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ABDOMINAL IMAGING

PICTORIAL ESSAY

Cross-sectional imaging of congenital and acquired abnormalities of the portal venous system

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ABSTRACT

Knowing the normal anatomy, variations, congenital and acquired pathologies of the portal venous system are important, especially when planning liver surgery and percutaneous interventional procedures. The portal venous system pathologies can be congenital such as agenesis of portal vein (PV) or can be involved by other hepatic disorders such as cirrhosis and malignancies. In this article, we present normal anatomy, variations, and acquired pathologies involving the portal venous system as seen on computed tomography (CT) and magnetic resonance imaging (MRI).

A wide range of congenital and acquired abnormalities may affect the portal venous system. They are sometimes encountered incidentally because of the widespread use of cross-sectional imaging modalities. Awareness of the presence of these abnormalities is important for adequate planning of surgical and percutaneous interventional procedures. The article reviews the normal anatomy, variations, and acquired pathologies involving the portal venous system as showed on computed tomography (CT) and magnetic resonance imaging (MRI).

Anatomy

The portal venous system refers to the veins draining the gastrointestinal tract and spleen to the liver, excluding the inferior rectum and anal canal. The superior mesenteric vein (SMV) and splenic vein (SV) behind the neck of the pancreas form the main portal vein (PV) (Fig. 1). The inferior mesenteric vein (IMV) may drain into either the SV or into the confluence directly. The PV divides into the right and left PV. The right PV divides into anterior and posterior branches that supply the right liver lobe. The left PV courses horizontally and branches out to supply the lateral and medial segments of the left lobe (1,2).

Congenital abnormalities

Variations of the portal vein

Anatomic variants of the branching pattern of the intrahepatic PV are present in approximately 20% of the population (1). The most common variant is trifurcation of the PV; the right PV does not exist and the PV divides into the right anterior, right posterior, and left branches (Fig. 2). Right anterior segmental branch arising from the left PV is another common variant (Fig. 3). Less common PV variations have been reported but their incidence has not been found to be higher than 2% (2). Awareness of these variants is crucial for planning surgery; for example, in a patient with right anterior segmental branch arising from the left PV, ligation of the left PV during left hepatectomy may lead to unexpected infarction of the anterior segment of the right liver lobe.

Congenital agenesis of the portal vein

Congenital agenesis of the PV is a rare malformation characterized by the absence of the PV and anomalous drainage of SMV and SV into the systemic circulation (Fig. 4). The PV is normally formed in 4–10 weeks of embryonic development via involution of the perintestinal vitelline venous loop. Atypical involution may cause a prebiliary, preduodenal, or duplicated PV, and excessive involution may result in the absence of the PV (3). Liver morphology is generally preserved. Congenital agenesis of the major branches of the PV is

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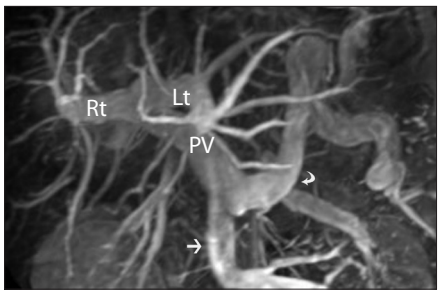


Figure 1. Coronal MIP reformation of contrast-enhanced magnetic resonance angiography (MRA) shows normal anatomy of the portal venous system in this patient with portal hypertension. Note the splenic *(curved arrow)* and superior mesenteric *(arrow)* veins form the main PV, which further divides into the right (Rt) and left (Lt) branches at the porta hepatis.

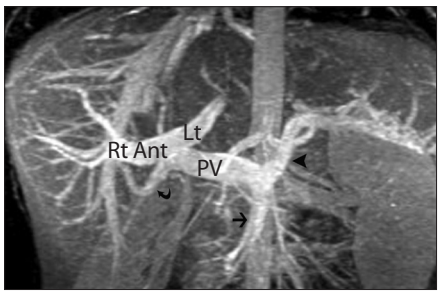


Figure 2. Coronal MIP reformation of contrast-enhanced MRA demonstrates trifurcation of the PV. No right PV is present; right anterior (Rt Ant), right posterior *(curved arrow)* and left PV arise from the main PV at the porta hepatis. Note SV *(arrowhead)* and SMV *(arrow)*.

