Acute Portal Vein Thrombosis: Current Trends in Medical and Endovascular Management

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Abstract

Acute portal vein thrombosis (PVT) is a relatively rare diagnosis with a nonspecific clinical presentation. Imaging plays an important role in establishing the diagnosis as well as the etiology and complications of acute PVT. Prompt diagnosis is essential to prevent catastrophic short-term complications including bowel infarction, sepsis, and possible death; missed diagnosis can also result in the long-term sequelae of portal hypertension. Differentiation of acute from chronic PVT is crucial as management strategies differ. Currently, guidelines for treating acute PVT recommend immediate initiation of systemic anticoagulation. Catheter-directed therapy may be used in combination with systemic anticoagulation in the setting of bowel ischemia or as an adjunct in patients with a contraindication to systemic anticoagulation. In this review article, we discuss the diagnosis and clinical features of acute PVT, focusing on current medical and endovascular management strategies including mechanical thrombectomy and fibrinolytic therapy.

Keywords

- portal hypertension
- portal vein thrombosis
- thrombolysis
- thrombectomy

Objectives: Upon completion of this article, the reader will be able to discuss the diagnosis and management of acute portal vein thrombosis as well as describe the indications for endovascular treatment of this condition.

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Acute portal vein thrombosis (PVT) is characterized as recent (<30 days) formation of thrombus within the main portal vein or its branches. Although acute PVT is an uncommon diagnosis, it can lead to serious morbidity, including

bowel infarction, as well as mortality. The clinical presentation of acute PVT can range from asymptomatic in partial PVT to sepsis and hypotension in the patients with acute complete PVT. The most common symptom in patients with acute PVT is nonspecific abdominal pain.¹

Because of the nonspecific symptomatology, the diagnosis is usually made by imaging, primarily contrast-enhanced computed tomography (CT) or ultrasound during the work-up for abdominal pain. If the diagnosis of acute PVT is made with Doppler ultrasound, further evaluation with CT or magnetic resonance imaging (MRI) (►Fig. 1) is warranted to evaluate (►Fig. 2) the extent of the thrombosis, assess for etiology, and provide information about associated complications, such as bowel ischemia.

It is important to differentiate acute from chronic PVT as both entities are managed differently. The presence of a cavernoma, a network of prominent collateral vessels in the porta hepatis, indicates chronic PVT. Diagnosis of malignant PVT is important as therapeutic strategies differ, which can include locoregional therapy or chemotherapy based on tumor type.² Imaging features that suggest malignant PVT

Fig. 1 (a) Grayscale imaging of the main portal vein demonstrating echogenic material within the lumen of the main portal vein (arrowhead). (b) Spectral Doppler ultrasound of the main portal vein demonstrating absent flow within the main portal vein.

include enhancing thrombus on contrast-enhanced CT or MRI, the presence of flow on Doppler ultrasound, or invasion of the wall of the portal vein.

There are several factors that increase risk for the development of acute PVT. Local factors include cancer within any abdominal organ, cirrhosis, extrinsic compression of the portal vein and/or its branches, postoperative patients, and

patients with abdominal infections or inflammation such as pancreatitis or inflammatory bowel disease. Local factors play a role in acute PVT ~21 to 30% of the time.^{3,4} Systemic risk factors include prothrombotic disorders, and are present in up to 70% of patients who present with acute PVT.⁵⁻⁸ Prothrombin gene mutation has a nearly 4.5-fold increased risk of PVT, while factor V Leiden mutation has a nearly twofold risk of acute PVT.^{8,9} Myeloproliferative disorders such as Janus kinase 2 (JAK2) mutation are found in 21 to 37% of patients with acute PVT.^{8,10-12} A prothrombotic state may also result from recent contraceptive use as well as deficiencies of antithrombin, protein C, and protein S.⁶⁻⁸

Timely therapy for acute PVT is essential to prevent propagation of thrombus and restore adequate venous outflow from the small bowel to prevent small bowel infarction and sequelae of portal hypertension. If a patient is found to have a local risk factor, work-up for prothrombotic disorders must still be performed, as 35% of patients may have both local and systemic risk factors for PVT. Approximately 25% of patients who develop acute PVT will have no identifiable risk factor.⁸ In this article, the data and rationale for medical and endovascular therapy will be reviewed, along with devices frequently used in endovascular procedures.

Medical Therapy

Currently, the first-line treatment for management of acute PVT is systemic anticoagulation. Prior studies have shown that without systemic anticoagulation, spontaneous recanalization occurs in only 16.7% of cases.¹³ Collective data from retrospective studies have shown when anticoagulation is initiated immediately and continued for 6 months, 50% of patients had complete recanalization, 40% had partial recanalization, and 10% of patients had no recanalization.^{14,15} Major complications from anticoagulation were relatively low, occurring in less than 5% of patients. Prospective multicenter data have demonstrated a 1-year recanalization rate of 38%, with no recanalization in any of the patients beyond the sixth month after initiation of

Fig. 2 (a) Axial postcontrast computed tomography (CT) image of the upper abdomen demonstrating low-density material within the main portal vein, consistent with thrombus (arrowhead). (b) Coronal reconstructed image demonstrating low-density material within the main portal vein, consistent with thrombus (arrowhead).

