Peliosis Hepatis: Spectrum of Imaging Findings

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OBJECTIVE. It is important to recognize the imaging characteristics of peliosis hepatis because peliotic lesions may mimic several different types of focal hepatic lesions

CONCLUSION. We illustrate the spectrum of imaging findings of peliosis hepatis, including sonography, CT, MR, and angiography.

Peliosis hepatis (also called hepatic peliosis) is a rare benign disorder causing sinusoidal dilatation and the presence of multiple blood-filled lacunar spaces within the liver [1]. "Peliosis" is a term derived from the Greek *pelios,* which means "dusky" or "purple," referring to the color of the liver parenchyma with peliosis. Similar blood-filled spaces may be seen in the spleen, lymph nodes, and other organs (including the bone marrow, lungs, pleura, kidneys, adrenal glands, stomach, and ileum) [2]. The size of the lesions may vary from 1 mm to several centimeters.

Cause and Pathogenesis

The cause of peliosis hepatis can be related to drugs (including anabolic steroids, oral contraceptives, corticosteroids, tamoxifen, diethylstilbestrol, azathioprine, 6-thioguanine, 6-mercaptopurine, and methotrexate); toxins (polyvinyl chloride, arsenic, and thorium oxide); chronic wasting diseases (e.g., tuberculosis, leprosy, and various malignancies, particularly hepatocellular carcinoma); and infection in AIDS (so-called bacillary peliosis caused by *Bartonella henselae* and *Bartonella quintana*). In addition, several other conditions are described as associated with peliosis hepatis, including sprue, diabetes mellitus, necrotizing vasculitis, and hematologic disorders. Moreover, peliosis hepatis may develop after renal or cardiac transplantation. In 20–50% of patients, no associated condition is identified.

The pathogenesis of peliosis remains poorly understood, with various investigators proposing that the primary event could be obstruction of hepatic outflow at the sinusoidal level, direct breakdown of sinusoidal borders, dilatation of the central vein of the hepatic lobule, or hepatocellular necrosis leading to cavity formation [1].

Pathology

The gross pathologic appearance of peliosis hepatis is that of multiple, irregularly shaped blood-filled hepatic cavities. The lesions typically involve the entire liver, but focal peliosis hepatis has been described. At microscopic examination, cystic dilated sinusoids filled with RBCs and bound by cords of liver cells can be seen (Fig. 1).

Pathologists originally classified peliotic lesions by the presence or absence of endothelium [1]. However, although the blood-filled cavities may have no endothelial lining, reendothelialization probably occurs rapidly. Thus, the continuity/rupture of the endothelial lining of the sinusoids is not a reliable criterion to define peliosis hepatis [2]. To distinguish peliosis hepatis from sinusoidal dilatation, lesions should show evidence of rupture of the reticulin fibers that support the hepatocytes and sinusoids [1]. This rupture may follow the intrinsic weakness of the fibers of the endothelial wall (peliosis hepatis of the phlebectatic type) or may be associated with focal hepatocyte necrosis (peliosis hepatis of the parenchymal type) [1].

Peliotic lesions in bacillary peliosis contain clumps of organisms (i.e., *B. henselae* or *B. quintana*) that stain with the Warthin-Starry technique [1]. Patients with bacillary peliosis often have peliosis of the spleen and lymph nodes and cutaneous angiomatous lesions.

