasis (Table 1) [1]. Anatomically it can occur in the intra or extrahepatic tract and/or involve the superior mesenteric vein and/or the splenic vein.

The presentation of PVT is very variable, and ranges from asymptomatic incidental findings to severe complications of portal hypertension, variceal bleeding in particular; the number of vessels involved (e.g. portal vein only vs. portal, splenic and mesenteric veins) and the degree of thrombosis (partial vs. complete) influence the clinical features, and the chance to respond to anticoagulation [1]. Additional complexity of PVT is due to the fact that it can occur in several different clinical scenarios, including cirrhosis, haematological diseases in non-cirrhotic subjects (mostly chronic myeloproliferative diseases), abdominal infections/surgery, central obesity [2] and intra- or extrahepatic malignancy, which all cause an hypercoagulable state (Table 1) [1]. In non-cirrhotic subjects, the term "PVT" should be replaced with the term "extrahepatic portal vein obstruction" (EPVHO), which does not include isolated splenic vein or mesenteric vein thrombosis [3]. Due to its complexity, major experts in this field recently suggested to define PVT as a syndrome rather than a disease itself [4]. In this complex scenario, the aim of this review is to describe how imaging methods can help the clinical hepatologist in all phases of the decision-making process related to PVT/EPVHO (in the review generally addressed as PVT).

2. The diagnosis of PVT: who should be suspected of PVT and how to investigate the presence of PVT and its severity

The diagnosis of PVT may occur in two major clinical scenarios: in asymptomatic and in symptomatic patients. If PVT is incidentally discovered the clinician will be asked to make a step backward and look for the risk factors leading to PVT, while in the case of symptomatic patients potentially at risk of carrying PVT the clinician will be asked to make a step forward and ask for the help of imaging to confirm or exclude the diagnosis of PVT. Even when a local risk factor is found or known, additional causes such as systemic prothrombotic conditions should be investigated [5].

The results of a recently published study suggest that the prognosis of incidentally detected splanchnic vein thrombosis is similar to that of clinically suspected splanchnic vein thrombosis suggesting that similar treatment strategies should be applied [6]. In all cases, after the diagnosis the clinician will need to carefully screen

* Corresponding author at: MEM F807, Murtenstrasse 35, CH-3010 Bern, Switzerland. Fax: +41 31 632 49 97.
E-mail address: annalisa.berzigotti@insel.ch (A. Berzigotti).

http://dx.doi.org/10.1016/j.dld.2016.11.013
1590-8658/

Table 1
Major risk factors of portal vein thrombosis in adults and children.

Major risk factors for bland portal vein thrombosis in adults
Liver cirrhosis
Neoplastic conditions
Overt myeloproliferative disease
Solid abdominal neoplasia (e.g. HCC, pancreatic carcinoma)
Thrombophilic conditions
Subclinical myeloproliferative disease (JAK 2—Mutation)
Antiphospholipid syndrome
Protein C and S deficiency
Antithrombin III deficiency
Prothrombin gene mutation
Factor V Leiden
Homozygous MTHFR mutation
Paroxysmal nocturnal hemoglobinuria
Hyperhomocysteinemia
Connective tissue disease
Hormonal contraception or replacement therapy
Personal history of deep vein thrombosis
Family history of deep vein thrombosis
Abdominal surgery
e.g. liver surgery, colon-rectal surgery, splenectomy, sleeve gastrectomy, cholecystectomy, pancreatectomy, hysterectomy
Abdominal inflammatory/infectious process
e.g. Acute pancreatitis, cholecystitis or cholangitis, liver abscesses, gastritis, inflammatory bowel disease, diverticulitis, cytomegalovirus hepatitis, tubercular lymphadenitis
Abdominal trauma
Major risk factors for benign portal vein thrombosis in children
Umbilical vein catheterisation
Neonatal sepsis
Abdominal infection
Cardiovascular malformation
Coagulation disorder
Abdominal surgery

for complications of PVT and plan the most appropriate therapy and follow up.

The main clinical presentation of acute PVT is abdominal pain, especially if the superior mesenteric vein is involved. In chronic PVT the clinical presentation is related to the development of pre-hepatic portal hypertension (ascites, variceal bleeding, encephalopathy) or portal cholangiopathy (jaundice, abdominal pain, cholangitis).

When PVT is suspected ultrasound is the first line imaging method to be used (Fig. 1), since it holds an accuracy ranging 88–98% for the detection of PVT with a sensitivity and specificity of 80–100% in the majority of studies [7–11]. The sensitivity of ultrasound is particularly high in complete PVT, while the risk of false negative results occurs only in incomplete PVT [12] and isolated superior mesenteric vein thrombosis [8].

In 2-D Gray-Scale ultrasonography a thrombus appears as a hypo/isoechoic material occupying part (partial thrombosis) or the entire vessel (complete thrombosis) (Fig. 2). The normal portal vein can be eventually replaced by multiple tortuous vessels with hepatopetal flow, a condition, named "cavernomatous transformation" or "cavernoma", that can be also easily detected with Doppler-ultrasound (Fig. 2). Colour/power and pulsed Doppler should be mandatorily used to confirm whether the vessel has a remnant blood flow, so helping differentiating high degree partial thrombosis to complete thrombosis.

The reliability of ultrasonography in the detection of PVT improves with the operator's experience and whenever PVT is clinically suspected ultrasonography should be performed by experienced operators [13]. Ultrasonography suffers from other limitations, such as reduced visualisation in obese individuals and in case of abundant bowel gas and impossibility to assess bowel ischemia. This should be suspected in case of ascites and/or high blood lactate levels.

Ultrasound is sufficient to diagnose PVT in patients with a good acoustic window, but when ultrasonography is insufficient to clarify whether PVT is present or absent (for instance in patients with insufficient visualisation), a second line cross-sectional imaging method should be considered to confirm or exclude the diagnosis.

Contrast-enhanced 4 Phases (pre-contrast, arterial, portal and late) CT (CECT) and contrast-enhanced MRI (CEMRI) can be used, being CT preferred in unstable patients with acute abdominal symptoms. Advantages of MR and CT over US include the possibility of detecting bowel ischemia, septic foci, and intraabdominal malignancies, and higher sensitivity in the detection of thrombosis in the splenic and superior mesenteric vein (Table 2). The drawbacks of CT are well known and include exposure to ionizing radiation, the risk of allergic reactions and nephrotoxicity. CEMRI is also contraindicated in patients with acute renal failure for the risk of nephrogenic systemic fibrosis [14]. The use of unenhanced magnetic resonance portography is currently under investigation [15–17] but it is not yet recommended in clinical practice.

Once PVT is diagnosed, CECT or CEMRI are mandatory to evaluate the extent of thrombosis and to allow a detailed mapping of porto-systemic collaterals (Fig. 1), crucial to the planning of interventions aimed at recanalising the PV system. It should be considered that clinical consequences of PVT mainly depend on the number of vessels completely occluded [1], as well on the degree of collateralization in chronic cases. Furthermore, the presence of ascites is a predictor of lack of response to anticoagulation, and should be reported [1]. Several classification/staging systems have been developed, but mostly rely upon anatomical considerations. The most commonly cited and used in clinical trials is the one proposed by Yerdel et al. [18]. However, there is no validated classification to be used in clinical practice to personalise risk assessment and guide therapy [4].