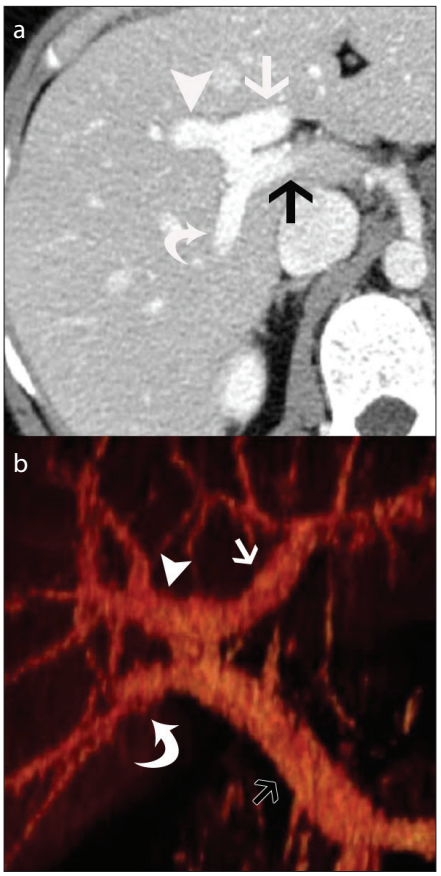


Figure 3. Axial contrast-enhanced CT **(a)** and coronal MIP reformation of CT angiography **(b)** show that the anterior branch of the right PV *(arrowhead)* is arising from the left PV *(white arrow)*. Note posterior branch of the right PV *(curved arrow)* arising from the main PV *(black arrow)*.

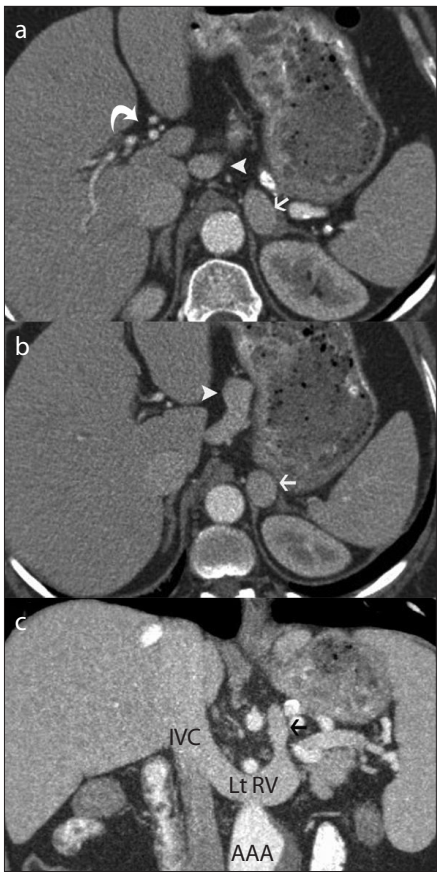


Figure 4. A 58-year-old man with a history of ischemic cardiomyopathy and congestive heart disease presented with progressively elevated liver enzymes and hepatic encephalopathy. Contrast-enhanced CT images **(a–c)** show only the hepatic artery *(curved arrow)* in the porta hepatis with absence of the main PV and its major branches. The spleno-mesenteric veins were drained by an anomalous collateral vein *(arrowheads)* arising from the portal venous confluence, coursing superiorly and eventually reaching the inferior vena cava through the left adrenal gland *(arrows)* and left renal (Lt RV) veins.

Main points

- A thorough knowledge of CT and MRI features of anatomical variations, congenital, and acquired abnormalities of the portal venous system will help radiologists to interpret these examinations correctly, which may play a significant role for patient management.
- CT and MRI allow noninvasive evaluation of the portal venous system for anatomical and functional abnormalities as well as vascular guidance to help percutaneous interventional procedures and surgical planning. These modalities have replaced conventional angiography in the assessment of the portal venous system.
- The spectrum of abnormalities of the portal venous system is broad and includes potentially life-threatening conditions such as portal venous gas. The key to successful recognition of abnormalities of the portal venous system depends on familiarity with the normal and variant anatomy and the characteristic imaging findings of abnormalities affecting the portal venous system.

common and should be distinguished from the absence of small calibered PV branches due to acquired atrophy of the liver lobes.

Portal vein aneurysms

Aneurysms of the PV represent only 3% of all aneurysms of the venous system (4). PV aneurysms occur mainly at the sites of the splenomesenteric venous confluence (Fig. 5), main PV (Fig. 6), and intrahepatic PV branches at bifurcation sites (5, 6). The pathogenesis of PV aneurysms is controversial and it can be congenital or acquired, as a result of weakening of the venous wall. Incomplete regression of the distal right vitelline vein and an inherent weakness of the vessel wall are proposed theories to support a congenital origin (7). Acquired factors may include portal hypertension, chronic hepatic disorder, necrotizing pancreatitis, trauma, and the sequelae of abdominal surgery. Most PV aneurysms need no treatment, since they typically do not show a significant increase in diameter and regular follow-up would be sufficient.

