

Hematologic Malignancies of the Liver : Spectrum of Disease

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- ◆ Hematologic malignancies include a wide spectrum of lymphoproliferative and myeloproliferative disorders with nodal and extranodal manifestations
- ◆ Hepatic involvement is a common extranodal manifestation of common and some rare hematologic malignancies

Primary and Secondary Hepatic Lymphoma

Posttransplant lymphoproliferative disorder

Myeloid sarcoma

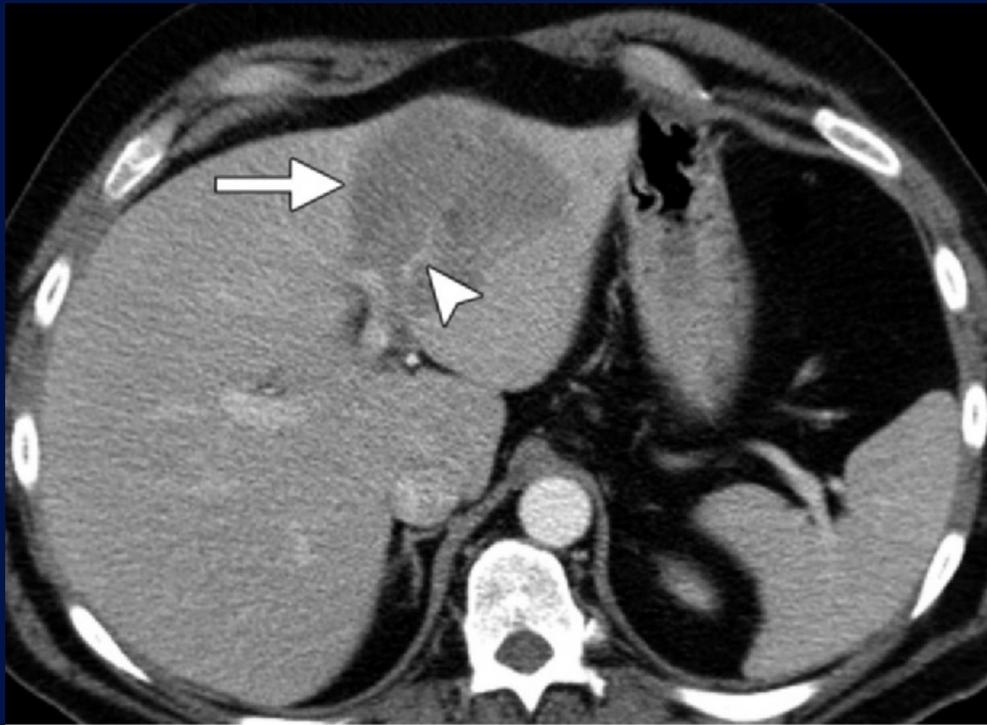
Multiple myeloma

Castleman disease

Hemophagocytic lymphohistiocytosis

- ◆ PHL is defined as lymphoma that is confined to the liver and perihepatic nodal sites at patient presentation, without distant involvement
- ◆ Right upper quadrant pain or jaundice, fever, weight loss
- ◆ Most cases of PHL are of B-cell lineage
- ◆ The most common imaging manifestation is a solitary discrete lesion, multiple lesions are seen in 35%–40% of patients
- ◆ Dominant liver masses are not typically seen in secondary lymphoma, but are characteristic of PHL. In contrast, multifocal lesions or diffuse infiltration is the most common pattern of secondary hepatic lymphoma
- ◆ Untreated nodules in secondary hepatic lymphoma are usually homogeneous, even when large, while the dominant masses in PHL are typically heterogeneously enhancing

- ◆ Numerous small discrete nodules (in a miliary pattern) are distributed throughout the liver in about 10% of cases of Hodgkin disease and secondary non-Hodgkin lymphoma of the liver
- ◆ Splenic lesions are not seen in patients with PHL but are seen in 30%–40% of patients with secondary non-Hodgkin lymphoma
- ◆ At CT, lymphomatous nodules commonly have enhance to a lesser degree than the liver parenchyma
- ◆ At MR imaging, the nodules tend to be hypo-or isointense on T1-weighted images and moderately hyperintense on T2-weighted images, with an enhancement pattern similar to that seen at CT



a.

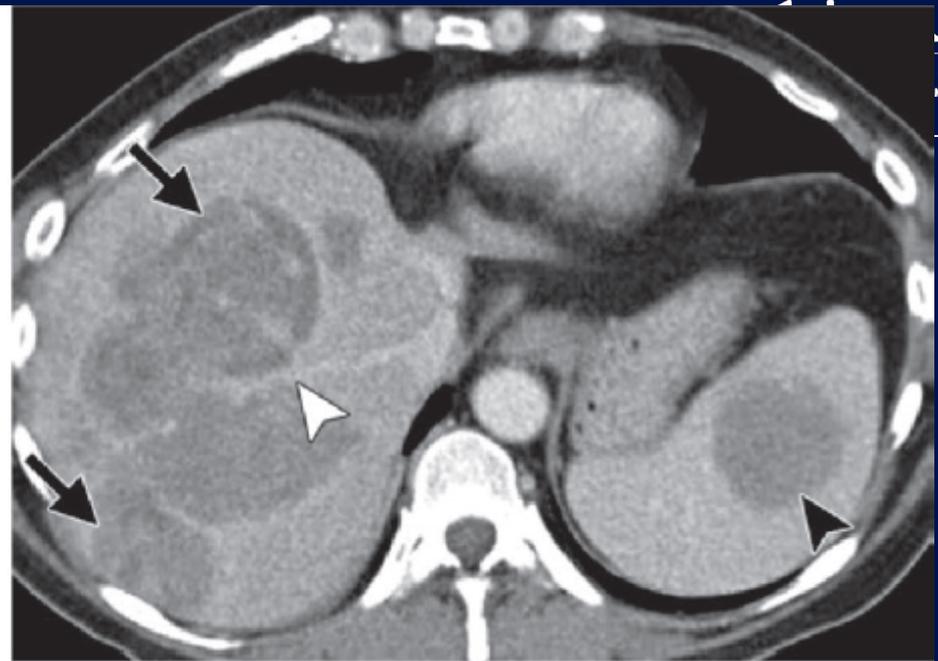


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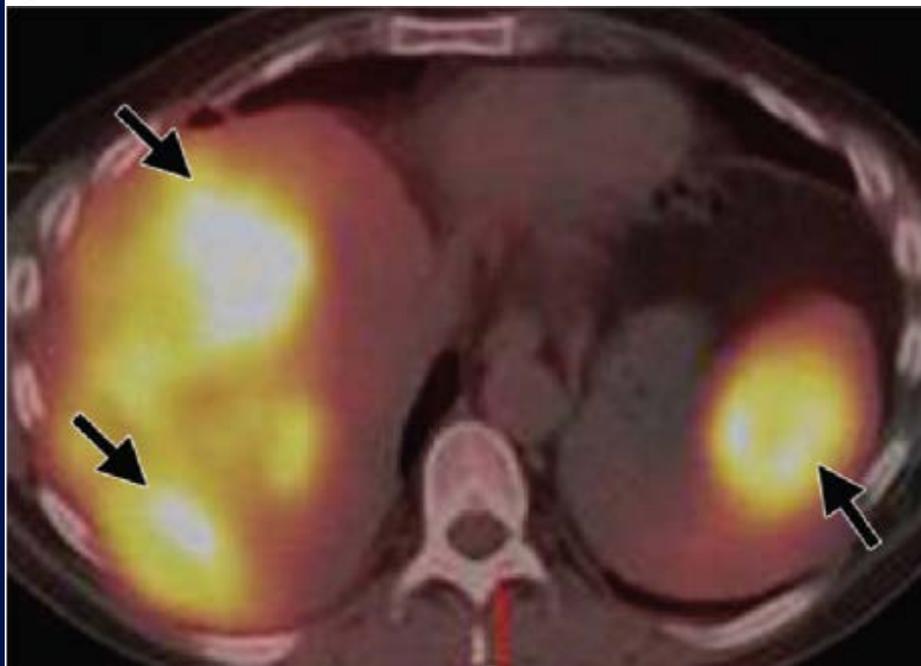
PHL in a 51-year-old man with night sweats. Axial CT images show a solitary mass in the left lobe of the liver (arrow), with a patent vessel (arrowhead in a) seen coursing through the mass. The spleen is uninvolved. Periportal nodes are seen (arrowheads in b), but there is no other abdominal adenopathy. Lymphoma was suspected and was proven at biopsy



2.

