

Congenital and acquired abnormalities of the portal venous system: multidetector CT appearances

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ABSTRACT

A knowledge of normal anatomy as well as the most frequent variants and abnormalities of the portal venous system are of great importance for liver surgery and interventional procedures. An understanding of the varied MDCT appearances of these abnormalities will allow more definitive diagnoses and help avoid false diagnoses.

Key words: • portal vein, abnormalities • portal vein, thrombosis • portal vein, gas • shunts, arterioportal • tomography, X-ray computed

The portal venous system is associated with a wide range of congenital abnormalities and acquired processes that can be detected incidentally with current imaging modalities. In this pictorial essay, we discuss and illustrate the normal anatomy as well as the congenital and acquired abnormalities of the portal venous system as shown by multidetector computed tomography (MDCT). Recognizing the varied features of these abnormalities of the portal venous system will help radiologists to correctly interpret images and prevent misdiagnoses.

Normal anatomy

The portal vein issues from the confluence of the superior mesenteric, inferior mesenteric, and splenic veins posterior to the neck of the pancreas. In its most common branching pattern, the portal vein divides at the porta hepatis into the right and left portal veins. As it courses cranially, the right portal vein first sends branches to the caudate lobe and then divides into anterior and posterior branches. The left portal vein first follows a horizontal course to the left and then turns medially toward the ligamentum teres (umbilical portion), supplying the lateral and medial segments of the left lobe (Fig. 1). The Cantlie line corresponds to the median fissure and is defined as a line passing through the gallbladder toward the inferior vena cava (IVC) (1–3).

Congenital agenesis of the portal vein branches

Congenital agenesis of the portal vein branches is a frequently reported congenital anomaly (4). An important distinction exists between congenital agenesis and atrophy secondary to a pathological process. Congenital agenesis is thought to be secondary to either the failure of the right and left portal veins to develop or thrombosis of the affected lobe or segment during embryonic growth (Fig. 2). Liver ultrasonography is most often capable of identifying such vascular abnormalities (5). However, the MDCT appearances of these anomalies must be known because MDCT may be the initial form of imaging used.

Preduodenal portal vein

The preduodenal portal vein (PDPV) is associated with anomalies that include intestinal malrotation and pancreatic, splenic, or cardiac anomalies. The PDPV passes ventral to the duodenum and the head of the pancreas, so the PDPV is clearly seen on CT images as a round structure in front of the pancreatic head (2, 4, 6). An L-shaped and convex caudal mesentericoportal vein, which can also be seen on CT angiography, is an angiographic feature typical of PDPV (Fig. 3). MDCT images are reliable for revealing other associated anomalies, such as azygos or hemiazygos continuation of IVC, short pancreas, and visceral heterotaxia.

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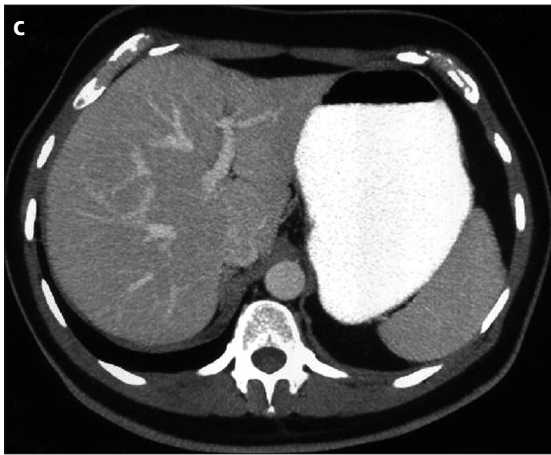
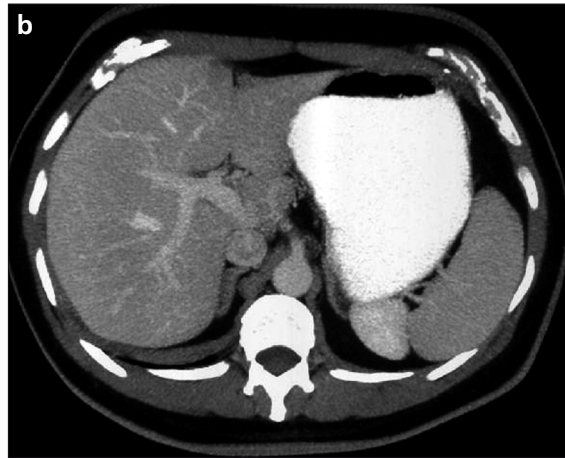
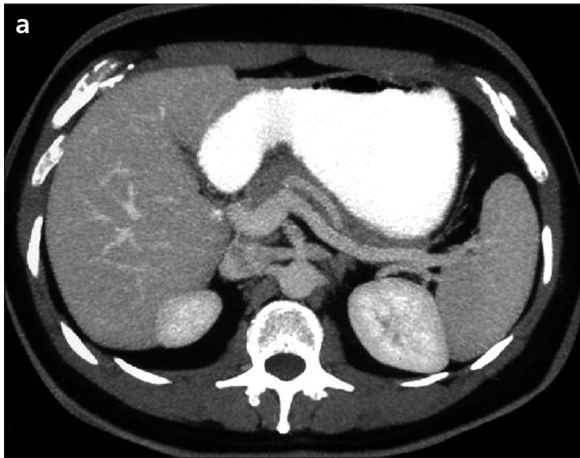