Intrahepatic vascular shunts

Intrahepatic vascular shunts may be congenital or acquired and are described as abnormal communications between the hepatic artery, the hepatic vein, and the PV. Congenital portosystemic shunts are rare anomalies that can be seen incidentally when an infant undergoes ultrasonography (US) for other reasons. Several forms of intrahepatic portosystemic shunts have been reported (8, 9). The most common form is a single large tube that connects the right PV to the inferior vena cava. The other form of intrahepatic portosystemic shunt is a com-

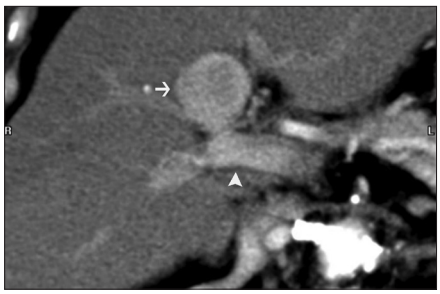


Figure 5. A 64-year-old woman with metastatic breast cancer. Axial contrast-enhanced CT scan shows a saccular aneurysm *(arrow)* arising from the main PV *(arrowhead)*.

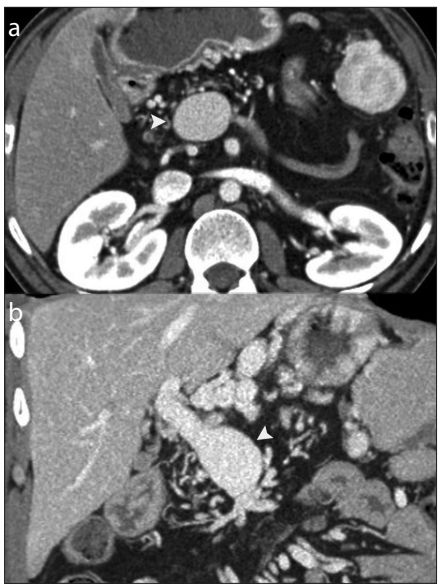
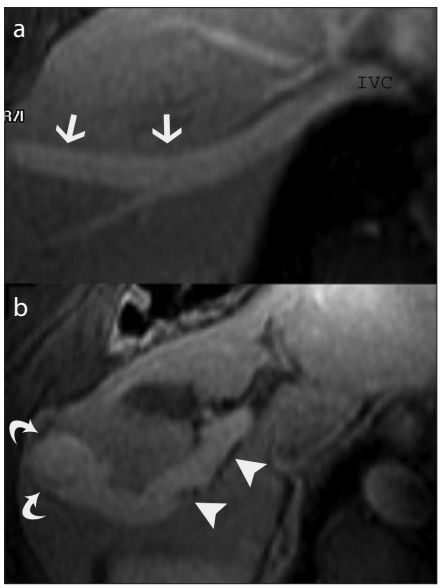


Figure 6. A 23-year-old man with cystic fibrosis, cirrhosis and portal hypertension. Axial **(a)** and coronal **(b)** contrast-enhanced CT images show marked fusiform aneurysmal dilation *(arrowhead)* of the portal venous confluence.



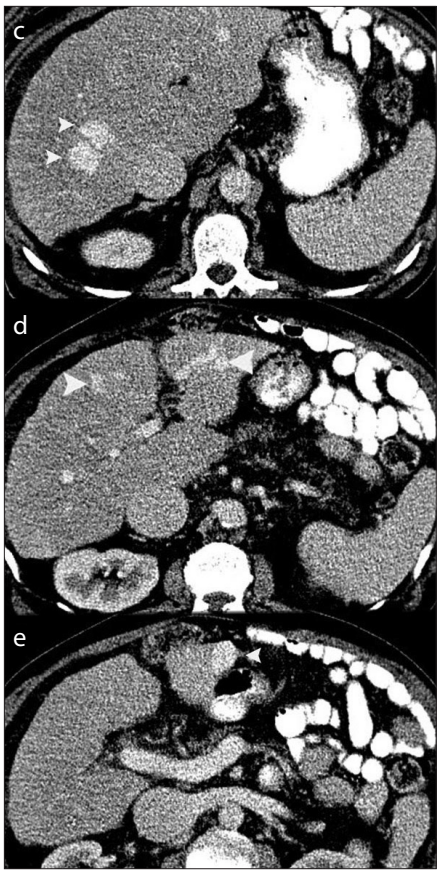


Figure 7. a–e. Axial contrast-enhanced MRI scans of a 73-year-old man with hepatic encephalopathy **(a,b)** show markedly enlarged right PV *(arrowheads)* connected directly to the right hepatic vein *(arrows)*, through a large segment *(curved arrows)* in the right liver lobe, compatible with congenital portosystemic shunt. Axial contrast-enhanced CT images of a 67-year-old man with painless jaundice **(c–e)** show multiple, markedly enhancing foci *(arrowheads)* in the liver, which continue with both distal branches of the PV and hepatic veins, consistent with intrahepatic portosystemic shunts.

munication between peripheral PV and hepatic vein branches through an aneurysm (Fig. 7). Most patients with intrahepatic portosystemic shunt have clinical signs of portal hypertension and cirrhosis, so correct radiologic diagnosis and proper treatment of this abnormality is important.