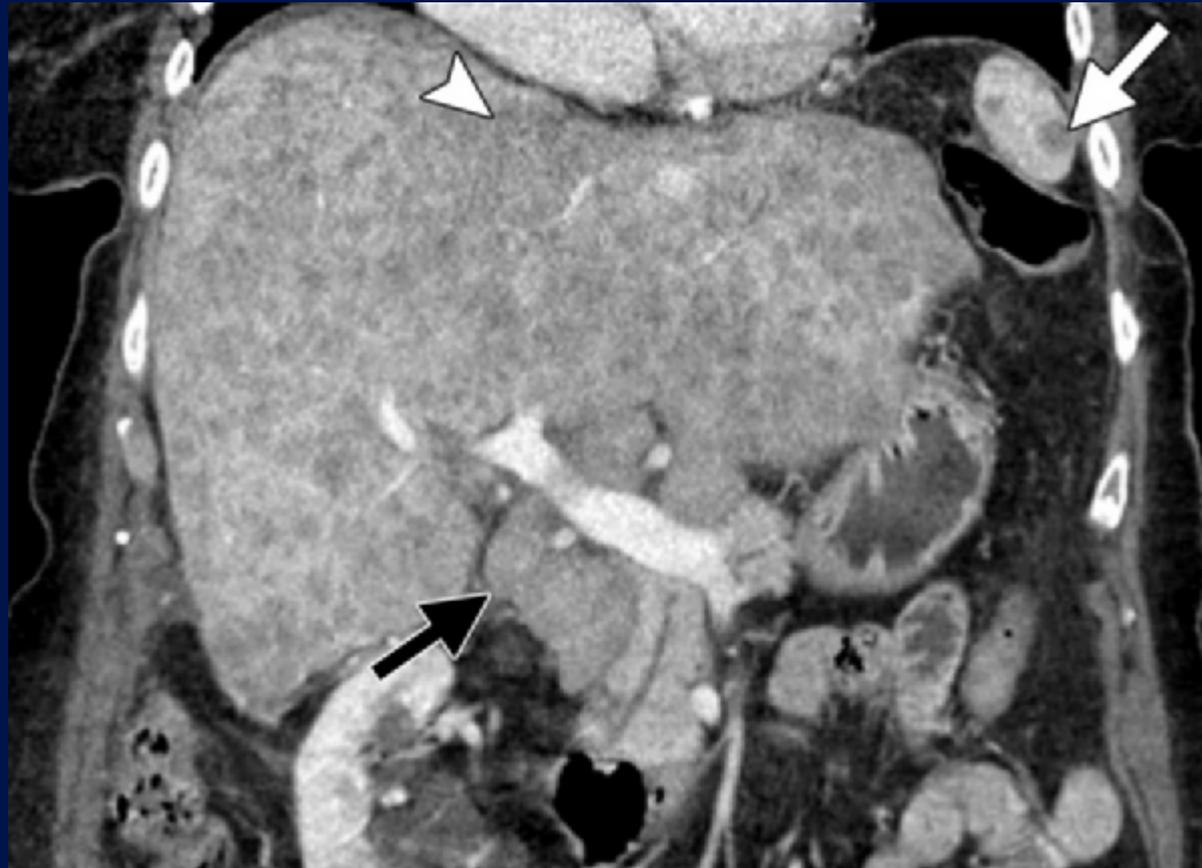


3a.

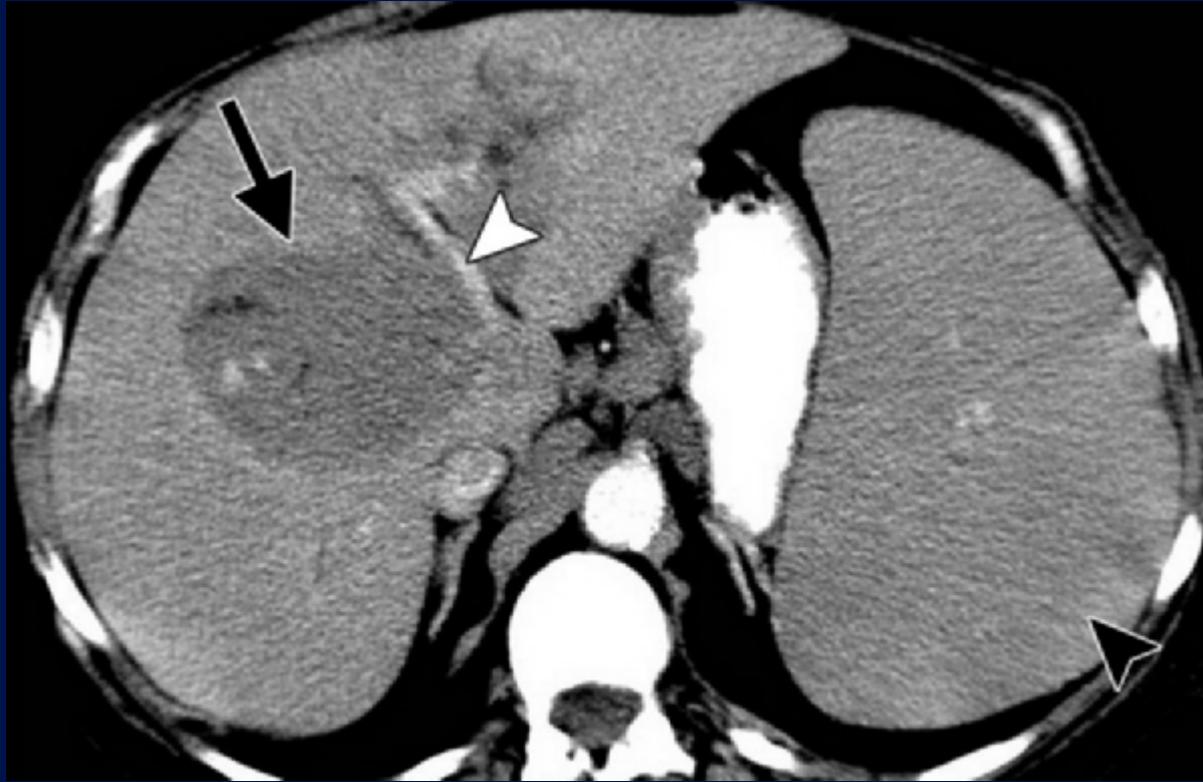


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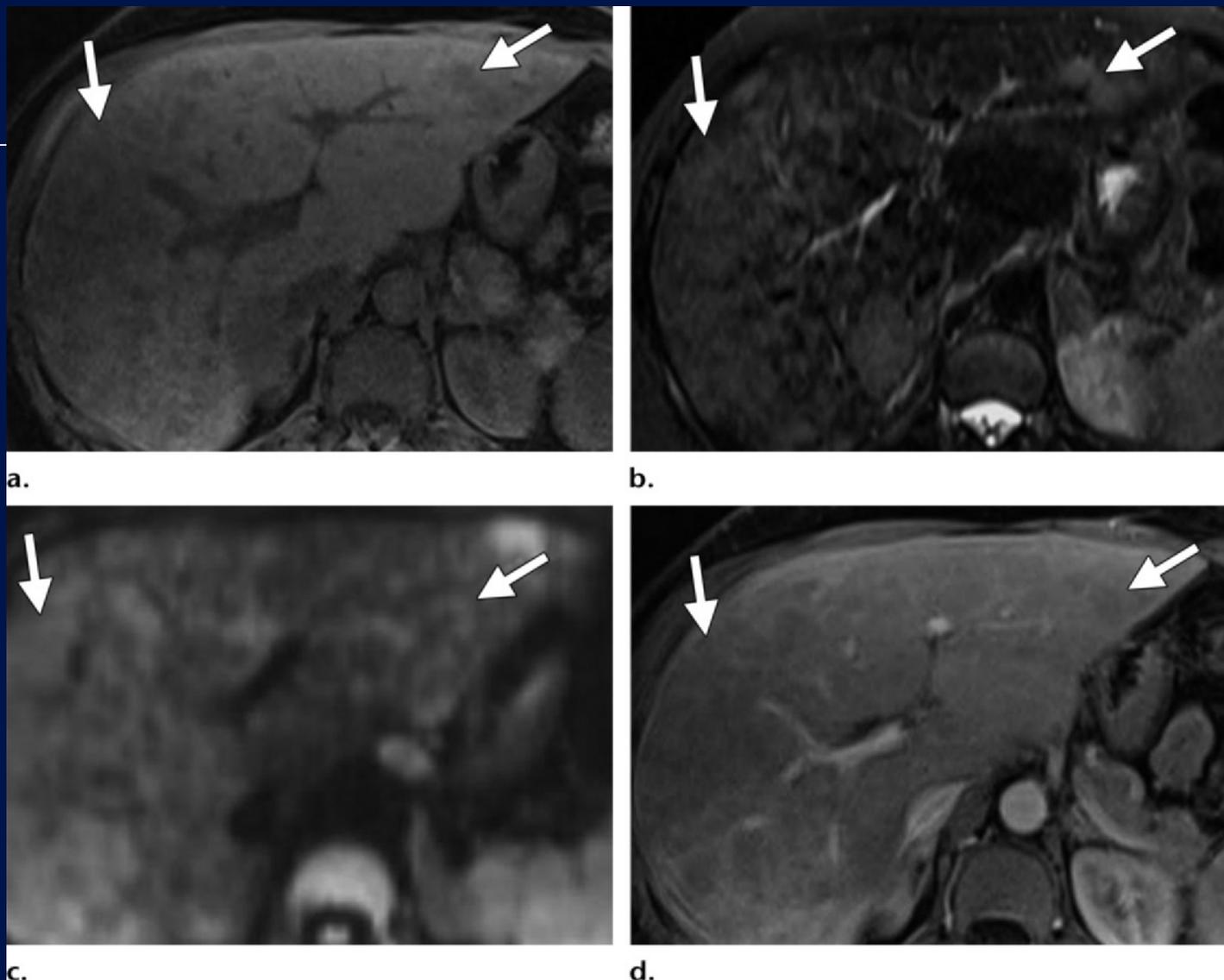
PHL in a 57-year-old man with fever and night sweats. Axial CT image shows two contiguous lesions (arrow and arrowhead) in the liver, with one lesion being dominant (arrow). Biopsy demonstrated lymphoma. No evidence of lymphoma outside the liver was seen at bone marrow biopsy or PET. (3) Secondary hepatic large B-cell non-Hodgkin lymphoma in an 83-year-old woman who presented for disease restaging. (a) Axial contrast-enhanced CT image shows multiple discrete homogeneously hypoenhancing hepatic masses (arrows) and a splenic mass (black arrowhead). The right hepatic vein (white arrowhead) is seen coursing through the mass without occlusion or constriction. (b) Axial fluorodeoxyglucose (FDG) PET/CT image shows avidly hypermetabolic lesions.