Figure 1. a–c. The normal anatomy of the portal vein in a 46-year-old man in an MDCT scan obtained during the portal venous phase. The axial image shows the main portal vein (MPV) formed by the union of the splenic and superior mesenteric veins behind the neck of the pancreas (a). The MPV courses to the porta hepatis, where it divides into the right and left branches (b). A section more cranial than b shows that the left portal vein passes transversely through the porta hepatis to supply the caudate and left lobes of the liver (c).

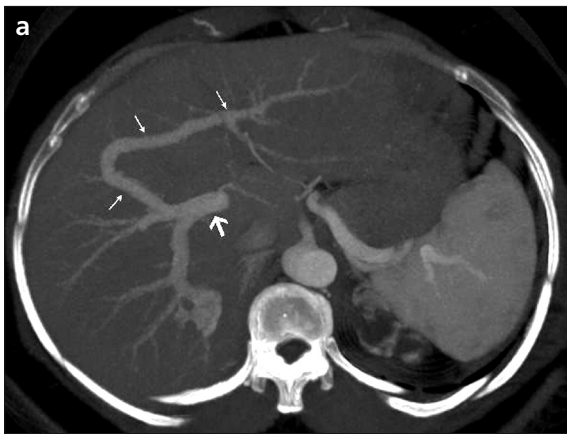


Figure 2. a, b. A 41-year-old woman with agenesis of the horizontal segment of the left portal vein. Two sequential axial thick-slab MIP CT images (a, b) reveal large aberrant vessels (thin arrows) running transversely from the right anterior portal branch (thick arrow) to the left segmental branches. Note the patency of the main portal vein (white arrowheads) and the splenic vein (black arrowheads).

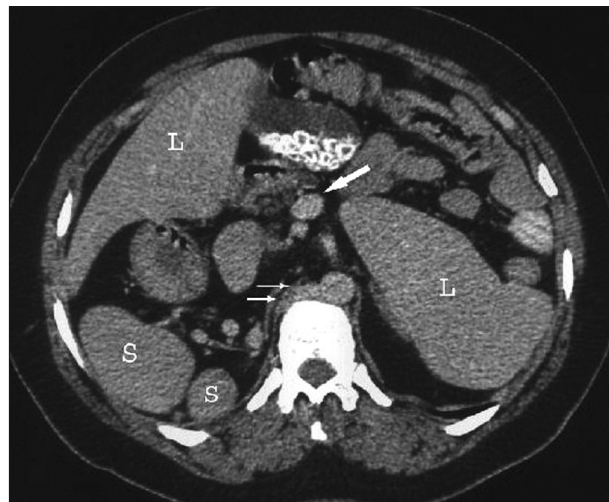


Figure 3. A 38-year-old woman with preduodenal portal vein and other congenital anomalies, including polysplenia (S), midline liver (L), and IVC absence with azygos continuation (double arrow). The axial image reveals preduodenal portal vein (arrow). Note gallbladder stones.