Fig. 3 Digital subtraction venography images demonstrate: (a) a large amount of thrombus within the main portal vein, extending into the proximal superior mesenteric vein; (b) creation of a transjugular portosystemic shunt; (c) mechanical thrombectomy used to fragment and aspirate clot from the portal and superior mesenteric veins; (d) small amount of persistent eccentric clot, remainder of portal vein and TIPS is patent.

anticoagulation treatment; factors that may predict failure of recanalization included presence of ascites, JAK2 mutation, and superior mesenteric venous thrombus. Furthermore, initiation of anticoagulation is essential; data have shown that if there is a delay the likelihood of recanalization is very low.⁸

Currently, the European Association for the Study of the Liver (EASL) recommends anticoagulation for at least 6 months, whereas the American Association for the Study of Liver Diseases (AASLD) recommends anticoagulation for at least 3 months in patients with acute PVT.^{16,17} If an underlying prothrombotic condition is identified, longer term systemic anticoagulation therapy would be recommended.

Endovascular Therapy

Endovascular treatment of acute PVT may be used in conjunction with systemic anticoagulation in the setting of imminent bowel infarction or as the sole therapy in patients with a contraindication to systemic anticoagulation. Several endo-

vascular techniques have been described for treating acute PVT via direct or indirect access into the portal system, including catheter-delivered fibrinolytic therapy and mechanical/suction thrombectomy with or without fibrinolytic therapy.

Direct percutaneous access can be obtained through a transhepatic approach, although there is a theoretical increased risk of bleeding and it may require embolization of the access tract. The portal system can also be directly accessed through creation of a transjugular intrahepatic portosystemic shunt (TIPS), trans-splenic access, and others. Indirect routes include infusion of thrombolytics into the superior mesenteric artery (SMA), exposing portal venous thrombus to fibrinolytic via the small bowel capillaries.

Mechanical Thrombectomy

Mechanical thrombectomy involves restoring flow within the main portal vein using balloon thrombectomy, rheolytic thrombectomy, and suction thrombectomy. Successful use of mechanical thrombectomy-assisted thrombolysis has been previously described in the literature, although no

prospective data are available to compare different techniques with outcomes.¹⁸

Balloon thrombectomy involves inflation of the balloon past the clot (Fogarty, Edwards Lifesciences, Irvine, CA) and retracting the inflated balloon over the guidewire to pull the clot into a patent vein, which then washes it away. This is usually followed by angioplasty to increase luminal gain and macerate any residual thrombus.

Rheolytic therapy uses high-velocity saline jets for maceration of clot, allowing for passive thrombus evacuation through the Bernoulli principle. The rheolytic thrombectomy (AngioJet device) has been used off label in the portal venous system. In addition to mechanical thrombectomy, this device also has a "power-pulse mode" to distribute thrombolytic agents into the clot.

Suction thrombectomy has evolved from the most basic setup of having a large-volume syringe on the end of a large-bore catheter to the use of vacuum-operated thrombectomy tools.

(► **Fig. 3**) The aspiration thrombectomy (Penumbra Indigo system) is a continuous suction device, most commonly used for neurovascular and peripheral interventions. Catheters are available in numerous sizes; the accompanying CT 8, flexible 8F aspiration catheter is the most likely size to be used for PVT intervention. Unlike the rheolytic thrombectomy (AngioJet device), the aspiration thrombectomy (Penumbra Indigo system) does not return volume to the patient and may lead to rapid volume loss if thrombus is not directly engaged at the end of the catheter.

Thrombolytic Therapy

If there is no contraindication to anticoagulation, another option is passive infusion of fibrinolytic medications (alteplase, Genentech, South San Francisco, CA) directly into acute thrombus. This is usually accomplished with a multiple side hole infusion catheter (Cragg-McNamara, Medtronic, Minneapolis, MN; or UniFuse, Angiodynamics, Latham, NY). These catheters are typically 4F to 5F and are available in a variety of working/infusion lengths.

Data from a retrospective review examining recanalization of acute noncirrhotic, nonmalignant PVT via transjugular thrombolysis with and without TIPS stent placement (Viatore, Gore Medical, Flagstaff, AZ) in 17 patients demonstrated complete recanalization in 52.9% (9/17) and partial recanalization in 41.2% (7/17).¹⁹ Cumulative 24-month secondary portal vein patency was 88.2%; only one patient developed cavernous transformation of the portal vein.¹⁹ Clinical success was demonstrated by prevention of bowel resection in 15/17 patients as well as no patient with portal hypertensive complications during follow-up. These data showed presence of the JAK2 mutation predicted necessity of TIPS implantation; furthermore, the risk of PVT recurrence or TIPS thrombosis was significantly higher in these patients.¹⁹ However, TIPS implantation in noncirrhotic patients remains a controversial issue as prior authors have demonstrated slow flow in the portal vein to be biggest predictor of PVT.²⁰

In a small series of 20 patients investigating thrombolysis via direct transhepatic access or indirectly through the SMA,

the majority of patient had partial recanalization 12/20 (60%).²¹ Only 3/20 patients (15%) had complete recanalization and there was no recanalization in 5/20 (25%) patients.²¹

Conclusion

Diagnosis of acute PVT is challenging; the managing clinician must have a high index of suspicion; prompt diagnosis is critical in instituting early therapy, as this can improve both short- and long-term outcomes. Although first-line therapy is systemic anticoagulation, endovascular therapy is indicated in patients with evidence of impending bowel ischemia or a concurrent contraindication to anticoagulation. Several endovascular techniques have now been described for treating acute PVT via direct and indirect access into the portal system and include mechanical techniques with or without thrombolytic therapy, as well as conventional fibrinolytic infusion via infusion catheters. Prospective data are needed to compare management techniques and evaluate outcomes.

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