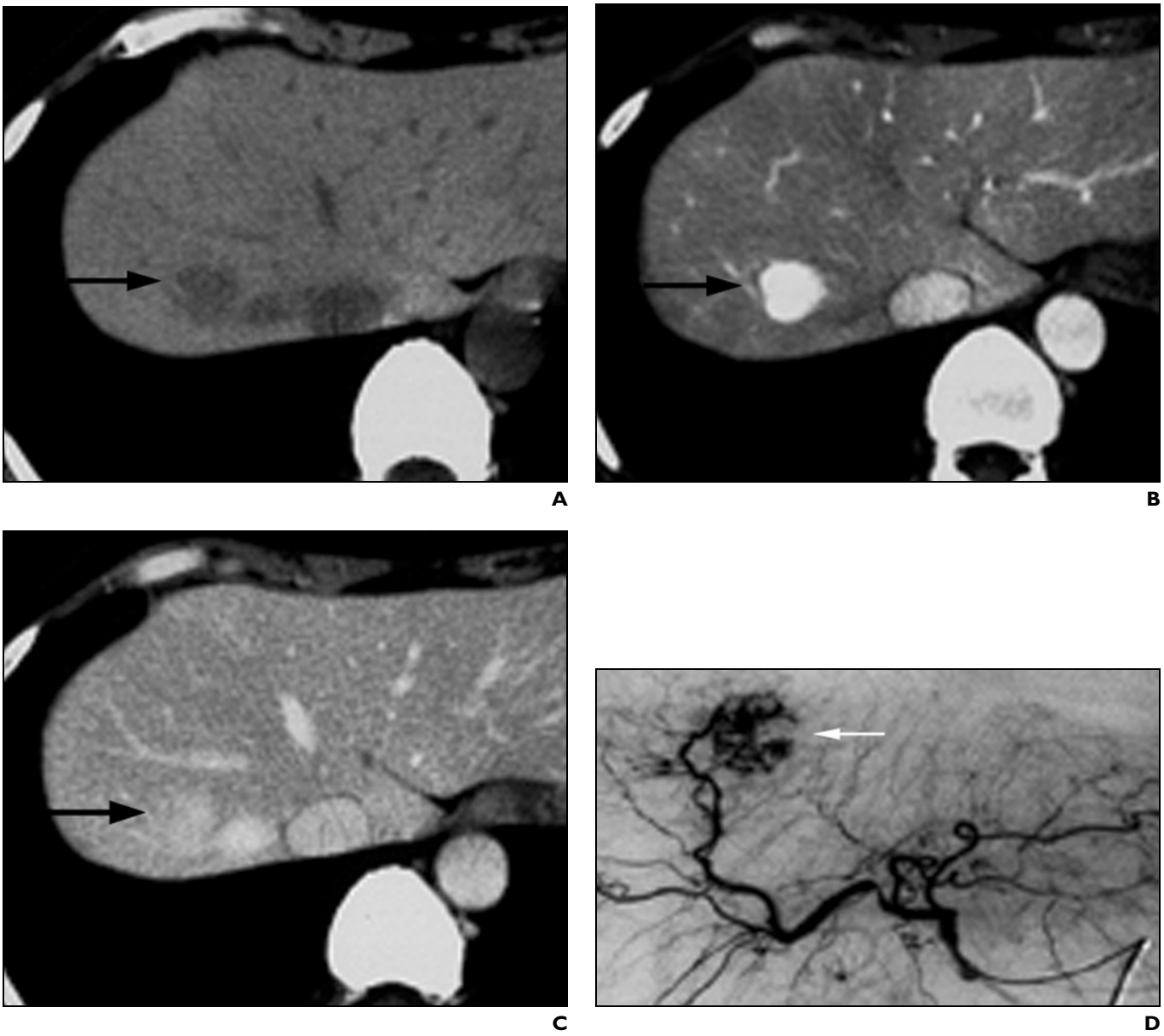


Fig. 1—51-year-old woman with history of benign ovarian tumor and incidentally discovered hepatic mass.
A, Transverse unenhanced CT image shows hypoattenuating lesion (*arrow*) within segment VIII of liver.
B, On contrast-enhanced CT during hepatic arterial phase, lesion (*arrow*) shows marked homogeneous contrast enhancement.
C, On contrast-enhanced CT during portal venous phase, lesion (*arrow*) shows washout of contrast but is still hyperattenuating compared with liver parenchyma. Overall, lesion has vessel-like enhancement at CT.
D, At angiography, lesion (*arrow*) is clearly depicted.
(Fig. 1 continues on next page)

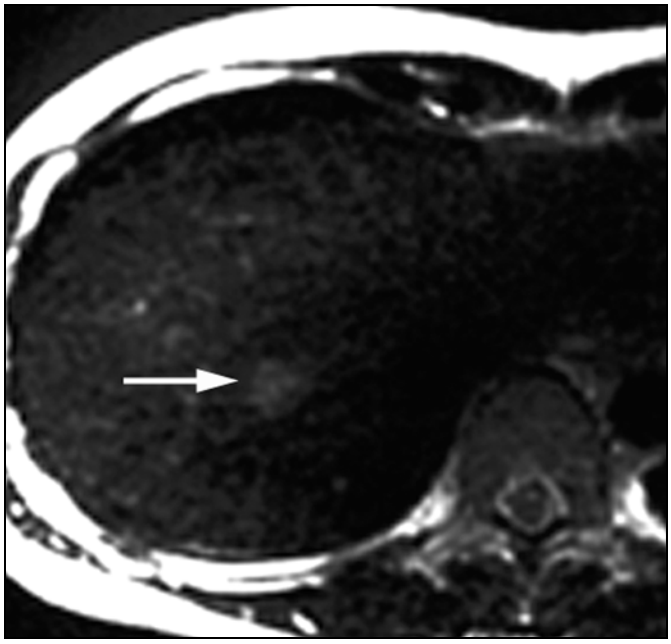
Clinical Presentation

Peliosis hepatis is often asymptomatic and therefore is diagnosed incidentally at autopsy [3]. In some instances, hepatomegaly, ascites, portal hypertension, cholestasis, and hepatic failure may be present. Severe abdominal pain may result from rupture and intraperitoneal hemorrhage. In the case of bacillary peliosis, lymphadenopathy with *B. henselae* and neurologic symptoms with *B. quintana* are typical findings.

Peliosis hepatis can occur at any age. Although a fetal form exists, peliosis hepatis usually develops in adults without regard to sex.

Natural History

The natural course of peliosis hepatis is regression after drug withdrawal, cessation of steroid therapy, or resolution of an associated infectious disease. A pseudotumoral and hemorrhagic evolution has also been described [4, 5]. Complications associated with



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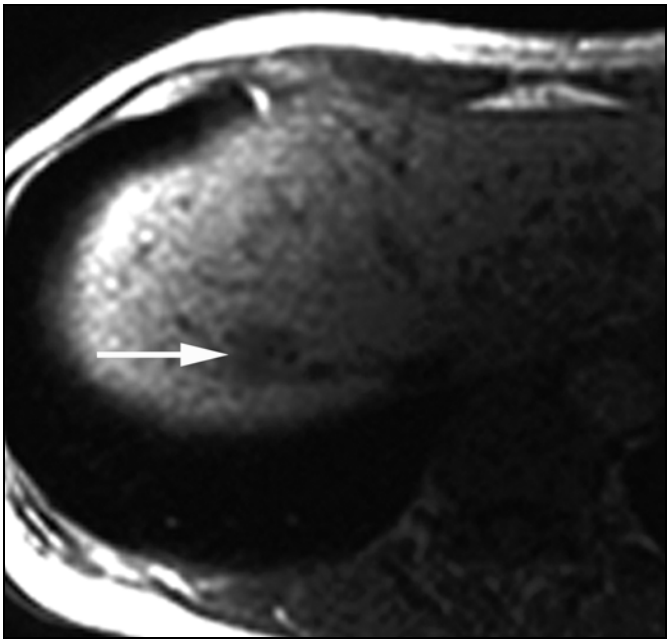

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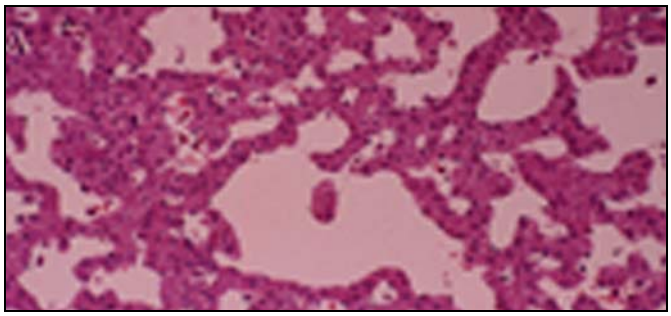

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Fig. 1 (continued)—51-year-old woman with history of benign ovarian tumor and incidentally discovered hepatic mass.
E, On T2-weighted MR sequence, lesion (*arrow*) is faintly hyperintense compared with liver parenchyma.
F, On T1-weighted MR sequence, lesion (*arrow*) is hypointense compared with liver parenchyma.
G, Multiple blood cysts are well shown at microscopic examination. (H and E, ×200)

peliosis hepatis include liver failure, portal hypertension, and liver rupture leading to hemoperitoneum or shock. In general, if untreated, peliosis hepatis may be rapidly fatal.