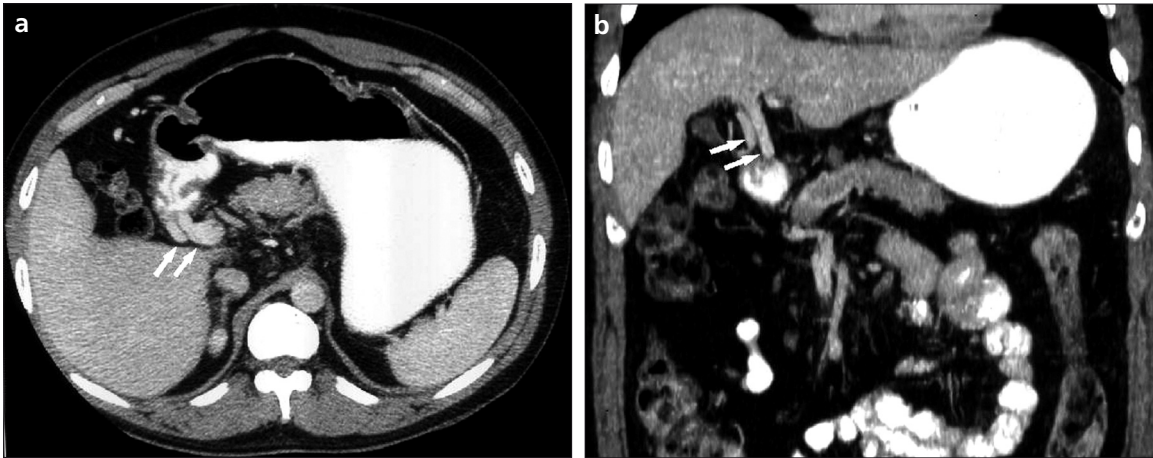


Figure 4. a, b. A 54-year-old man with a duplication of the portal vein. Axial (a) and coronal (b) MDCT images show two separate portal veins (arrows) coursing to the porta hepatis.



Figure 5. A 48-year-old woman with chronic liver disease and portal hypertension. Axial MDCT imaging reveals extrahepatic portal vein aneurysm (arrow). Note collaterals in the abdominal wall.

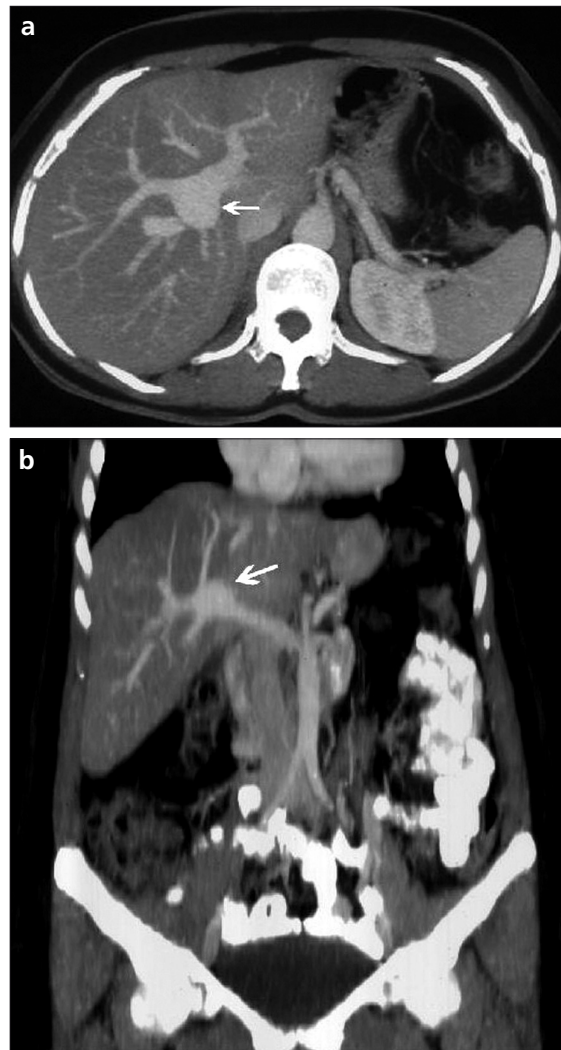


Figure 6. a, b. A 35-year-old woman with no underlying liver disease. Axial (a) and coronal (b) thick-slab MDCT images show the intrahepatic portal vein aneurysm (arrow).

Duplication of the portal vein

Duplication of the portal vein is an uncommon anomaly revealed by imaging as two separate portal veins that course upward to the porta hepatis (2, 7). Complications of this anomaly include variceal bleeding, portal hypertension and duodenal obstruction. MDCT allows multiplanar reconstructions and is helpful in distinguishing this anomaly from other disease entities (Fig. 4).

