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

Mustafa Özbayrak
Servet Tatlı

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).

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 perirectal 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
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-
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 arteriportal shunts. Arteriportal 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.
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 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 metastasis (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.
A 59-year-old female being treated with chemotherapy, consistent with metastatic liver disease. Numerous small venous channels (arrowheads) replace the main PV, a condition that typically develops secondary to chronic PV thrombosis. Gas within the portal venous system

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

Figure 19. a, b. A 75-year-old woman with metastatic lung cancer who had undergone laparotomy 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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