Imaging Findings

The imaging findings of peliosis hepatis are variable depending on the pathologic patterns of disease, various stages of the blood component of the lesions, and concomitant hepatic steatosis.

Sonographic Findings

Conventional gray-scale sonography shows homogeneous hypoechoic lesions in patients with hepatic steatosis, hyperechoic lesions in patients with a healthy liver (Fig. 2), and heterogeneously hypoechoic lesions if complicated by hemorrhage. Doppler studies can show evidence of both perinodular and intranodular vascularity (Fig. 2).

Recently, the use of a sonographic contrast agent (Levovist [Schering], a galactose and palmitic acid compound) has been shown to provide a "fast surge" central-echo enhancement in peliotic lesions [6], a finding consistent with the target sign described below.

CT Findings

On unenhanced CT, peliotic lesions usually appear as multiple areas of low attenuation [3] (Fig. 3). CT findings vary with the size of lesions, presence or absence of thrombus within the cavities, and presence of hemorrhage. In particular, peliotic lesions may be spontaneously hyperattenuating to liver parenchyma in certain patients (probably related to intralesional hemorrhage). In addition, if peliotic cavities are smaller than 1 cm in diameter, CT findings may be normal [2]. Calcifications within peliotic lesions have also been described.

On contrast-enhanced CT, peliotic lesions can be hypoattenuating to liver parenchyma in the early acquisitions (Fig. 4) and tend to become progressively isoattenuating with time. In addition, some lesions can also show areas of increased attenuation. Notably, larger cavities communicating with sinusoids display the same attenu-

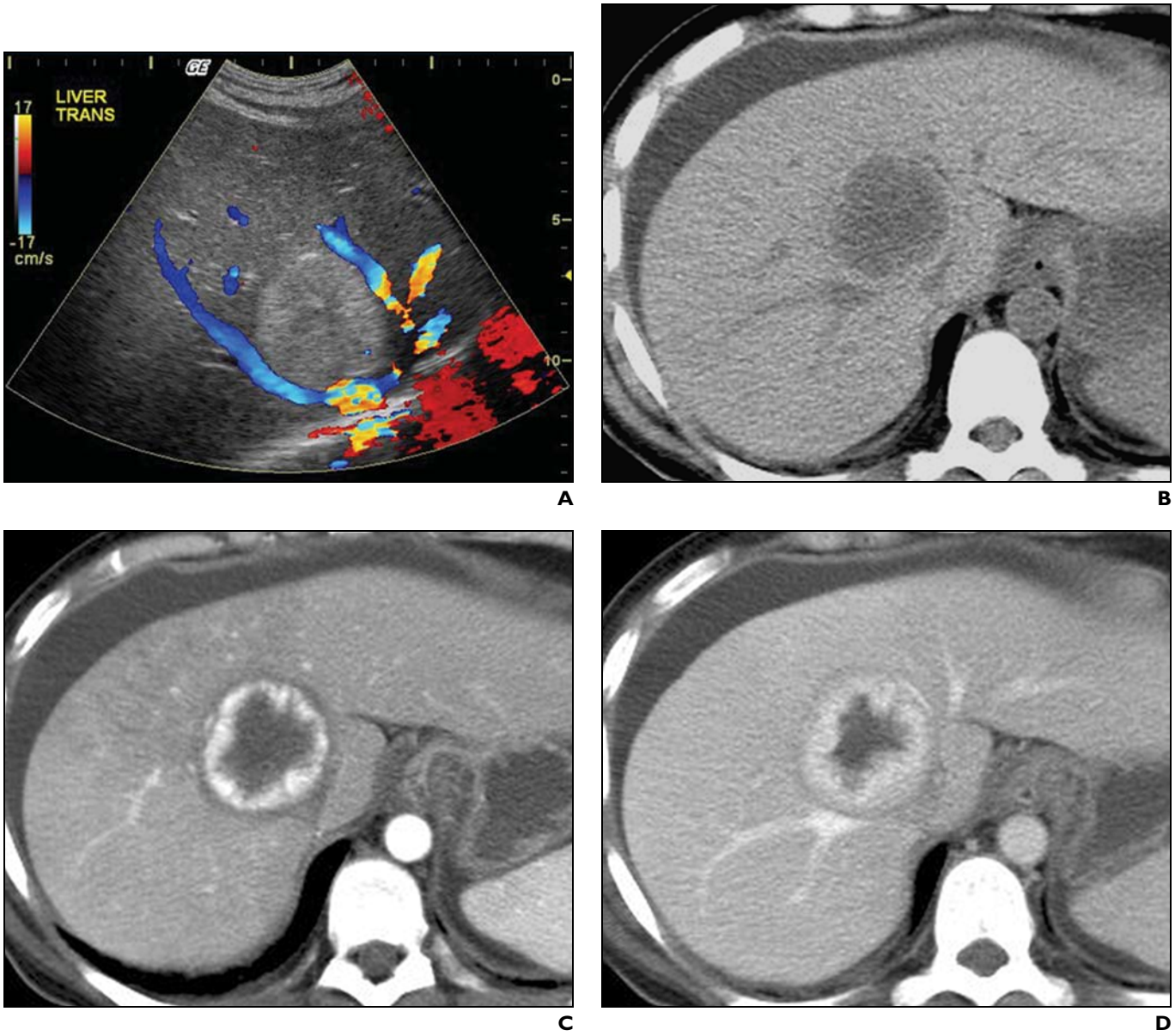


Fig. 2—42-year-old woman with peliosis hepatis.
A, Doppler sonographic study shows 4-cm diameter slightly heterogeneously hyperechoic lesion within healthy liver parenchyma. Notably, lesion has no mass effect on middle and right hepatic veins.
B, Transverse unenhanced CT image shows focal hypoattenuating lesion within right lobe of liver. Ascites is also evident.
C, In arterial phase of contrast enhancement, lesion shows marked peripheral ring enhancement. Lesion enhancement is isodense to aorta.
D, In portal venous phase, lesion enhancement is isodense to intrahepatic vessels. Note centripetal progression of contrast enhancement, which simulates hemangioma. Peliotic lesions have typically centrifugal progression of contrast enhancement, but centripetal enhancement can also be observed.