Portal vein aneurysm

Portal vein aneurysm, which can be congenital or acquired, is an uncommon entity. Portal venous system aneurysms represent 3% of all venous aneurysms (8). Most of these aneurysms are acquired, as a significant number of portal vein aneurysms are detected in patients with underlying hepatocellular disease and portal hy-

per-tension (Fig. 5). Some portal vein aneurysms are found in children and young adults who have no evidence

of liver disease or portal hypertension. Intrahepatic aneurysms have a tendency to occur at bifurcations (Fig. 6).

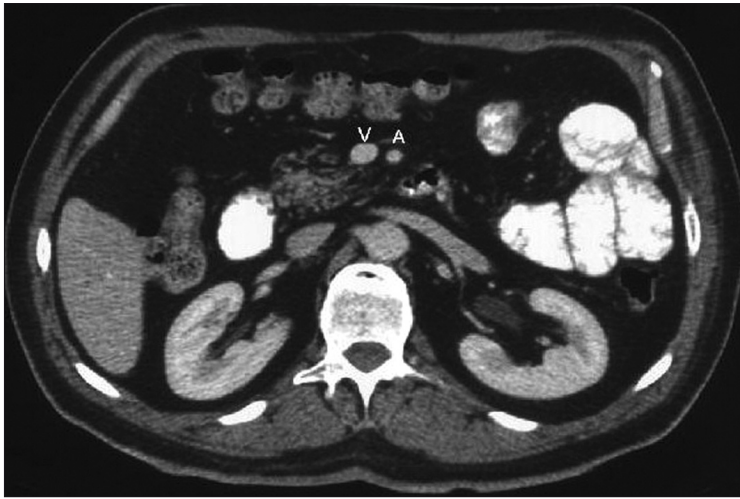


Figure 7. An axial MDCT image through the upper abdomen showing a normal relationship between the superior mesenteric artery (A) and the superior mesenteric vein (V).

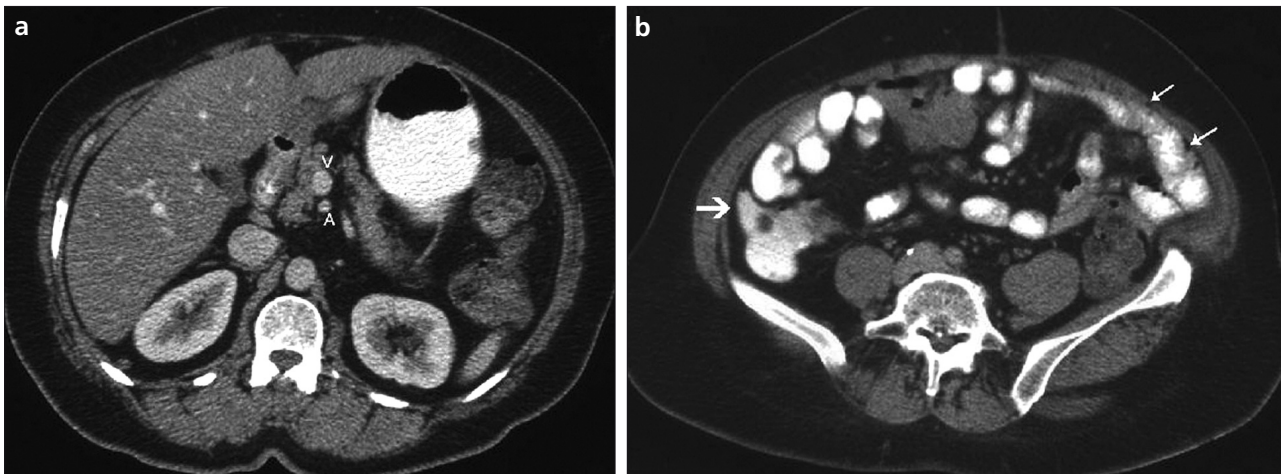


Figure 8. a, b. An axial MDCT image obtained through the upper abdomen (a) showing a vertical orientation of the superior mesenteric artery (A) and the superior mesenteric vein (V). Axial MDCT scan obtained through the lower abdomen (b) shows contrast agent-filled small bowel (*thin arrows*) on the left and colon (*thick arrow*) on the right.