Arterioportal shunt is a connection between the hepatic arterial branch and the portal venous system that leads to redistribution of arterial flow into the PV. Liver neoplasm, cirrhosis, and iatrogenic reasons (Fig. 8) are the most common causes of acquired arterioportal shunts. Arterioportal shunt can also occur in liver trauma (Fig. 9). Transient hepatic attenuation difference (THAD) refers to parenchymal enhancement during the arterial phase to compensate reduced portal flow and occurs secondary to a tumor, inflammation, thrombosis or compression of a PV branch. THADs appear as wedge-shaped areas during early hepatic arterial phase that return to nearly normal attenuation during portal venous phase (Fig. 10) (10).

Acquired abnormalities

Portosystemic collaterals

Portosystemic collaterals develop in patients with end-stage cirrhosis or portal hypertension to compensate for increased portal blood flow that cannot be sustained due to increased intrahepatic venous pressure. Contrast-enhanced CT at portal venous phase should be the first-line imaging modality when portosystemic collaterals are suspected. Varices appear on CT scans as well-defined, round, tubular, or serpentine configurations and enhance with contrast agent similar to other adjacent veins. Due to increased venous pressure and stasis, the portal venous flow is redirected towards the systemic circulation through many portosystemic shunts including gastroesophageal (Fig. 11), paraumbilical, splenorenal, inferior mesenteric, and retroperitoneal pathways. Among these collateral pathways, esophageal varices are the clinically most important, because they are the most common cause of gastrointestinal hemorrhage. Information about other collateral pathways is also relevant, especially when interventional procedures or surgery is planned. On CT scans, esophageal varices appear as enhancing intraluminal protrusion with scalloped contours and related wall thickening. The paraesophageal varices are situated outside the wall of the esophagus. They can be so massive that they may mimic a posterior mediastinal lesion. Therefore, CT is more sensitive to demonstrate esophageal varices than US. Portosystemic venous collaterals may also develop in response to occlusion of the PV and its tributaries due to neoplastic, infectious, or inflammatory processes (most frequently pancreatitis, causing SV occlusion) (Fig. 12) and iatrogenic reasons.

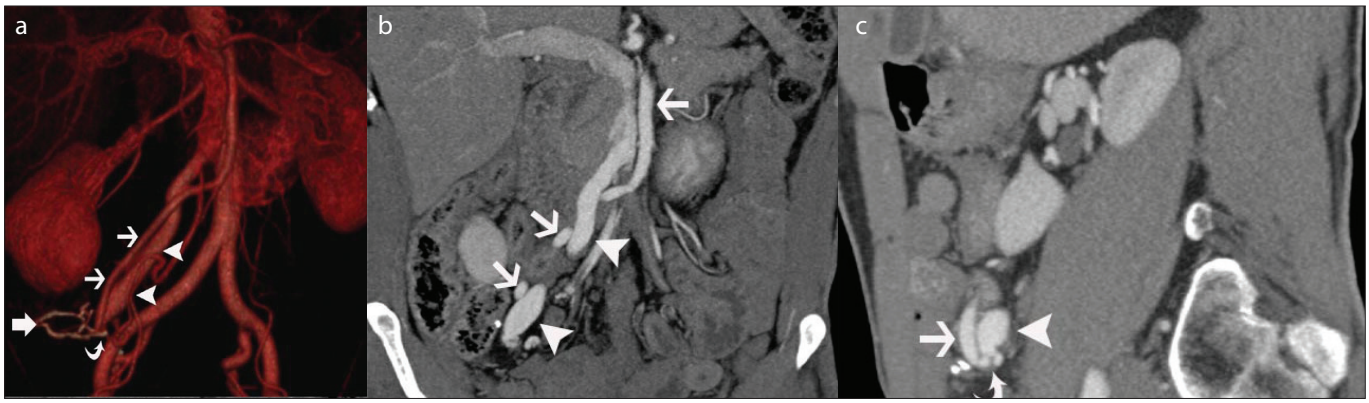


Figure 8. a–c. A 53-year-old male patient with the history of prior iliocecal resection due to cecal volvulus presented with prominent bruit in the right lower quadrant. Coronal volume rendering **(a)** and coronal **(b)** and sagittal **(c)** maximum intensity projection (MIP) CT angiography images show marked enhancement of the SMV *(arrowheads)* during arterial phase consistent with an arteriovenous fistula *(curved arrow)* between the SMA *(arrows)* and SMV. Note surgical sutures at the region of the fistula *(thick arrow)*.