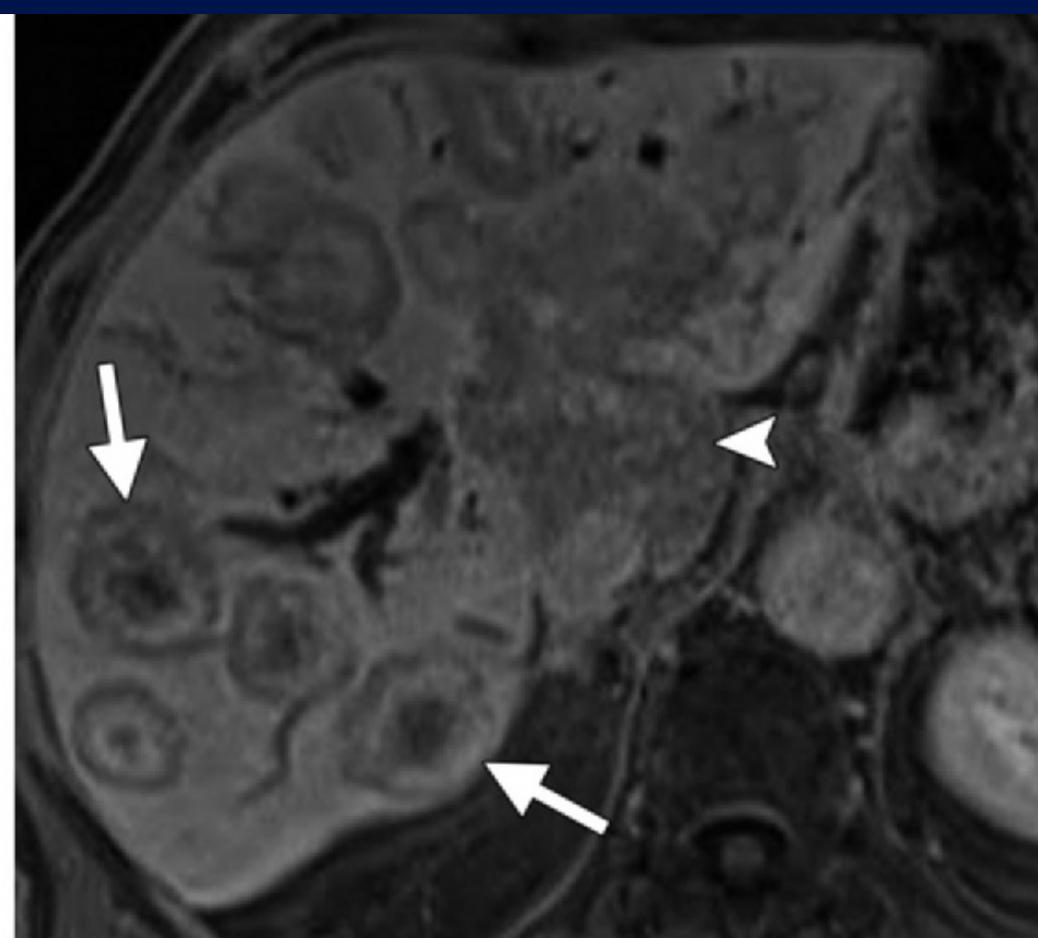
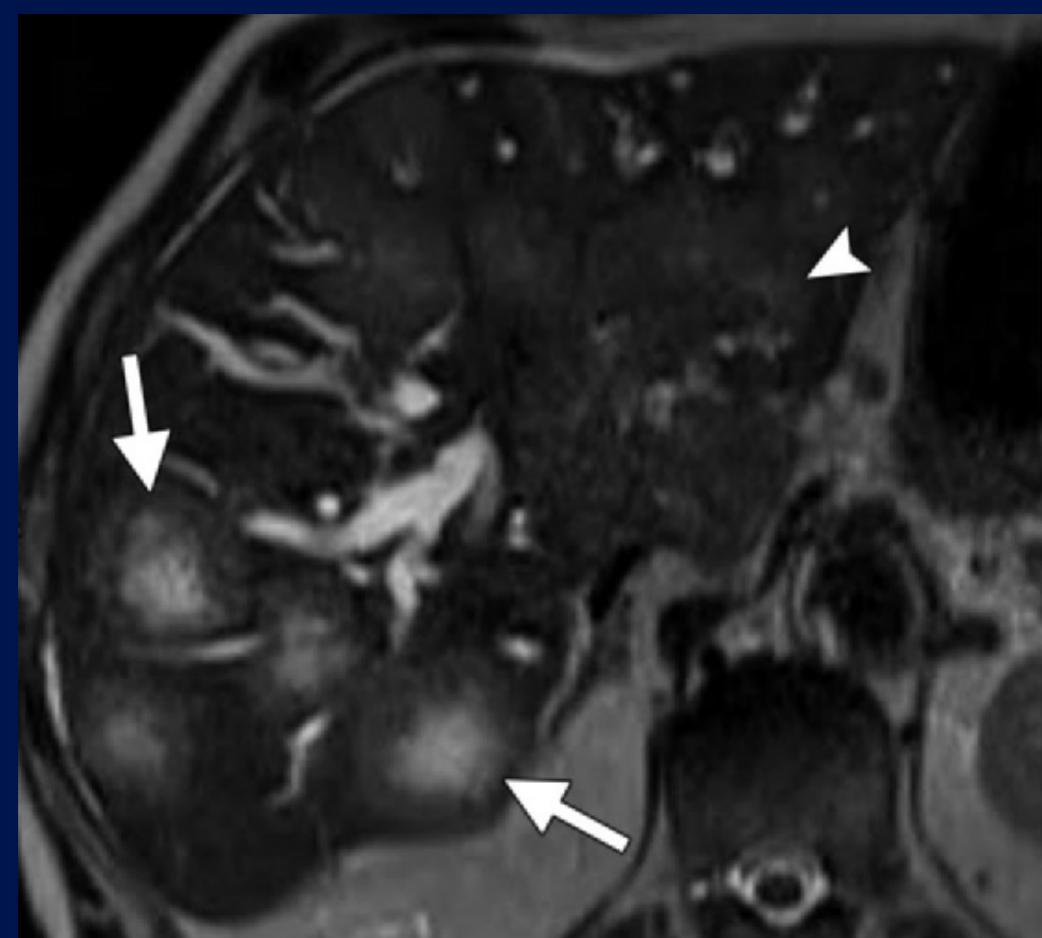
Miliary pattern in an 82-year-old man with secondary large B-cell hepatic lymphoma who presented with abdominal fullness. Coronal CT image shows multiple solid hypoenhancing masses (arrowhead) in a miliary pattern throughout the liver. Moderate periportal adenopathy (black arrow) and splenic lesions (white arrow) are also seen



77-year-old woman with known lymphoma who presented with splenomegaly at physical examination. Axial CT image shows hepatomegaly with discrete, predominantly homogeneous, hypoenhancing masses (arrow). An unaffected vessel (white arrowhead) is seen coursing through the mass, and splenomegaly is also seen (black arrowhead). The combination of hepatosplenomegaly and vessel encasement without occlusion suggests a hematologic disorder.