Most reported extrahepatic portal vein aneurysms occur at the confluence of the superior mesenteric and splenic veins (1, 9). Most people with a portal venous system aneurysm are asymptomatic, although portal venous system aneurysms can cause symptoms. External compression and rupture are rare complications. Thrombosis of a portal venous system aneurysm occurs frequently and can lead to the development of portal hypertension with clinically severe consequences (10). Color Doppler sonography and CT have been considered accurate and reliable methods for both the diagnosis and follow-up imaging of portal venous system aneurysms and their complications.

Inverted relationship between the superior mesenteric vein and superior mesenteric artery

On cross-sectional imaging, the superior mesenteric vein (SMV) is generally seen lying on the right ventral aspect of the superior mesenteric artery (SMA) (Fig. 7). Conversely, deviation from the normal relationship between the SMV and SMA is a useful indicator of malrotation (Fig. 8). In most quiescent malrotation, the SMA and SMV will assume a vertical relationship or show left-right inversion. However, abnormalities of SMA-SMV orientation are not entirely diagnostic because some patients with malrotation will have a normal relationship, and a vertical or inverted relationship can also be seen in patients without malrotation (Fig. 9) (2, 11, 12).

Portal vein thrombosis

Portal vein thrombosis occurs in various clinical settings, with the most common being liver cirrhosis. In a number of cases, no identifiable cause for the portal vein thrombosis can be found. Thrombosis of the portal vein may partially or totally occlude its flow (4). Unenhanced CT may show focal high attenuation in the portal vein and venous enlargement when the thrombosis is acute. On contrast-enhanced CT, thrombi usually manifest as low-attenuation intraluminal lesions combined with the enlargement of the involved portal vein (Fig. 10). Chronic thrombosis can lead to the formation of multiple collateral channels and subsequent cavernous transformation of the portal vein (4). Calcification is occasionally seen in chronic thrombosis of the portal vein.

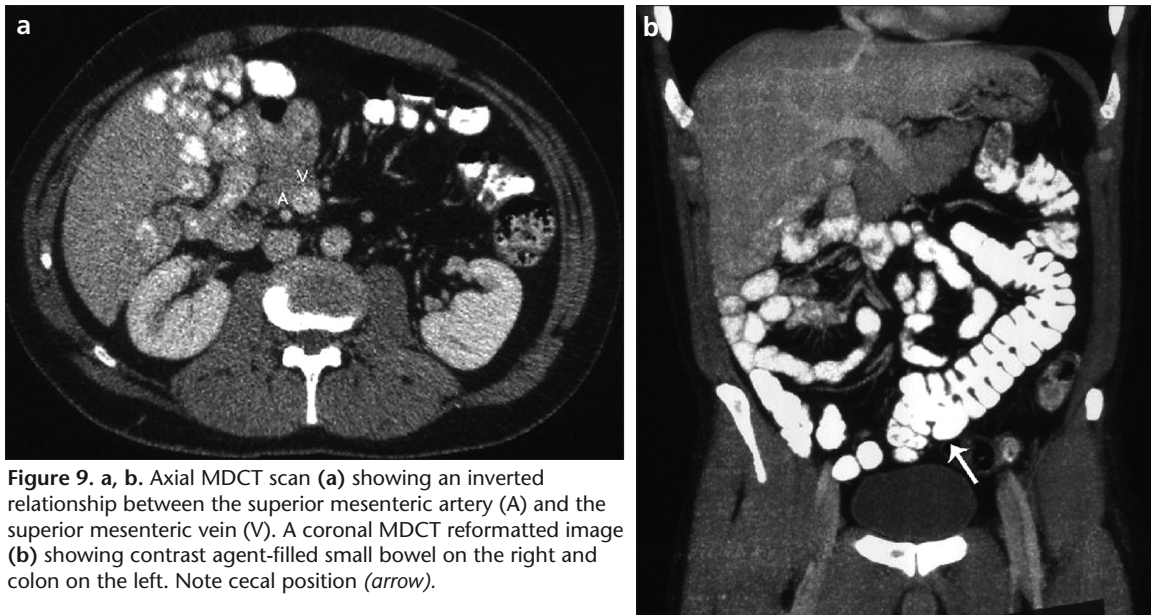


Figure 9. a, b. Axial MDCT scan (a) showing an inverted relationship between the superior mesenteric artery (A) and the superior mesenteric vein (V). A coronal MDCT reformatted image (b) showing contrast agent-filled small bowel on the right and colon on the left. Note cecal position (arrow).