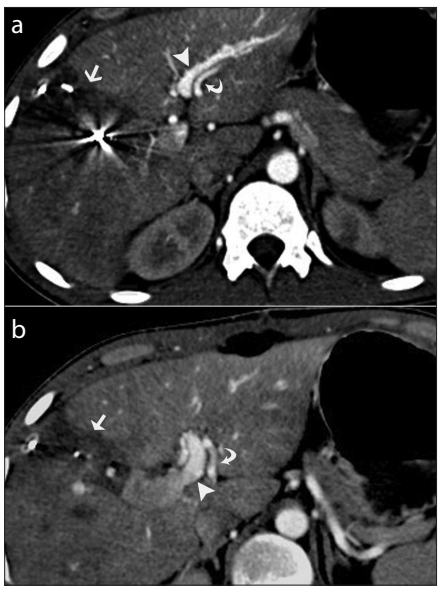


Figure 9. A 19-year-old male who sustained a stab wound to the liver and was treated with embolization of the right hepatic artery. On follow-up CT angiography **(a, b)**, there was marked enhancement of the left PV branch *(arrowheads)* during arterial phase due to a fistula from the left hepatic artery *(curved arrows)*. Note changes in the right liver lobe *(arrows)* secondary to prior surgery and embolization.

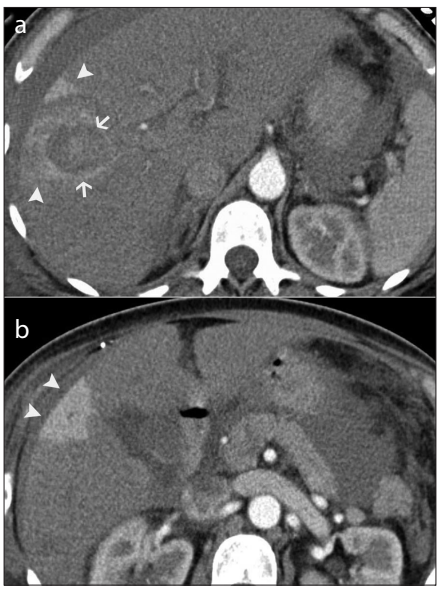


Figure 10. A 49-year-old female with breast cancer metastasized to the liver, which was treated with cryoablation. Axial contrast-enhanced CT scan **(a, b)** shows markedly enhanced peripheral wedge-shaped area *(arrowheads)* in the right liver lobe during the early arterial phase representing an arteriovenous shunt due to thrombosis of the distal branch of the right PV. Note ablated metastatis *(arrows)*.

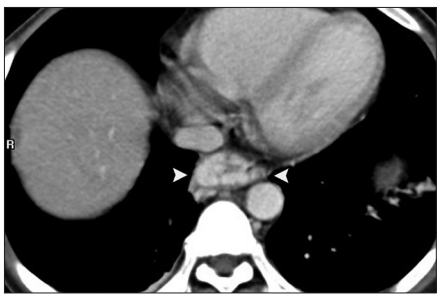


Figure 11. Axial contrast-enhanced CT scan shows marked esophageal varices *(arrowheads)*.

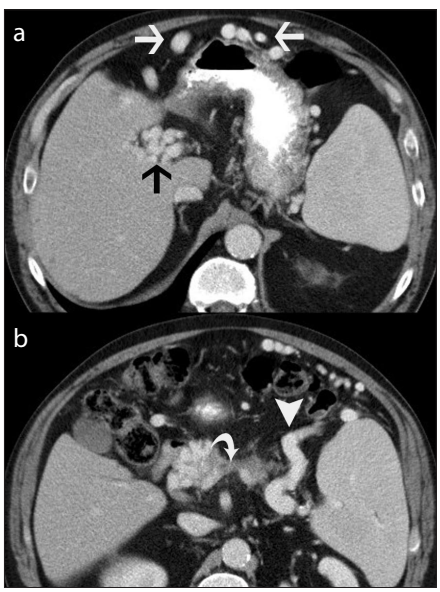


Figure 12. A 59-year-old man with locally advanced pancreatic islet cell tumor of the pancreas. Axial contrast-enhanced CT **(a, b)** shows large venous structures *(white arrows)* adjacent to the greater curvature of the stomach, representing markedly dilated short gastric vein secondary to chronically occluded splenic vein near the portal venous confluence due to patient's known pancreatic cancer *(curved arrow)*. Visualization of this vein on CT or MRI scan already indicates high-grade stenosis or occlusion. Note cavernous transformation of the PV *(black arrow)* and dilated SV *(arrowhead)* in the splenic hilum.

Radiologists should be familiar with the various imaging findings of these portosystemic shunts, as some of these cases can be treated with interventional techniques such as endoscopic sclerotherapy or ligation for prevention of variceal bleeding (11, 12).