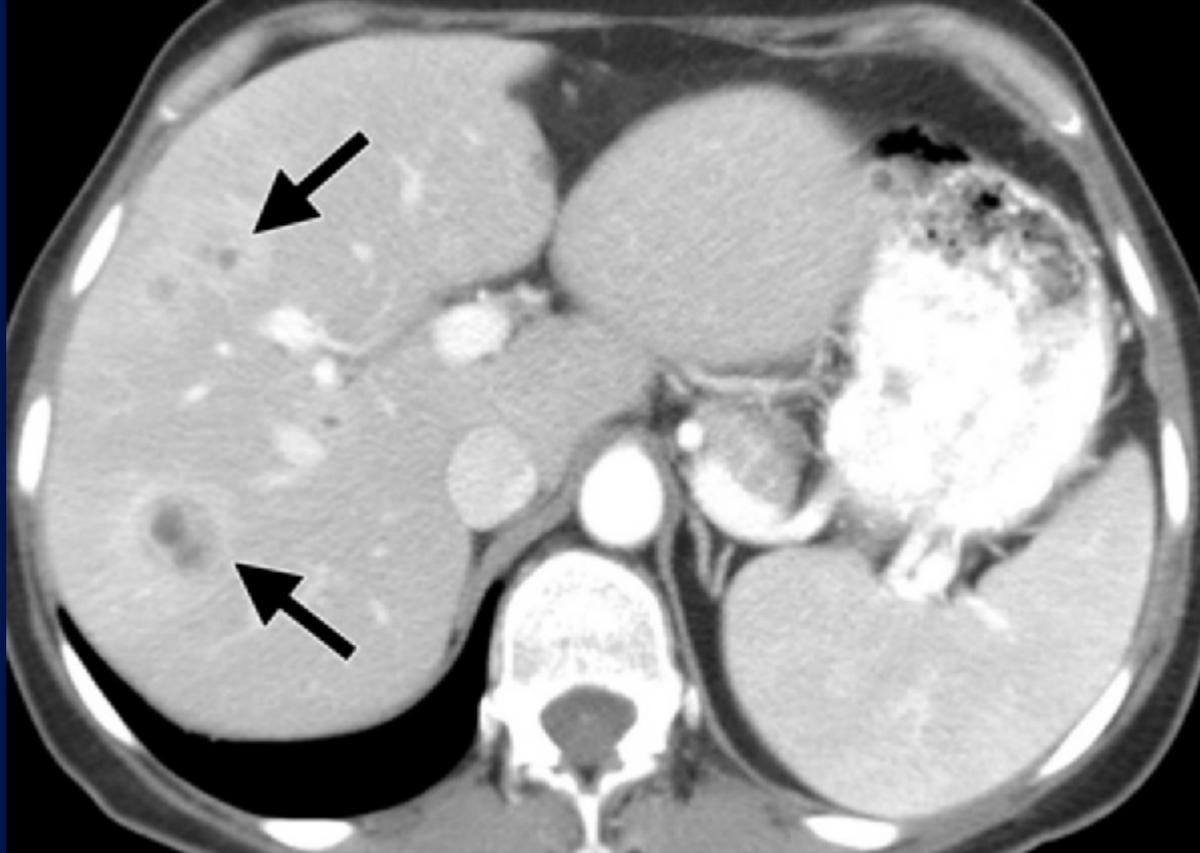


Hepatic lymphoma in a 56-year-old. Axial T1-weighted (a) and T2-weighted (b) MR images show numerous nodules (arrows), some confluent, which are hypointense on the T1-weighted image and hyperintense on the T2-weighted image. (c) Diffusion-weighted MR image ($b = 500 \text{ sec/mm}^2$) shows the lesions (arrows) as hyperintense relative to the liver. (d) Axial gadolinium-enhanced MR image shows mildly hypoenhancing lesions (arrows).

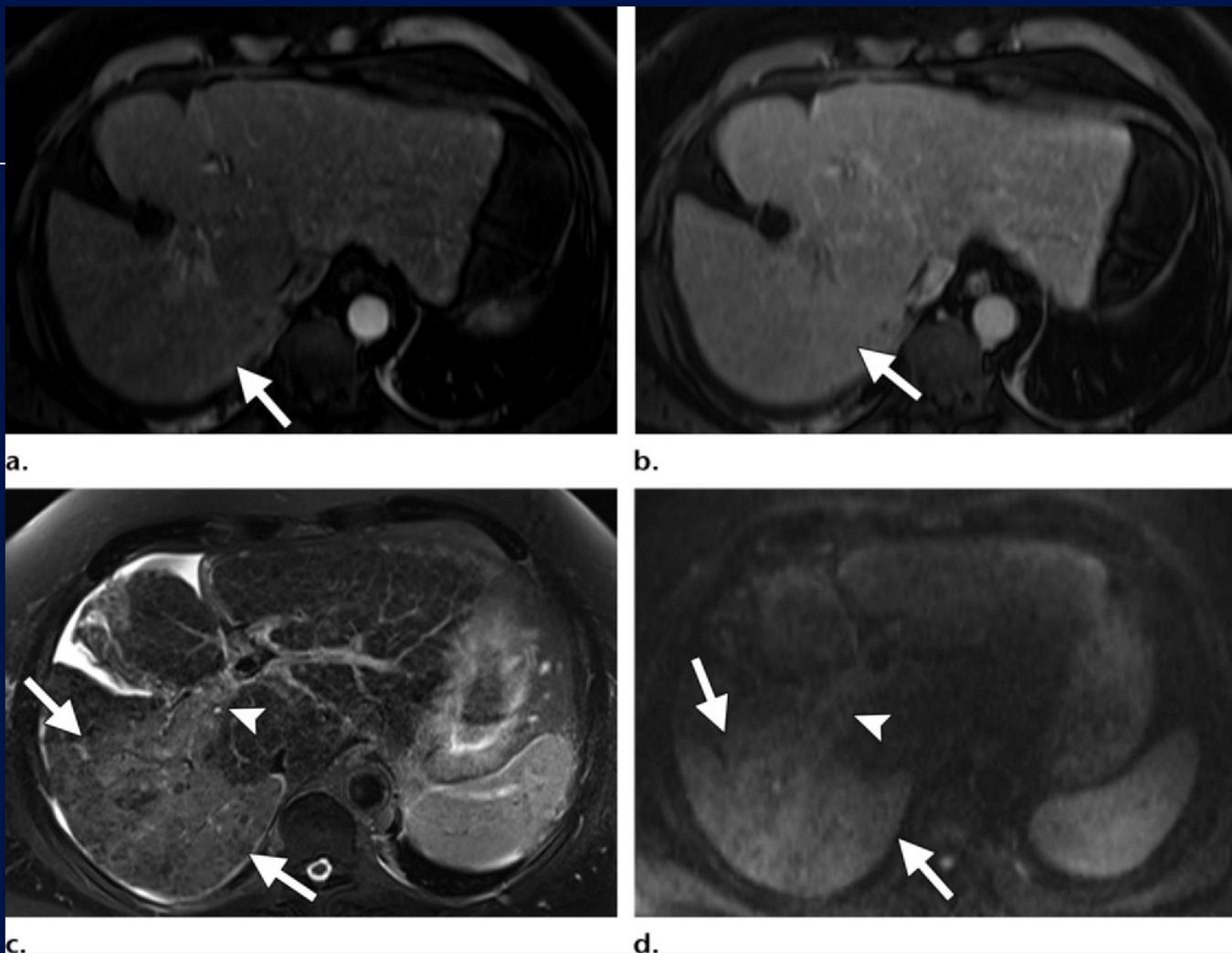


80-year-old man with large B-cell secondary hepatic lymphoma. (a) Axial T2-weighted MR image shows multiple hepatic lesions (arrows) with a targetlike appearance, with a hyperintense center and a hypointense periphery. A more infiltrative mass (arrowhead) in the porta hepatis extends into the left lobe and obstructs the bile duct. (b) Axial gadolinium-enhanced MR image shows the targetlike lesions (arrows) with poorly enhancing centers. The ill-defined infiltrating mass (arrowhead) is seen in the porta hepatis.

Differential Diagnosis



52-year-old woman with AML who presented with fever. Axial CT image shows small, ill-defined, low-attenuating hepatic masses (arrows) with surrounding hyperemia. Guided liver biopsy demonstrated fungal microabscesses.