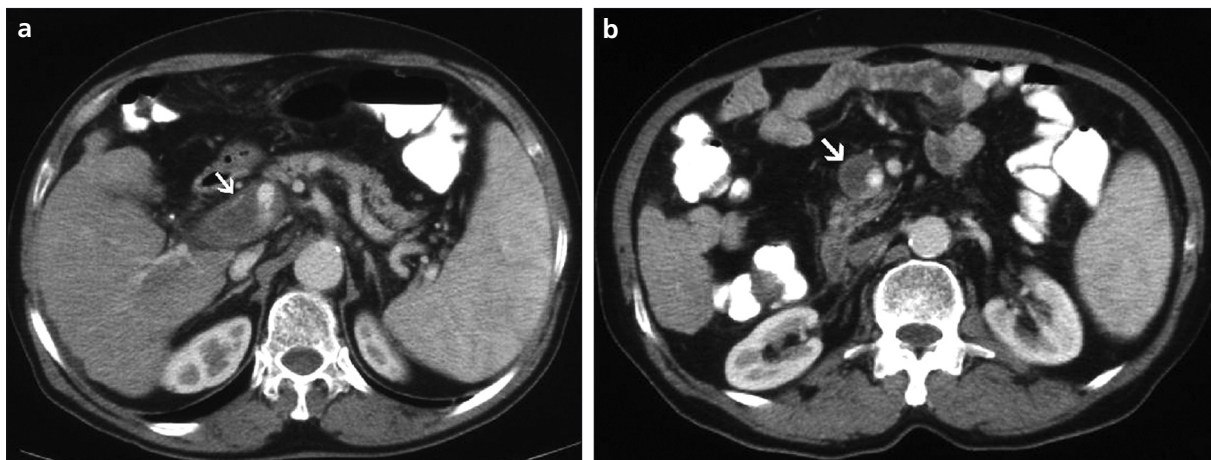


Figure 10. a, b. A 56-year-old woman with portal vein thrombosis. Axial MDCT scan demonstrates an enlargement of the portal vein and a low-attenuation intraluminal thrombus (a, arrow). A more caudal section (b) shows a thrombus in the superior mesenteric vein (small arrow). Note the nodularity of the liver surface due to cirrhosis.

Cavernous transformation of the portal vein

Cavernous transformation of the portal vein is defined as a masslike network of intertwined veins in the hepatoduodenal ligament and porta hepatis that provides an alternative pathway by replacing a stenosed or occluded portal vein (4, 13, 14). Cavernous transformation has been demonstrated by sonography as early as 6–20 days after the thrombotic event (Fig. 11).

Pseudothrombus

Pseudothrombus in the portal vein caused by laminar flow is often observed during an arterial-phase CT scan. This is a common flow-related artifact

resulting from the incomplete mixing of enhanced blood in splenic veins with unenhanced blood in superior mesenteric veins (13). Delayed-phase imaging will usually demonstrate homogenous portal venous enhancement (Fig. 12).

Arterioportal shunts

Arterioportal shunts may be congenital (vascular malformations in Osler-Weber-Rendu syndrome) or acquired (hepatic tumors, trauma, interventional procedures, cirrhosis) (Fig. 13) and consist of a communication between the hepatic artery and the portal venous system.

Dynamic CT performed during the hepatic arterial phase shows an ear-

ly and marked enhancement of the main portal vein, segmental branches or major tributaries with an attenuation approaching that of the aorta, as well as an early enhancement of the portal vein with nonenhancement of the splenic and mesenteric veins (1, 13).

Hepatic involvement is believed to be a common manifestation of hereditary hemorrhagic telangiectasia (15). Contrast-enhanced CT commonly shows a prominent hepatic artery. Dynamic study may also demonstrate the presence of an arterioportal shunt and telangiectases as a diffuse heterogeneous enhancement of the hepatic parenchyma (Fig. 14).