Cavernous transformation of the portal vein

Cavernous transformation of the PV consists of dilatation of multiple venous channels within or around the chronically occluded PV. Three different etiopathogenic theories have been proposed about cavernous transformation of the PV: 1) it is a congenital malformation which replaces an undeveloped PV (13); 2) it is a hemangioma of the paracholedochal veins, and 3) it develops as an end-product of thrombosis of the PV (14). However, little is known about the evolution of cavernous transformation or the intra-extra hepatic blood flow modifications that follows obstructed PV. On contrast-enhanced CT scans, small, numerous, tortuous enhancing vessels replacing the main PV is the characteristic imaging finding (Fig. 13). The development of these

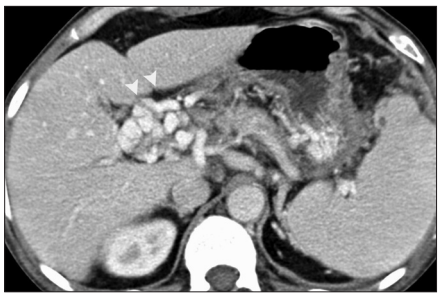


Figure 13. A 53-year-old man with a history of HIV, alcohol abuse, and several episodes of pancreatitis. Axial contrast-enhanced CT scan shows "cavernous transformation", in which numerous small venous channels *(arrowheads)* replace the main PV, a condition that typically develops secondary to chronic PV thrombosis.

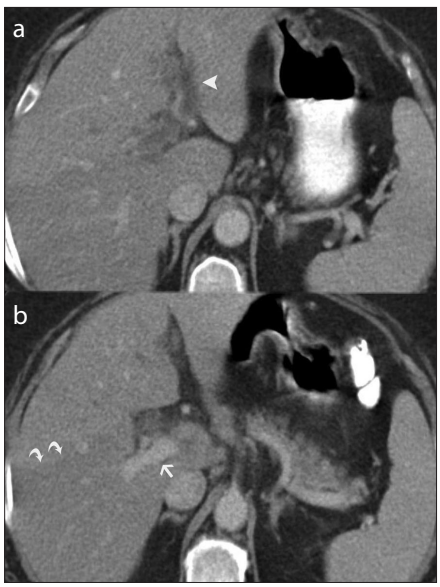


Figure 14. A 59-year-old female being treated for diverticulitis. Contrast-enhanced CT images **(a, b)** show hypoattenuating-filling defect in the left PV *(arrowhead)*, consistent with thrombus. Note the difference in enhancement pattern between the right and left lobes *(curved arrows)* and the the patent right branch of the PV *(arrow)*.

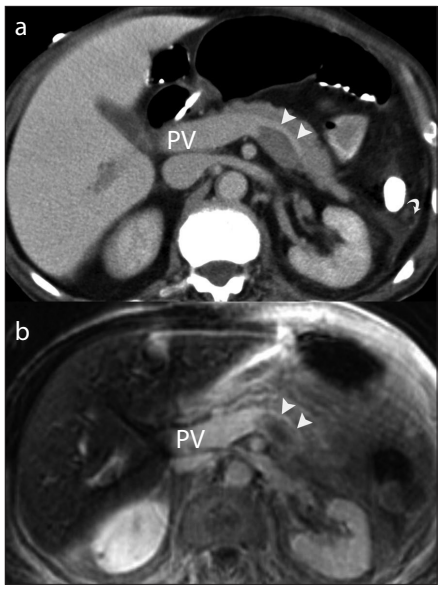


Figure 15. A 75-year-old man one-week status post splenectomy due to lymphoma. Axial contrast-enhanced CT **(a)** and MRI **(b)** scans show a filling defect *(arrowheads)* in the SV consistent with acute thrombosis. Note post splenectomy changes in the left upper quadrant *(curved arrow)*.

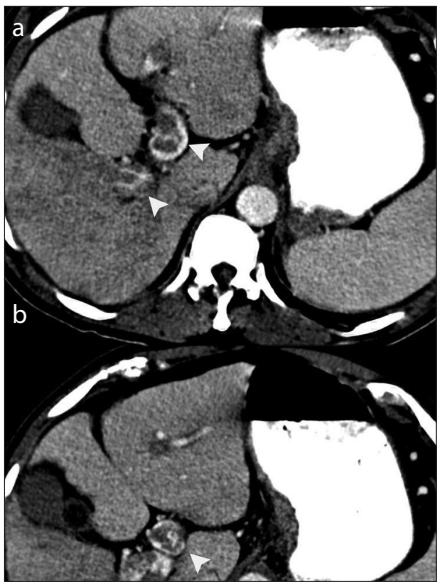


Figure 16. A 59-year-old man who presented with abdominal pain, weight loss, and anorexia. Axial contrast-enhanced CT scan **(a, b)** showed PV thrombosis involving the main PV and its right branch *(arrowheads)* in addition to heterogeneous enhancement in the right hepatic lobe. Fine needle aspiration biopsy revealed a hepatocellular carcinoma.

channels is essential to drain splenic and mesenteric venous flow into the intrahepatic portal branches and to sustain adequate hepatic blood flow and normal liver function. However, these small channels are usually inadequate to bypass the whole splenomesenteric inflow, and signs of portal hypertension subsequently develop.