67-year-old man with cirrhosis and elevated α -fetoprotein levels. (a, b) Axial gadolinium-enhanced arterial phase (a) and venous phase (b) MR images show no discernible abnormality in the right lobe of the liver (arrow). (c) Axial T2-weighted MR image shows definite signal hyperintensity in the right posterior lobe (arrows), with extension of high signal intensity into the right portal vein (arrowhead). (d) Diffusion-weighted MR image ($b = 500 \text{ sec/mm}^2$) shows signal hyperintensity in the right lobe (arrows) and right portal vein (arrowhead).

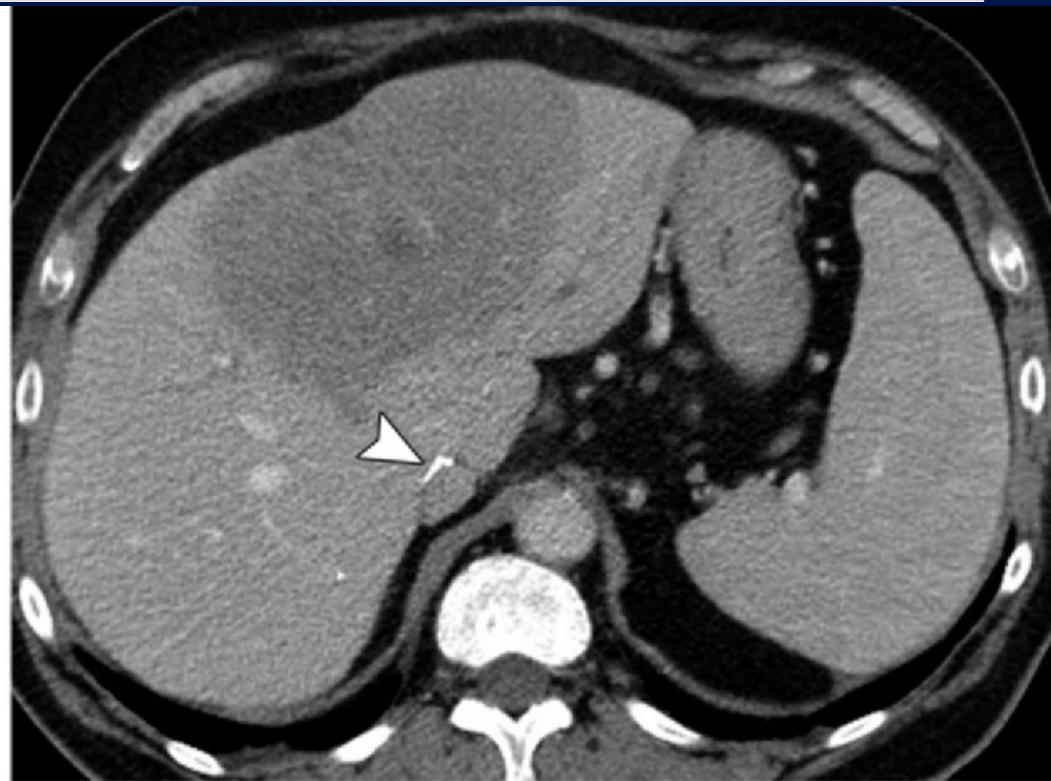
Posttransplant Lymphoproliferative Disorder (PTLD)

- ◆ The incidence of PTLD and the prognosis vary according to the organ transplanted, recipient age, and intensity of immunosuppression therapy
- ◆ The risk for developing PTLD is greatest within 1 year of transplantation and declines over time thereafter
- ◆ PTLD has a high propensity for extranodal involvement (80%). The liver is the most commonly involved abdominal organ (50%), followed by the small bowel (25%) and kidneys (17%)

- ◆ The most common manifestation of PTLN in the liver is one or more poorly enhancing masses
- ◆ Occasionally, an ill-defined, heterogeneous, infiltrating mass is seen
- ◆ A third pattern, characterized by a mass in the porta hepatis with biliary tree involvement and periportal lymphadenopathy, may be a characteristic feature of PTLN in liver transplant recipients
- ◆ Other imaging manifestations of PTLN include splenomegaly or splenic lesions, gallbladder or bowel wall thickening, biliary obstruction, and adenopathy.

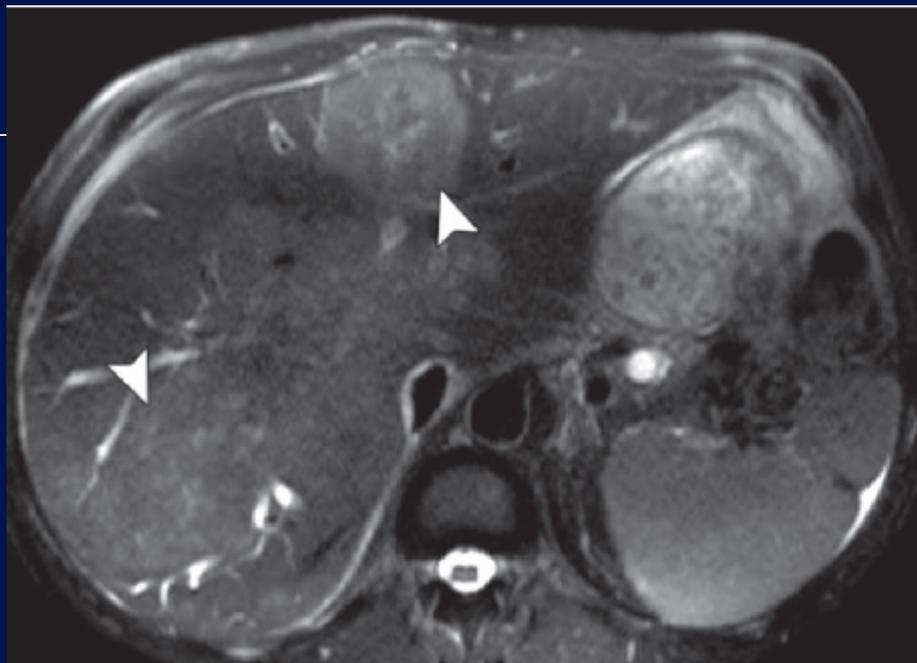


a.