ation of blood vessels, whereas thrombosed cavities have the same appearance as nonenhancing nodules [2]. More often, during the arterial phase of contrast enhancement, peliotic lesions typically show early globular enhancement (vessel-like enhancement) (Figs. 2 and 5) and multiple small accumulations of contrast material in the center of the lesions (the so-called target sign) [3] (Fig. 3). During the portal venous phase, a centrifugal progression of enhancement without a mass effect on hepatic vessels is usually observed [3]; however, a centripetal progression of enhancement can also be seen [7] (Fig. 2). On

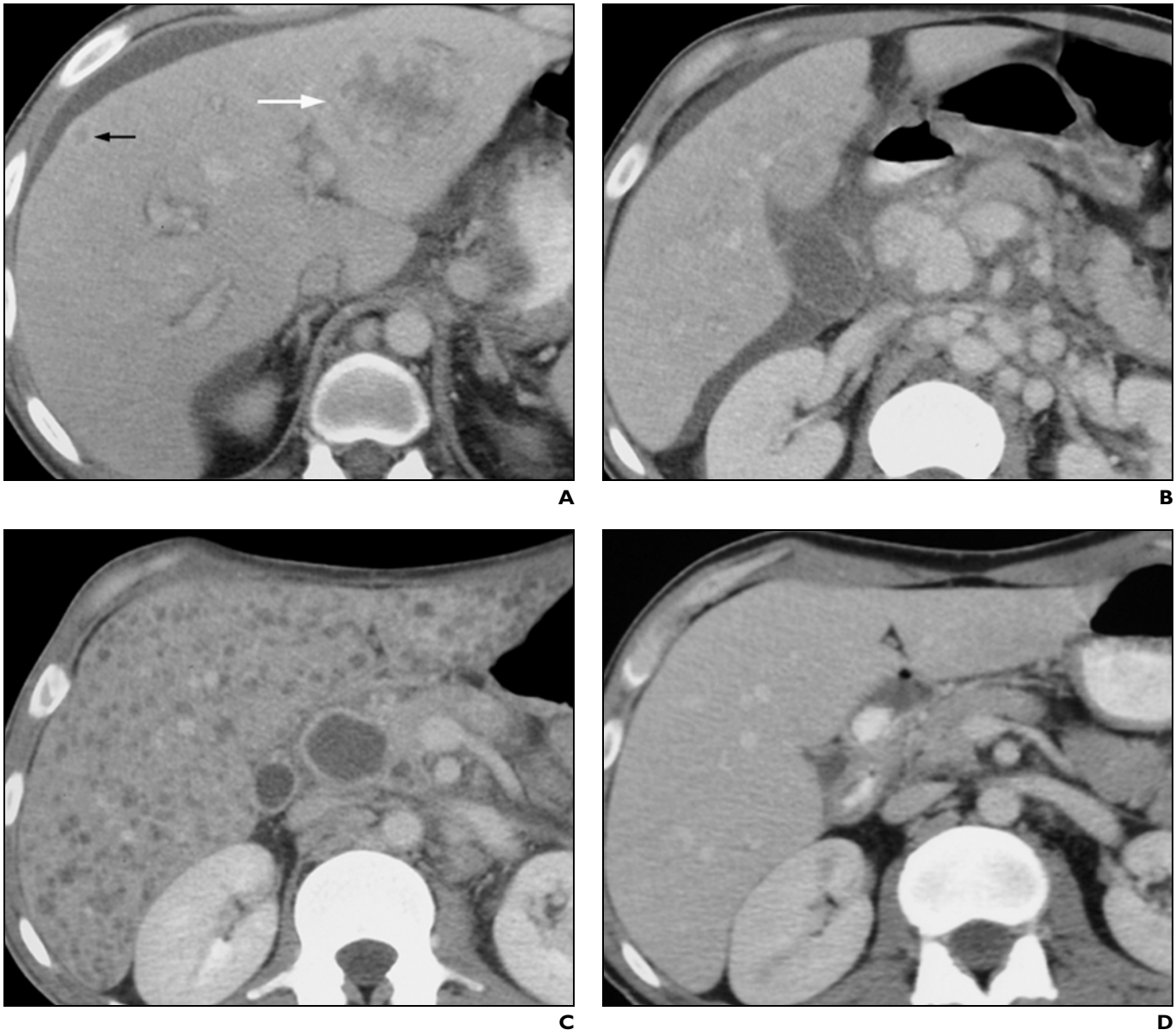


Fig. 3—34-year-old man with AIDS and bacillary peliosis.
A, Transverse contrast-enhanced CT image shows large ill-defined, hypoattenuating lesion (*white arrow*) with heterogeneous peripheral enhancement within left liver lobe. Smaller subcapsular hypoattenuating lesion (*black arrow*) with ring enhancement can also be seen in right liver lobe.
B, Transverse contrast-enhanced CT image (different scan level) shows multiple enlarged lymph nodes, feature typically seen in bacillary peliosis.
C, Transverse contrast-enhanced CT image obtained after 9 months shows progression of disease with multiple hypoattenuating lesions disseminated within liver parenchyma with multiple small accumulations of contrast material in center of lesions (so-called target sign).
D, After treatment with antibiotics, transverse contrast-enhanced CT image shows resolution of liver lesions and lymphadenopathy.

the delayed phase, late diffuse homogeneous hyperattenuation can also be seen in the phlebectatic type of peliosis hepatis (because of the lack of hemorrhagic parenchymal necrosis) [3]. This accumulation of contrast material in the delayed phase can be useful in the differential diagnosis with other focal hepatic lesions that do not show blood pooling. In some instances, small (< 2 cm) peliotic lesions may also show hyperattenuation on both arterial and portal venous phase images (Fig. 1).