Portal vein thrombosis

PV thrombosis is a rare condition in general population, but a relatively frequent entity in liver cirrhosis. Hypercoagulable states, intraabdominal infectious diseases (Fig. 14) such as pancreatitis, and surgery are the other most frequent causes of acute PV thrombosis. Doppler US is the single most useful imaging procedure for evaluation of PV thrombosis; however, it is expertise dependent, and some manifestations related with PV thrombosis such as varices and SMV thrombosis may be missed with Doppler US. In current clinical practice, dynamic contrast-enhanced CT scan is regarded as a precise and convenient diagnostic modality of PV thrombosis. Unenhanced CT scan may show venous enlargement and focal high attenuation in the PV during the acute stage. In later stages of the thrombosis linear areas of calcification can be seen within the thrombus. The correct discrimination between a bland thrombus and a tumor thrombus in patients with hepatocellular carcinoma may not always be possible with CT. Bland thrombus appears as a low attenuating, unenhancing filling defect that obstructs the lumen of the PV and its tributaries either partially or totally (Fig. 15). Hepatocellular carcinoma (Fig. 16) or, less commonly, metastatic liver disease (Fig. 17) may extend into the PV and cause tumor thrombus. MRI can help diagnose tumor thrombus and differentiate it from a bland thrombus. Tumor thrombus appears T1 hypointense and T2 hyperintense, whereas bland thrombus typically appears hypointense on both T1- and T2-weighted images. Besides, tumor thrombus enhances after contrast administration. Recently developed four-dimensional (4D) flow MRI achieves high spatial and temporal resolution in short scan times, which allows comprehensive assessment of portal venous system hemodynamics in a single breath hold. Visualization of PV hemodynamics with 4D flow MRI in a patient with suspected PV thrombosis shows not only a lack of flow in the PV, but also abnormal flow ways in other vessels (15, 16). The tumor thrombus may also be differentiated from bland thrombus using diffusion-weighted imaging (DWI); ratio of apparent diffusion coefficient (ADC) of the thrombus to ADC of the primary tumor lower than 2 and similar signal intensity of the thrombus as the primary tumor suggest a tumor thrombus (17). Since DWI does not require contrast use, it can be particularly useful in patients with contraindications to contrast agent. Intrahepatic cholangiocarcinoma (Fig. 18) tends to encase the PV rather than intraluminal tumor thrombus formation. Although spontaneous resolution of thrombosis can occur in some patients, partial or complete recanalization is seen more frequently in patients treated with anticoagulation therapy.