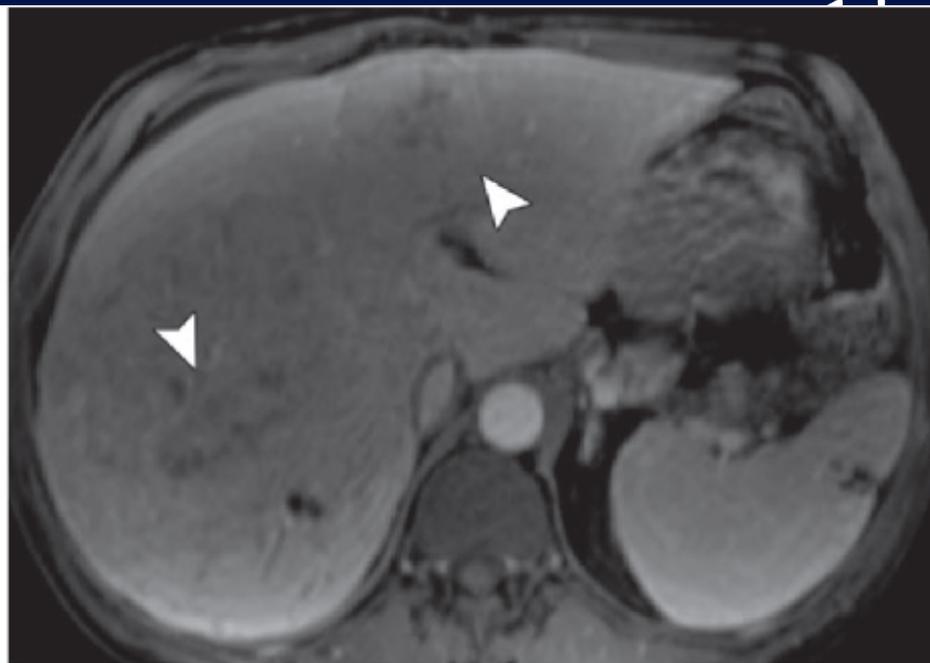


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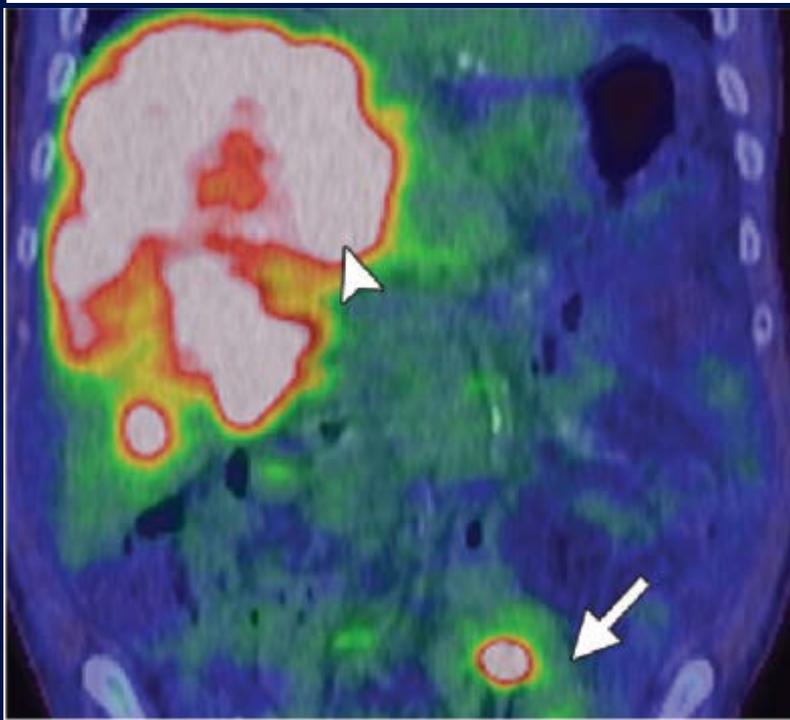
61-year-old man who presented with abdominal fullness 9 months after orthotopic liver transplant. Axial arterial phase (a) and venous phase (b) CT images show a large, well-defined, hypoenhancing mass in the left lobe, with a preserved small artery (arrow in a). Surgical clips from inferior vena cava anastomosis are seen (arrowhead). Biopsy demonstrated PTLD.



a.



b.

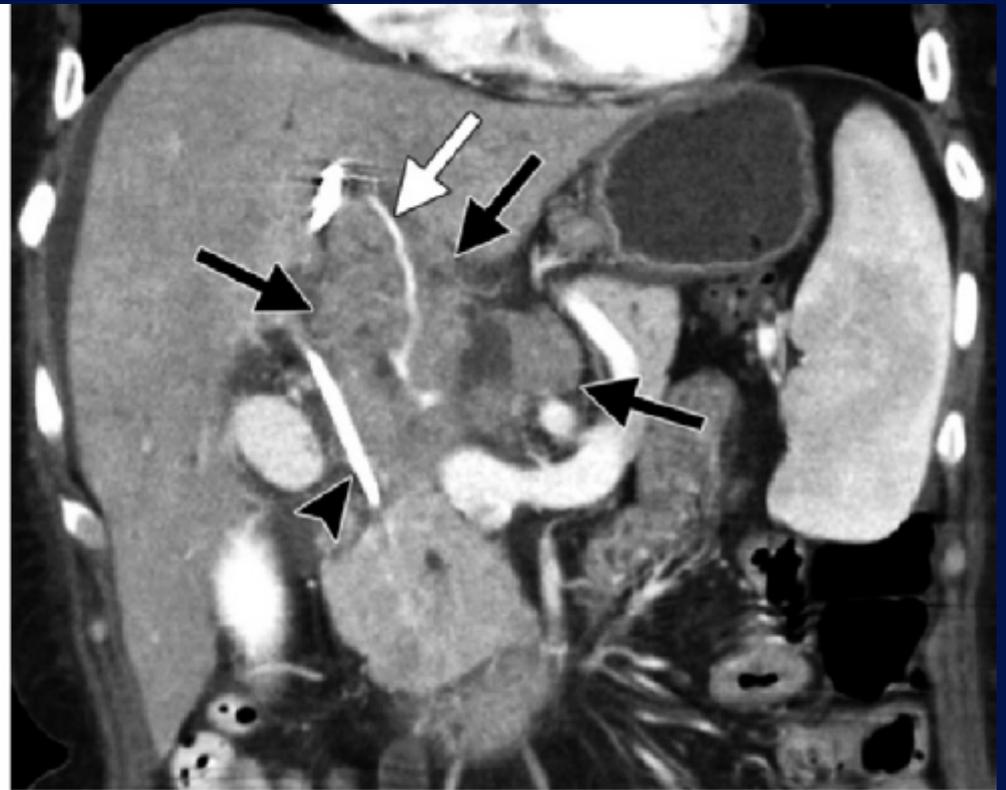


c.

PTLD in a 56-year-old man with elevated liver function test results 6 months after kidney transplant. (a) Axial T2-weighted MR image shows multiple poorly defined, mildly hyperintense hepatic masses (arrowheads). (b) Axial contrast-enhanced venous phase MR image shows barely visible lesions (arrowheads) in both lobes. (c) Coronal fused FDG PET/CT image shows the lesions as avidly hypermetabolic (arrowhead). Note the transplanted kidney (arrow).



13a.



13b.

Periportal infiltrative pattern of PTLD in a 69-year-old woman with previous liver transplant who presented with abdominal pain. (a) Axial CT image shows a poorly defined mass (arrow) in the periportal region. The mass is causing biliary obstruction, and a biliary stent is seen (black arrowhead). Focal nonocclusive portal vein thrombosis is also seen (white arrowhead). (b) Coronal reformatted CT image shows the mass (black arrows) and biliary stent (arrowhead). The hepatic artery (white arrow) runs through the mass without occlusion.

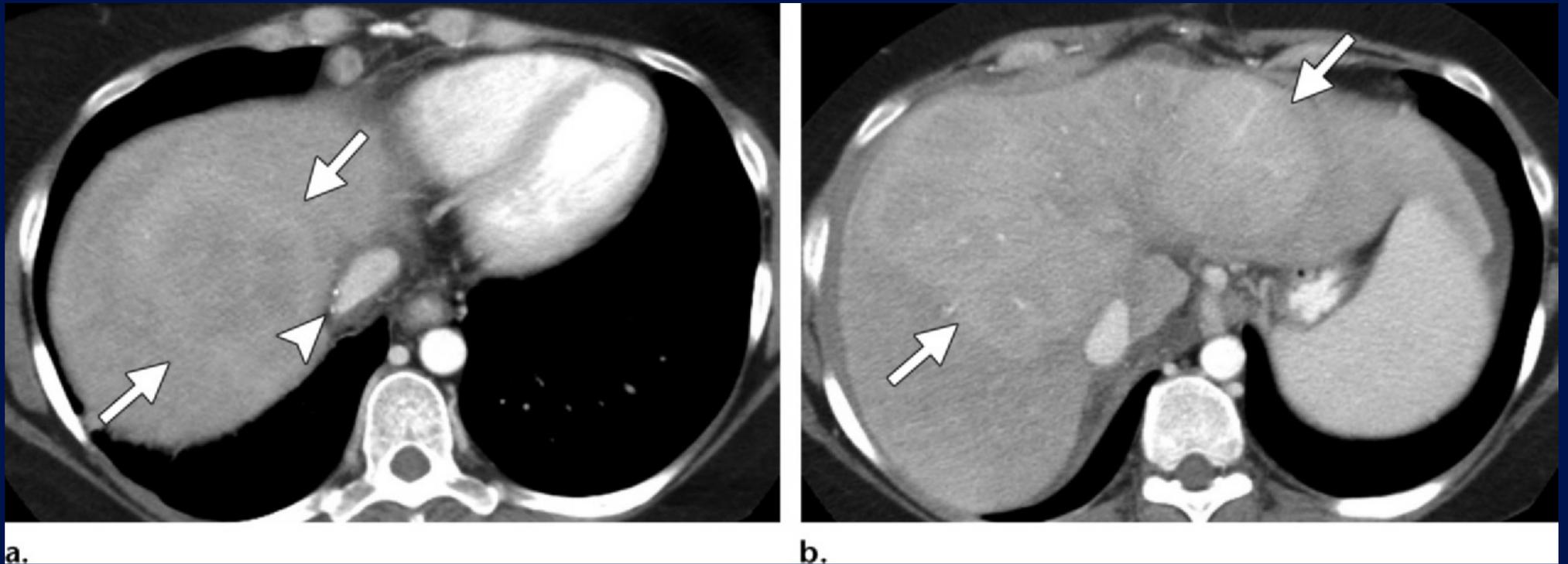


14a.



14b.

PTLD manifesting as a periportal mass in a 35-year-old man with a history of liver transplant who presented with abdominal tenderness. (a) Coronal reformatted CT image shows a large periportal mass (black arrows) enveloping but not occluding the main portal vein (arrowhead). The tumor also involves the liver (white arrow). (b) Coronal reformatted CT image obtained after medical therapy shows complete resolution of the mass.