MR Findings

The signal intensities of the lesions on MR examination largely depend on the age and status of the blood component. On T2-weighted sequences, peliotic lesions are usually hyperintense to liver parenchyma

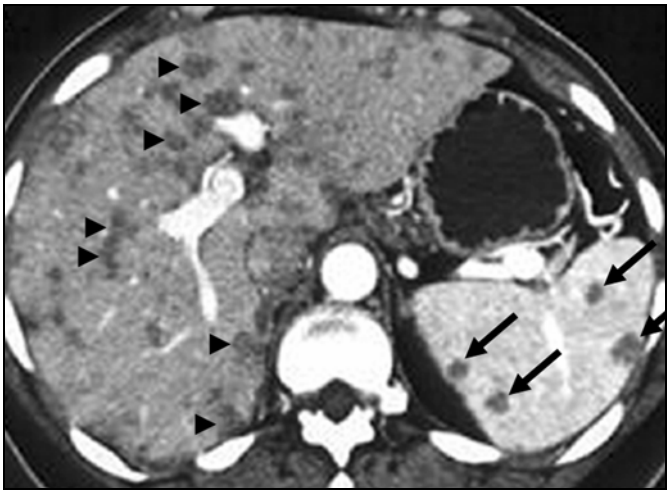
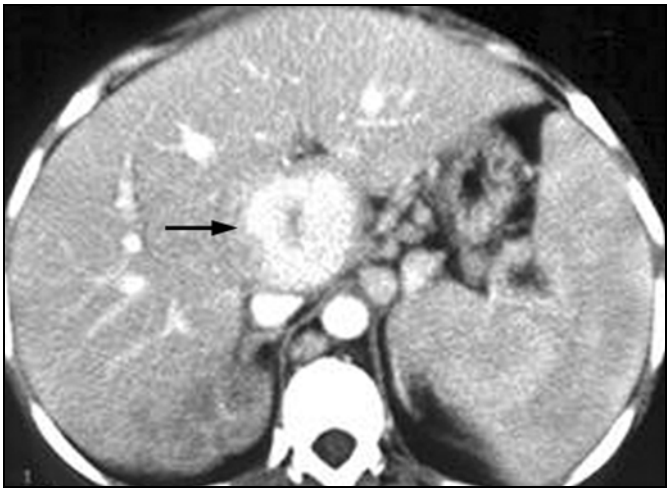


Fig. 4—46-year-old woman with history of abdominal pain and peliosis hepatis. Transverse contrast-enhanced CT image shows multiple hypoattenuating lesions (*arrowheads*) disseminated within liver parenchyma. Multiple similar lesions can also be seen within spleen (*arrows*).



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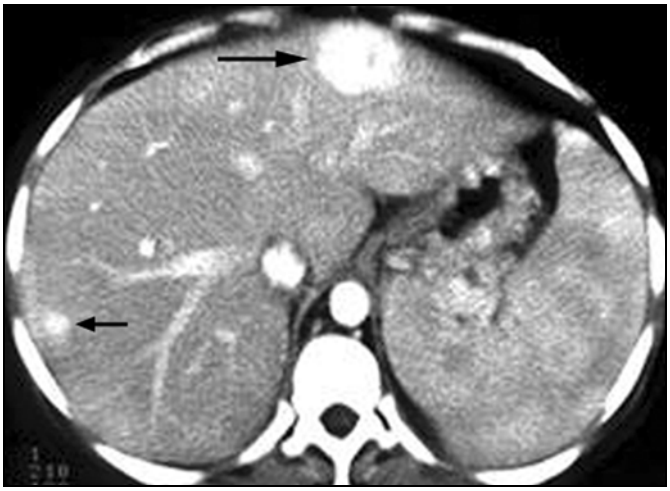

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Fig. 5—34-year-old woman with AIDS and bacillary peliosis. **A,** Transverse contrast-enhanced CT image shows focal hyperattenuating lesion (*arrow*) within caudate lobe. Lesion has central hypoattenuation. **B,** Transverse contrast-enhanced CT image (different scan level) shows two additional focal hyperattenuating lesions within left lobe (*large arrow*) and right lobe (*small arrow*) of liver.

with multiple foci of high signal, likely attributable to hemorrhagic necrosis [3, 4] (Figs. 1, 6, and 7). On T1-weighted sequences, the lesions are hypointense because of the presence of subacute blood (Figs. 1, 6, and 8), although isointense (Fig. 7) and hyperintense foci are also described in the literature [5]. On T1-weighted images after contrast material injection, peliotic lesions usually show enhancement (Figs. 6, 7, and 8). Similar to CT, the enhancement is typically centrifugal (from the center to the periphery of the lesion); however, a recent report described an unusual centripetal enhancement pattern that may be confused with that of a hemangioma [7]. Cystic cavities may reveal an enhancing rim that represents a hematoma. In addition, on fat-suppressed T1-weighted images in the delayed phase after administration of gadobenate dimeglumine, strong contrast enhancement with a branching appearance can also be observed because of the vascular component of the lesion [4] (Fig. 8).

Angiographic Findings

On angiography, peliotic lesions appear as multiple vascular nodules (i.e., accumulations of contrast material) during the late arterial phase. The enhancement of peliotic lesions typically is more distinct during the parenchymal phase and persists during the portal venous phase [8] (Fig. 1).

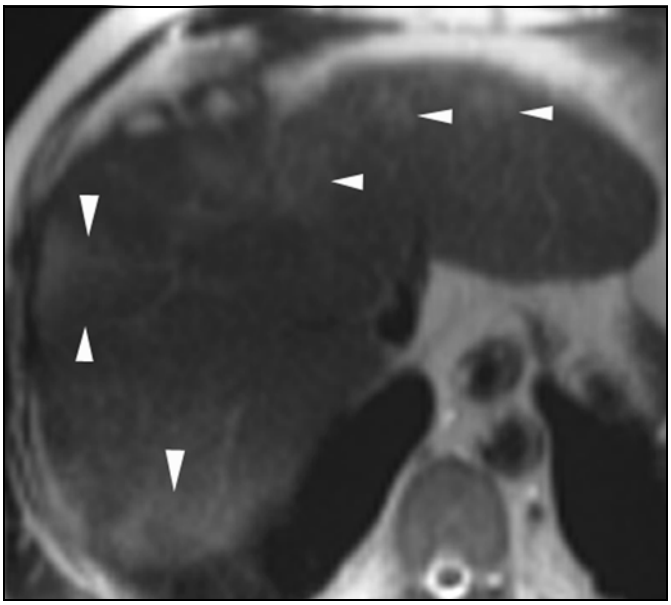
Differential Diagnosis

Hepatic Adenoma

Similar to peliosis, hepatic adenoma might also be associated with the long-term use of estrogens. In the case of diffuse peliosis hepatis, the differential diagnosis is relatively easy. In addition, the presence of fat in some adenomas is a useful sign to make a differential diagnosis. In certain instances, however, focal peliosis can be difficult to differentiate from adenomas. In these patients, biopsy is often required to reach a definitive diagnosis.