Gas within the portal venous system

There are a wide variety of causes for presence of gas within the portal venous

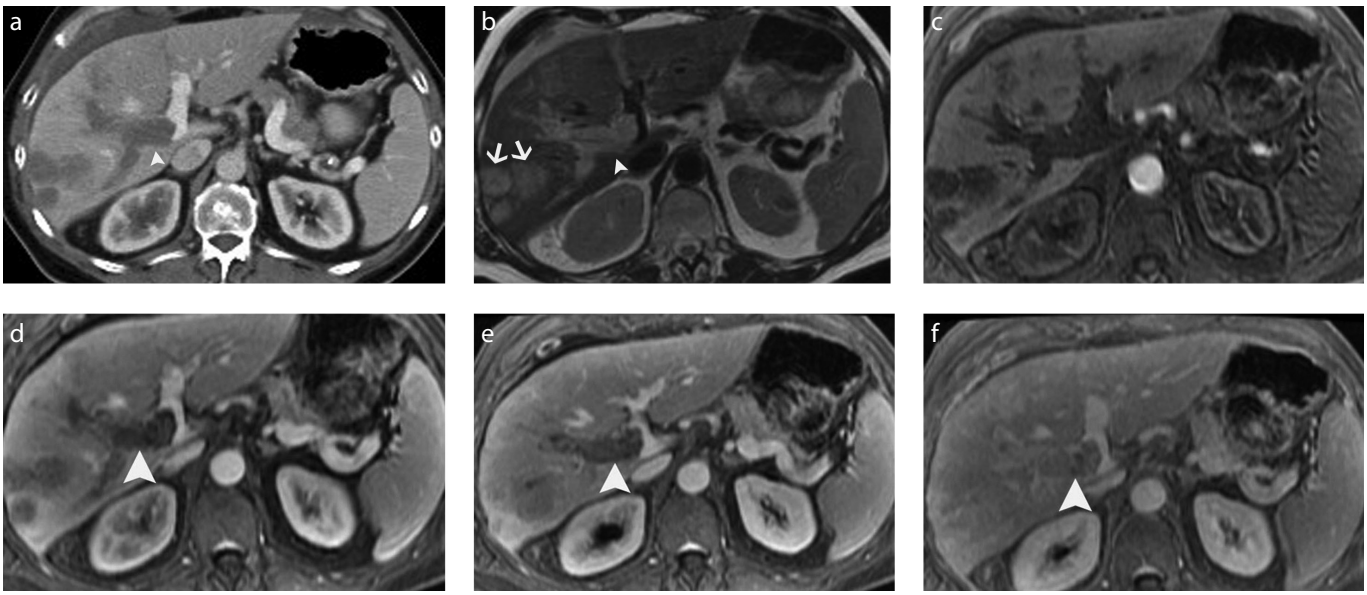


Figure 17. a–f. A 35-year-old man with metastatic gastrointestinal stromal tumor (GIST). Axial contrast-enhanced CT image **(a)** shows hypoattenuating filling defect *(arrowhead)* in the right PV, which could represent either tumor or bland thrombus. The patient was referred to MRI for further characterization. The filling defect *(arrowhead)* appeared hyperintense on T2-weighted **(b)** and hypointense on T1-weighted **(c)** images compared to liver parenchyma, similar to metastatic liver lesions *(arrows)*. Dynamic contrast-enhanced images **(d–f)** show that the thrombus *(arrowheads)* gradually and markedly enhances, consistent with intravenous tumor extension.

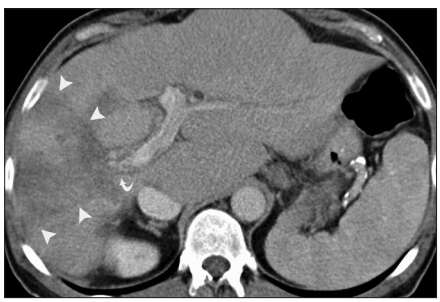


Figure 18. A 68-year-old man with a history of cholangiocarcinoma developed in the setting of chronic inflammatory bowel disease and sclerosing cholangitis. Axial contrast-enhanced CT scan shows a large hypoattenuating area *(arrowheads)* in the right hepatic lobe representing patient's known cholangiocarcinoma and encasement of the right PV *(curved arrow)*.

system. The primary factors that favor gas development are bowel ischemia and mesenteric vascular accident. PV gas resulting from bowel ischemia has been associated with a poor prognosis, with a mortality rate that ranges from 75% to 90% (18). However, advanced imaging modalities such as CT have increased the sensitivity for detection of portomesenteric vein gas and today it is not considered a grave sign as it was once assumed to be. Recent studies indicated mortality rates as low as 29% (19). On CT, air in the PV appears as branching streaks with air attenuation that can reach to the capsule of the liver (Fig. 19). Air in the PV has tendency to accumulate in the intrahepatic branch of the left PV vein due to its more vertical localization. Intrahepatic PV gas can be differentiated from pneumobilia because of its distribution. While gas in the biliary radicals is usually central, PV gas is typically peripheral and reaches the capsule of the liver.

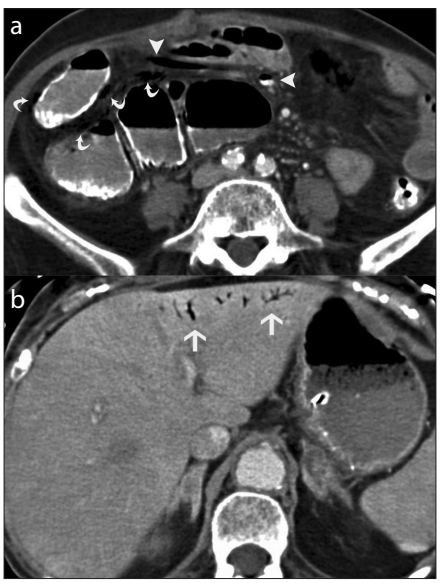


Figure 19. a, b. A 75-year-old woman with metastatic lung cancer who had undergone laparatomy due to small bowel obstruction, presenting with tender abdomen. Axial contrast-enhanced CT images show gas in the mesentery **(a)** *(curved arrows)*, SMV *(arrowheads)* and intrahepatic PV **(b)** *(arrows)* due to ischemic colitis.

Conclusion

A thorough information on CT and MRI features of anatomical variations and congenital and acquired abnormalities of the portal venous system permits the radiologists to identify these correctly; these features can play a significant role for patient management.

Conflict of interest disclosure

The authors declared no conflicts of interest.

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