54-year-old woman who underwent orthotopic liver transplant for cirrhosis with HCC and presented for routine HCC screening. Axial CT images show a large mildly hyperenhancing mass (arrows), with patent vessels seen coursing through the mass in b. Surgical clips are seen in the inferior vena cava (arrowhead in a). The findings are suspicious for PTLD rather than recurrent HCC. Biopsy demonstrated PTLD

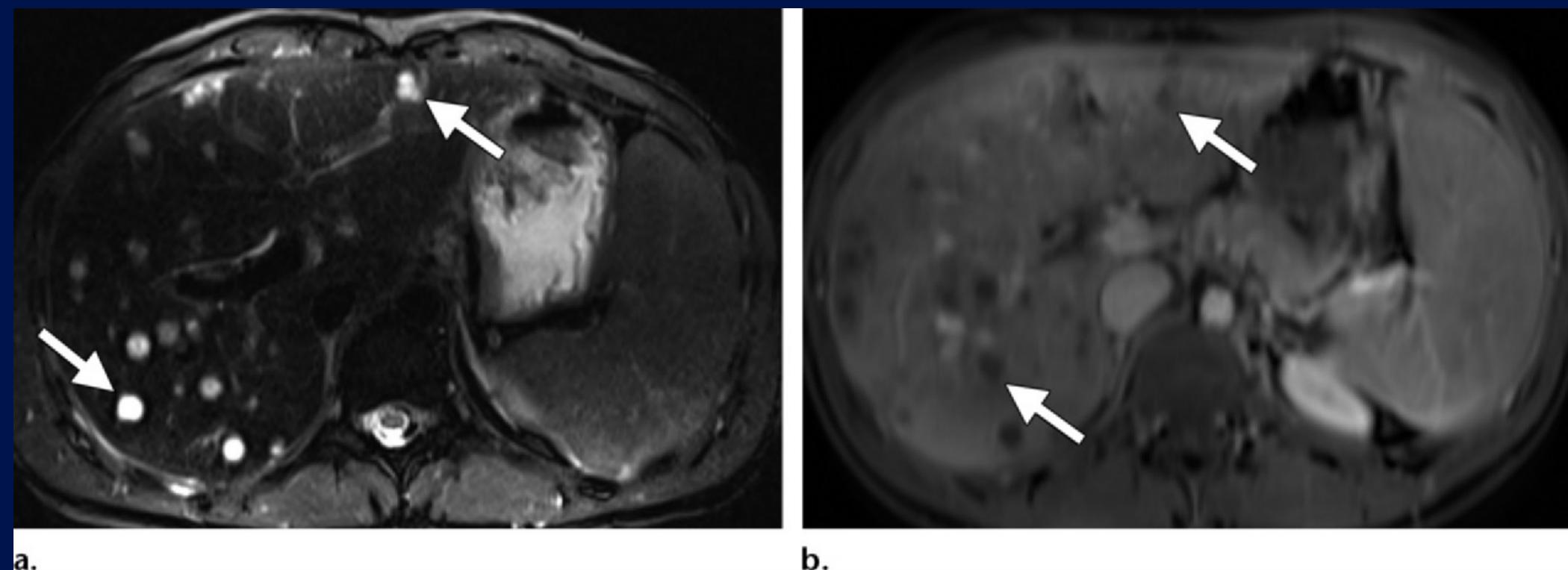
- ◆ Myeloid sarcoma is a rare extramedullary proliferation of immature myeloid cells. It is most commonly seen in patients with AML and occurs in 3%–5% of these patients
- ◆ At immunohistochemical analysis, myeloid sarcoma stains positive for myeloperoxidase, which results in green staining of the lesions (hence the term *chloroma*)
- ◆ A diagnosis of myeloid sarcoma indicates a poor outcome, irrespective of the clinical manifestations

- ◆ The most common sites of myeloid sarcoma involvement are the bones, lymph nodes, soft tissues, skin, and breasts. Less common sites are the genitourinary tract, gastrointestinal system, head and neck, and thorax
- ◆ The imaging features of hepatic myeloid sarcoma are nonspecific and are similar to those of hepatic lymphoma
- ◆ The diffuse hepatic sinusoidal infiltration of leukemic cells may result in intra- or extrahepatic biliary duct obstruction



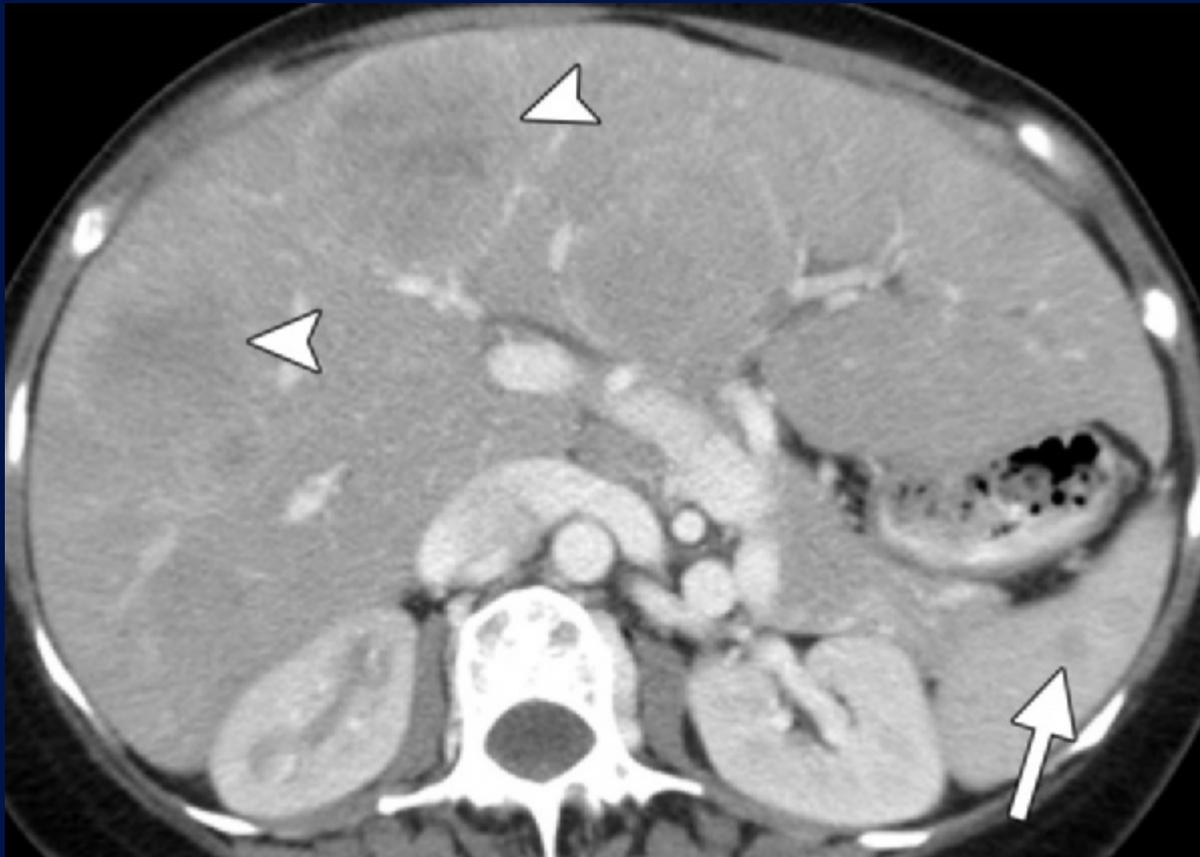
64-year-old man with myelofibrosis. Axial nonenhanced CT image shows a poorly defined hypoattenuating hepatic mass (arrowhead). The finding is nonspecific, but myeloid sarcoma is a leading consideration for a large mass in a patient with a myelogenous hematologic disorder. Biopsy demonstrated myeloid sarcoma.

Differential Diagnosis



Acute GVHD in a 21-year-old man with fever and an elevated white blood cell count 6 weeks after hemopoietic stem cell transplant. (a) Axial T2-weighted MR image shows multiple hyperintense hepatic nodules (arrows) with hypointense rims. The spleen is not affected. No adenopathy was seen. (b) Axial gadolinium-enhanced T1-weighted MR image shows poor central enhancement of the nodules (arrows), with mild peripheral enhancement. A fungal infection was suspected, but biopsy demonstrated acute GVHD.