Hemangioma

The typical enhancement pattern of hemangiomas (i.e., peripheral ring or globular enhancement with centripetal progression) is opposite of peliosis hepatis, and therefore differential diagnosis can be achieved in most patients. In addition, hemangiomas may be rather large lesions with a mass effect on the hepatic vessels, whereas peliotic



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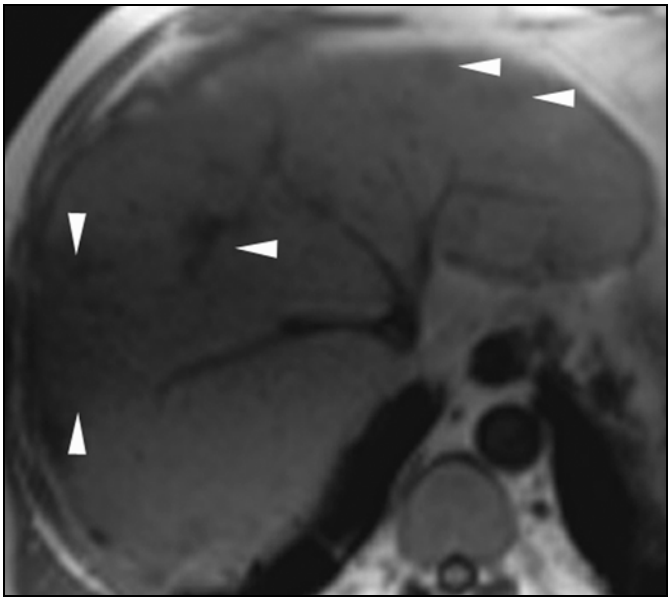

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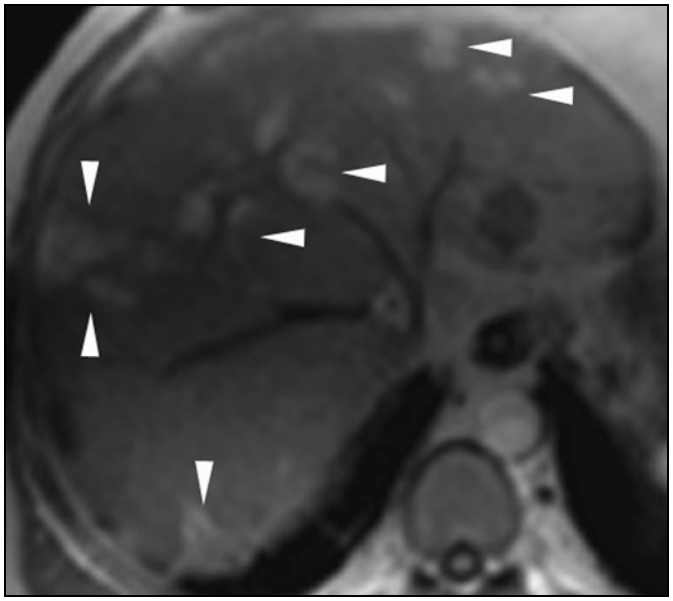

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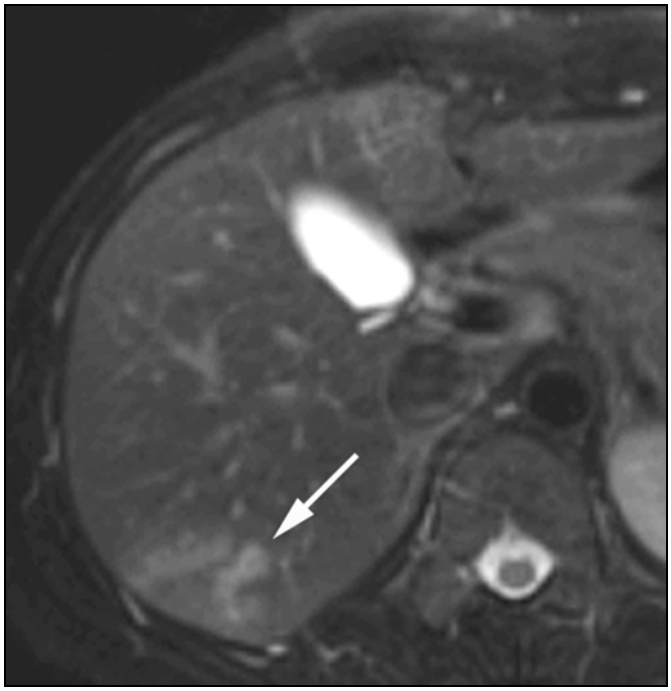
Fig. 6—57-year-old man affected by non-Hodgkin's lymphoma and peliosis hepatis caused by multiple chemotherapy treatments.
A, Transverse T2-weighted MR image faintly depicts multiple ill-defined lesions (*arrowheads*) with heterogeneous signal hyperintensity in both liver lobes.
B, Transverse T1-weighted MR image before contrast material injection faintly depicts multiple ill-defined, hypointense lesions (*arrowheads*) in both liver lobes.
C, On transverse T1-weighted MR image obtained after contrast material administration, lesions (*arrowheads*) show peripheral or complete contrast enhancement. Percutaneous biopsy (not shown) at two different sites showed peliosis hepatis. At follow-up after 1 year (not shown), lesions were still present but slightly smaller.

lesions usually show no mass effect on hepatic vessels.

Focal Nodular Hyperplasia

Focal nodular hyperplasias are typically homogeneously hyperattenuating masses on the arterial phase and isoattenuating on the portal venous and delayed phases. These lesions often have a central scar with low attenuation on the arterial and portal venous phases and enhancement on the delayed phase images. When such typical imaging characteristics of focal nodular hyperplasia are present, the differential diagnosis with peliosis hepatis can be achieved easily. Atypical forms of focal nodular hyperplasia may not show the characteristic enhancement patterns and the central scar just described, however, and thus pose some problems in the differential diagnosis with peliosis hepatis.