Figure 11. a–c. Portomesenteric venous thrombosis in a 43-year-old man with cirrhosis. Three sequential axial MDCT images show a cavernous transformation (**a**, *arrows*) at the porta hepatis, a large filling defect (**b**, *large arrow*) at the level of portovenous confluence with evidence of small-vessel collateralization (*small arrows*), and a thrombus in the SMV (**c**, *thick arrow*). Perihepatic fluid is present.

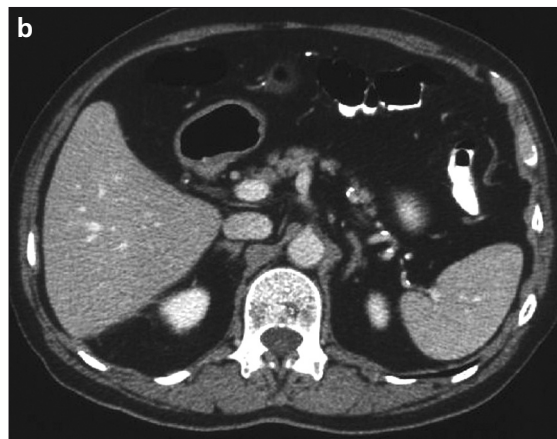
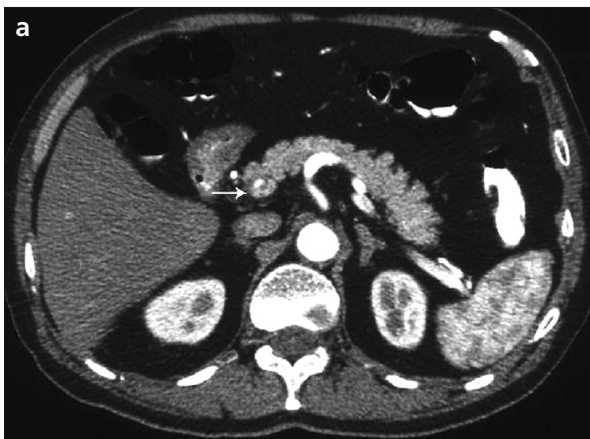


Figure 12. a–d. Pseudothrombosis of the main portal and the splenic veins. Axial MDCT scans obtained from the upper abdomen during the hepatic arterial phase (**a**, **c**) show a low-attenuation mass (*arrow*) mimicking portal vein thrombosis in the main portal vein and splenic vein. Portal venous phase CT scans (**b**, **d**) show normal enhancement of the portal venous confluence.



Figure 13. Neoplastic arterioportal fistula in a patient with hepatocellular carcinoma. The axial CT scan in the arterial phase shows an early enhancement of the portal vein and an increased perfusion of the hepatic lobes. The MDCT scan shows a tumor (*star*) invading the left and the right portal veins (*arrows*) and tumoral thrombus within the veins.

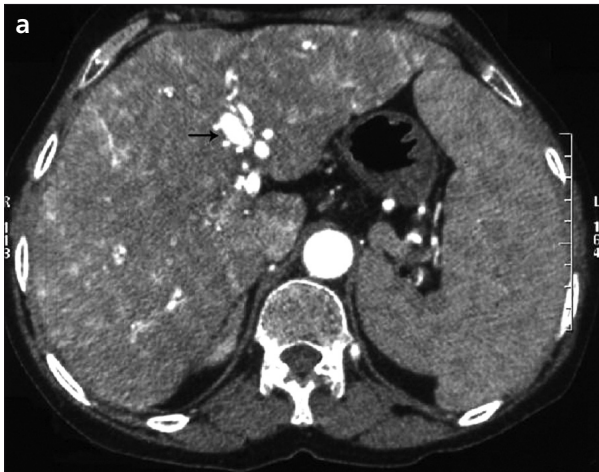


Figure 14. a, b. Axial arterial-phase MDCT images in a 46-year-old woman with hereditary hemorrhagic telangiectasia of liver. CT scan showing diffuse parenchymal heterogeneity and numerous telangiectases (*a*). Early filling of the left portal vein (*arrow*) during the arterial phase is consistent with an arterioportal shunt. Dilated common hepatic artery (*a, white arrow*) is seen.