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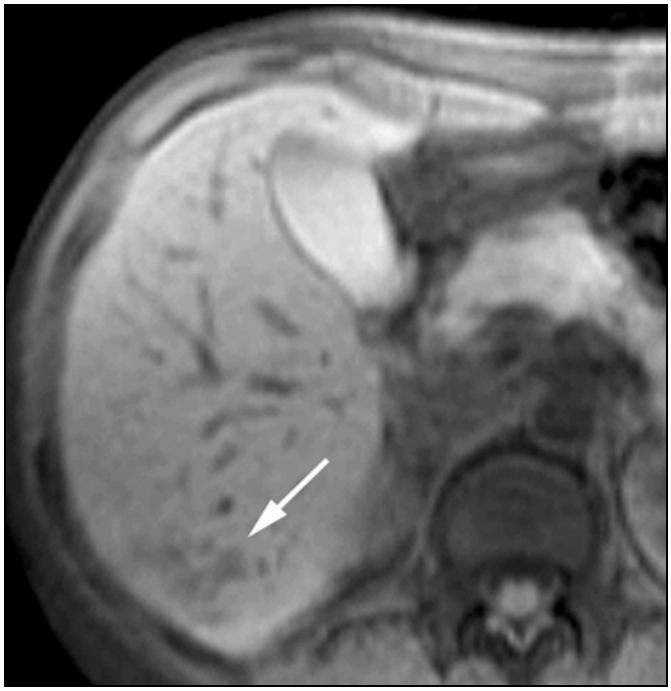

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Fig. 7—33-year-old woman with peliosis hepatis and history of oral contraceptive use.
A, Transverse fat-suppressed T2-weighted image shows ill-defined, heterogeneously hyperintense lesion (*arrow*) within segment VI of liver.
B, On transverse fat-suppressed T1-weighted image before contrast material injection, lesion (*arrow*) is heterogeneously hypointense to liver parenchyma.
(Fig. 7 continues on next page)

Hepatic Abscess

The differential diagnosis between peliosis hepatis and hepatic abscess is extremely important to avoid the percutaneous drainage of peliotic lesions, which can be dangerous and even fatal [9]. With regard to imaging criteria, a pyogenic abscess usually presents as a mass with a multiseptated or cluster-of-grapes appearance with nonenhancing contents.

Hypervascular Metastases

Although some hypervascular metastases with fibrotic change can show mild hyperattenuation in the delayed phase, hypervascular metastases are usually totally hypo- or isoattenuating in the delayed phase of contrast enhancement because of the rapid washout of contrast material. Thus, in general, peliotic lesions are rarely confused with hypervascular metastases.

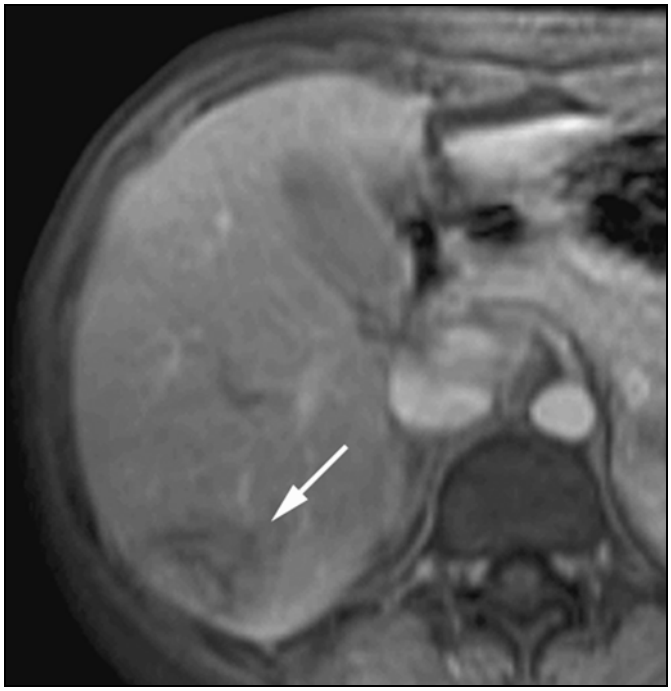
Hepatocellular Carcinoma

Hepatocellular carcinoma is usually hyperattenuating in the arterial phase with rapid washout in the portal venous phase and iso- or hypoattenuation in the delayed phase. Although rare, the possibility that peliosis hepatis may mimic the presence of hypervascular hepatocellular carcinoma has been reported in the literature. In these patients, biopsy is often necessary to reach a definitive diagnosis.

Treatment

The correct diagnosis of peliosis hepatis is important because withdrawal of the offending drug or toxin can resolve the disease and prevent serious complications such as hepatic failure or death related to intraabdominal hemorrhage. Because of its potential complications, surgical resection of the involved liver parenchyma should always be considered. In HIV-related peliosis hepatis caused by *B. henselae,* clinical improvement has been documented with the use of antibiotics (i.e., erythromycin).

In conclusion, peliosis hepatis is a rare clinical entity characterized by the presence of multiple blood-filled lacunar spaces within the liver. Awareness of the imaging findings of this disorder is important to suggest the diagnosis. Peliosis hepatis should always be considered in the differential diagnosis of atypical focal hepatic lesions in patients with the clinical conditions described earlier.



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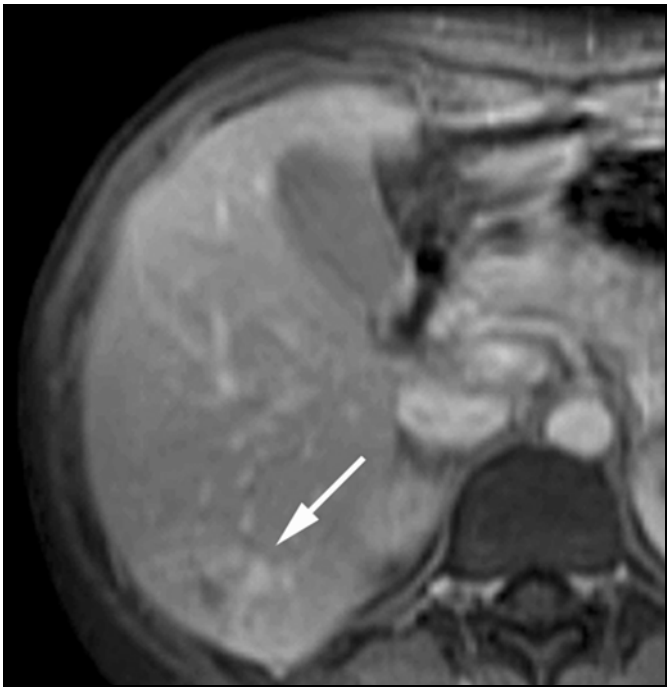

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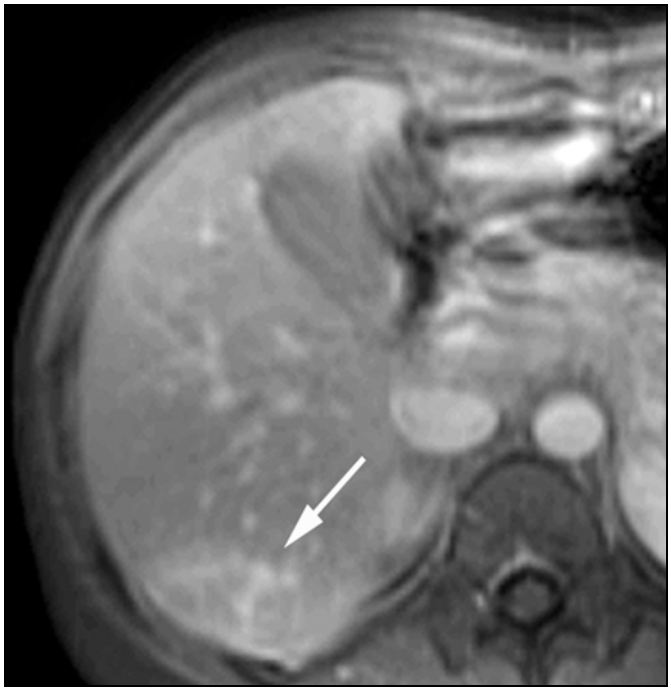

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Fig. 7 (continued)—33-year-old woman with peliosis hepatis and history of oral contraceptive use.
C–E, After gadolinium injection, lesion (*arrow*) shows progressive enhancement from arterial (**C**) to portal venous (**D**) and delayed (**E**) phases, with "branching" appearance. Because of progressive fill-in of contrast enhancement, findings were initially thought to be consistent with hemangioma. Percutaneous biopsy of lesion (not shown) was consistent with hepatocellular adenoma. However, histologic examination performed on resected surgical specimen showed peliosis hepatis. Centripetal enhancement in peliotic lesions is rather uncommon finding; branching appearance of contrast enhancement in delayed phase is useful sign that may help in diagnosis.

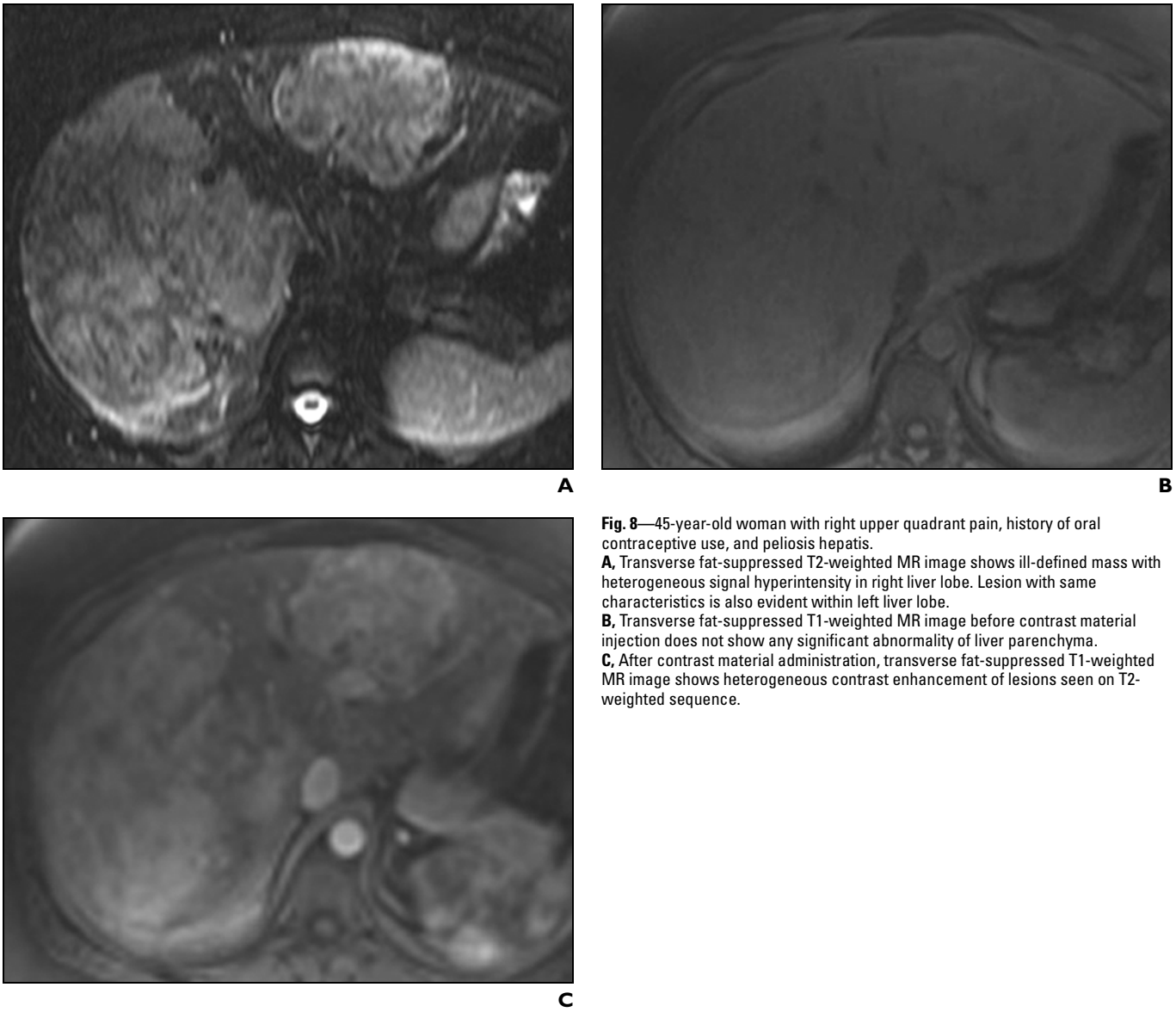

Fig. 8—45-year-old woman with right upper quadrant pain, history of oral contraceptive use, and peliosis hepatis.
A, Transverse fat-suppressed T2-weighted MR image shows ill-defined mass with heterogeneous signal hyperintensity in right liver lobe. Lesion with same characteristics is also evident within left liver lobe.
B, Transverse fat-suppressed T1-weighted MR image before contrast material injection does not show any significant abnormality of liver parenchyma.
C, After contrast material administration, transverse fat-suppressed T1-weighted MR image shows heterogeneous contrast enhancement of lesions seen on T2-weighted sequence.

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