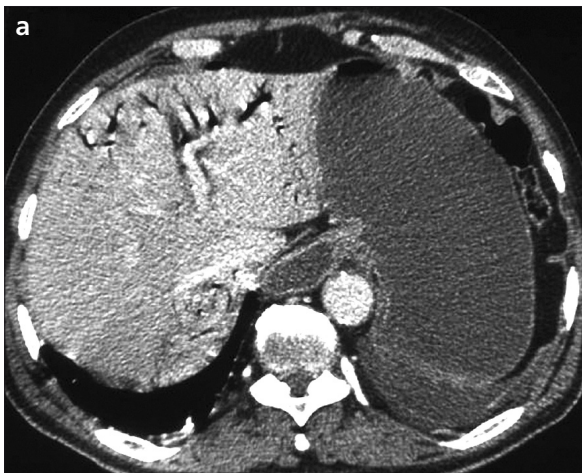


Figure 15. a, b. Intestinal pneumatosis in a 58-year-old man with progressive worsening of abdominal pain after aortobifemoral bypass grafting. Portal venous phase axial MDCT scan (*a*) obtained in a patient with mesenteric ischemia shows gas distributed in a branching pattern at the periphery of the liver. Coronal MDCT reformatted image (*b*) shows pneumatosis within the wall of the jejunum (*large arrow*) and ascending colon (*small arrow*).

Gas in the portal venous system

On CT scans, air in the portal vein manifests as ramifying streaks with air

attenuation that can reach the capsule at the periphery of the liver (1). Air has a propensity to accumulate in the

intrahepatic radicles of the left portal vein due to its more ventral location (Fig. 15).

References

1. Gallego C, Velasco M, Marcuello P, Tejedor D, De Campo L, Frieria A. Congenital and acquired anomalies of the portal venous system. *Radiographics* 2002; 22:141–159.
2. Ito K, Matsunaga N, Mitchell DG, et al. Imaging of congenital abnormalities of the portal venous system. *AJR Am J Roentgenol* 1997; 168:233–237.
3. Atasoy C, Ozyurek E. Prevalence and types of main and right portal vein branching variations on MDCT. *AJR Am J Roentgenol* 2006; 187:676–681.
4. Corness JA, McHugh K, Roebuck DJ, Taylor AM. The portal vein in children: radiological review of congenital anomalies and acquired abnormalities. *Pediatr Radiol* 2006; 36:87–96.
5. Chevallier P, Oddo F, Baldini E, Peten EP, Diaine B, Padovani B. Agenesis of the horizontal segment of the left portal vein demonstrated by magnetic resonance imaging including phase-contrast magnetic resonance venography. *Eur Radiol* 2000; 10:365–367.
6. Talus H, Roohipur R, Depaz H, Adu AK. Preduodenal portal vein causing duodenal obstruction in an adult. *J Am Coll Surg* 2006; 202:552–553.
7. Dighe M, Vaidya S. Case report. Duplication of the portal vein: a rare congenital anomaly. *Br J Radiol* 2009; 82:32–34.
8. Blasbalg R, Yamada RM, Tiferes DA. Extrahepatic portal vein aneurysms. *AJR Am J Roentgenol* 2000; 174:877.
9. Pickhardt PJ, Bhalla S. Intestinal malrotation in adolescents and adults: spectrum of clinical and imaging features. *AJR Am J Roentgenol* 2002; 179:1429–1435.
10. Koc Z, Oguzkurt L, Ulasan S. Portal venous system aneurysms: imaging, clinical findings, and a possible new etiologic factor. *AJR Am J Roentgenol* 2007; 189:1023–1030.
11. Clark P, Ruess L. Counterclockwise barber-pole sign on CT: SMA/SMV variance without midgut malrotation. *Pediatr Radiol* 2005; 35:1125–1127.
12. Ito K, Higuchi M, Kada T, et al. CT of acquired abnormalities of the portal venous system. *Radiographics* 1997; 17:897–917.
13. Agarwal A, Jain M. Multidetector CT portal venography in evaluation of portosystemic collateral vessels. *J Med Imaging Radiat Oncol* 2008; 52:4–9.
14. Ianora AA, Memeo M, Sabba C, Cirulli A, Rotondo A, Angelelli G. Hereditary hemorrhagic telangiectasia: Multi-detector row helical CT assessment of hepatic involvement. *Radiology* 2004; 230:250–259.
15. Duncan ND, Trotman H, Seepersaud M, Dundas SE, Thame M, Antoine M. Obstruction of the duodenum by a preduodenal portal vein in situs inversus. *West Indian Med J* 2007; 56:285–287.