- ◆ Extraosseous myeloma was once thought to be rare, but autopsy series have shown extraosseous disease in up to 64% of patients with myeloma
- ◆ The lymph nodes, pleura, and liver are the most commonly involved organs
- ◆ Hepatic involvement may be unifocal, multifocal, or diffuse
- ◆ Liver involvement may be asymptomatic or may manifest as hepatomegaly, jaundice, ascites, or fulminant liver failure.
- ◆ At CT, the most common finding is hepatomegaly. Focal hepatic lesions are typically hypoattenuating, without calcification or substantial contrast enhancement
- ◆ Myelomatous lesions are usually hyperintense on T1-weighted and T2-weighted MR images. There often is minimal enhancement on gadolinium-enhanced images

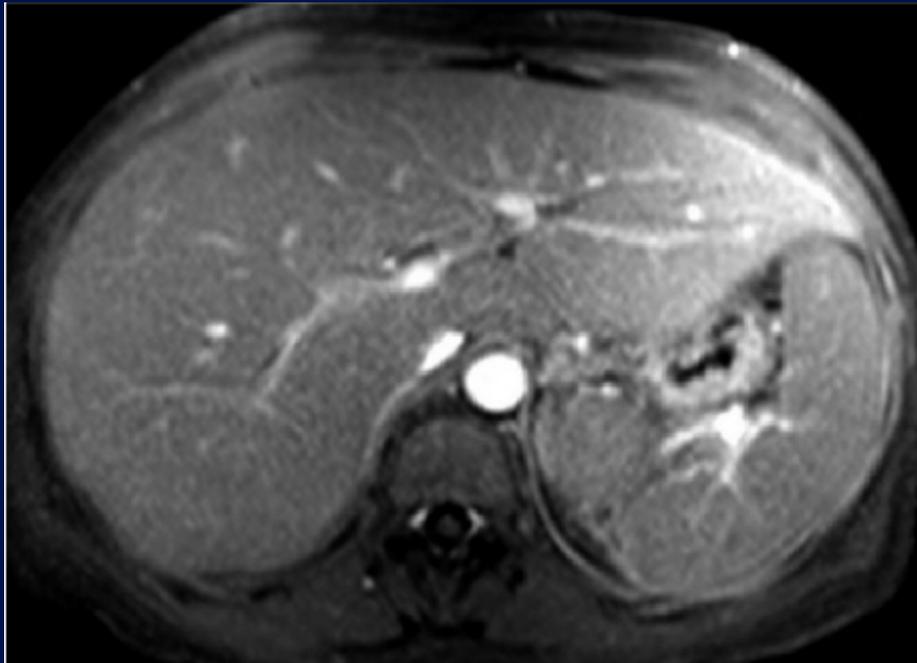


37-year-old woman. Axial CT image obtained to locate a possible primary malignancy shows multiple solid lesions in the liver (arrowheads) and spleen (arrow). The lesions are mildly hypoenhancing and do not show calcification. Biopsy of one of the liver lesions demonstrated myeloma.

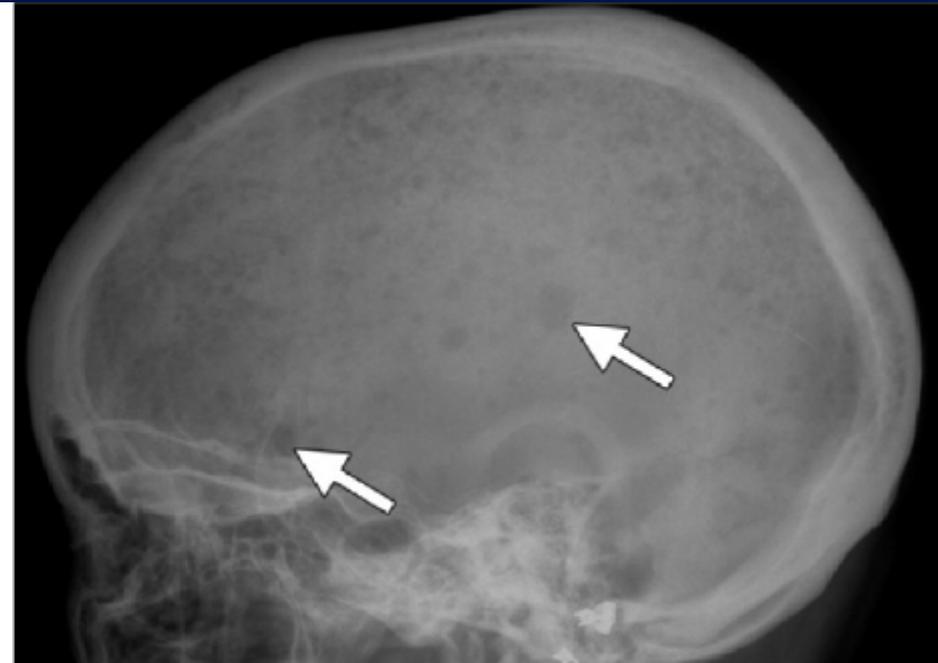
61-year-old woman with known myeloma and abnormal liver function test results. (a) Axial T2-weighted MR image shows minimally hyperintense hepatic lesions (arrowheads). (b) Axial gadolinium-enhanced MR image does not depict the hepatic lesions. (c) Lateral skull radiograph shows numerous lytic lesions (arrows), a finding consistent with the diagnosis of extraosseous myeloma.



a.



b.



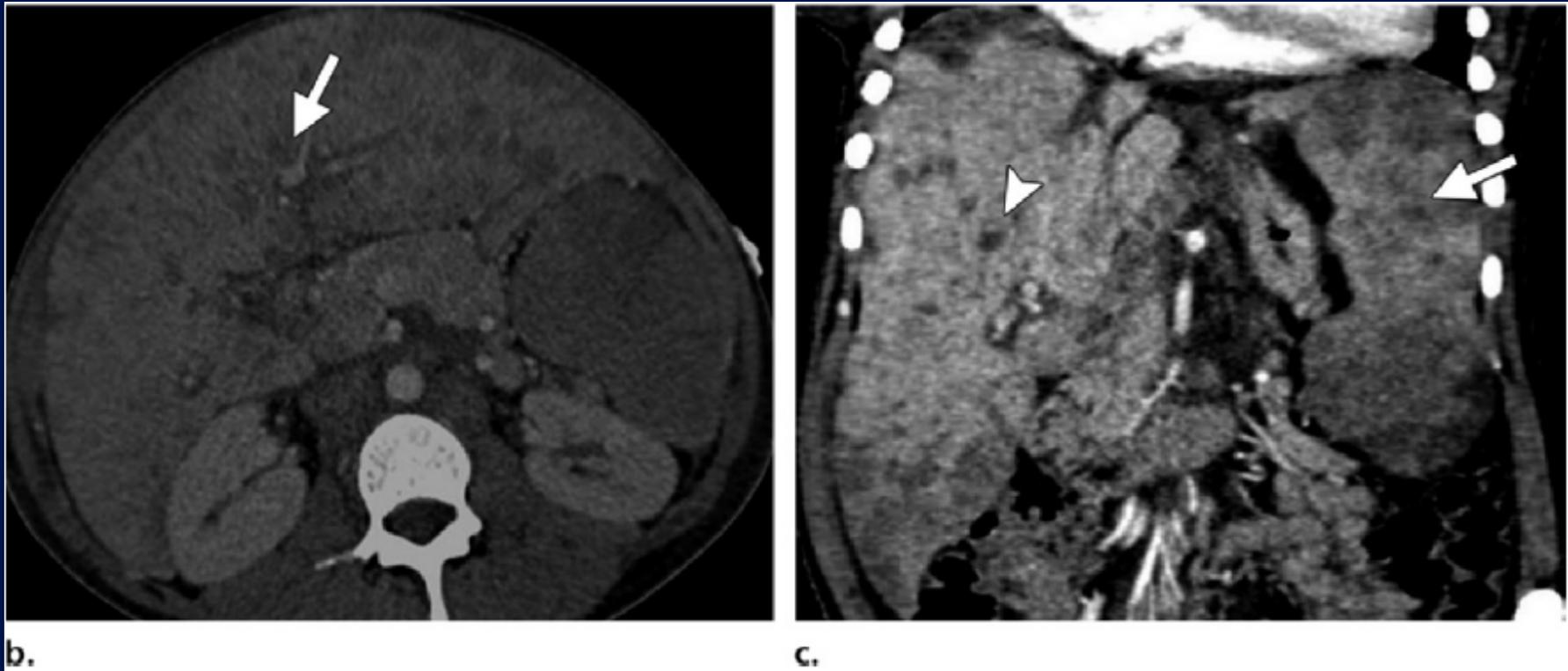
c.

- ◆ Castleman disease (angiofollicular lymph node hyperplasia or giant lymph node hyperplasia) is a nonclonal lymphoproliferative disorder and is one of the more common causes of lymphadenopathy
- ◆ Extralymphatic sites of involvement include the lungs, larynx, parotid glands, pancreas, meninges, and muscles. Liver involvement is uncommon
- ◆ Castleman disease is classified as hyaline vascular (90%) or plasma cell type (10%)
- ◆ The plasma cell type typically demonstrates less intense contrast enhancement compared with the hyaline vascular form



47-year-old woman. Axial contrast-enhanced CT image shows a homogeneously hyperattenuating periportal nodal mass (arrowhead) and retroperitoneal nodes. Biopsy of a mediastinal lymph node (not shown) demonstrated Castleman disease.

- ◆ HLH, or hemophagocytic syndrome, is a multisystem disorder characterized by cytokine dysfunction that results in uncontrolled proliferation of activated cytotoxic T cells, antigen-presenting cells, macrophages, and histiocytes
- ◆ Early clinical signs include fever (90% of cases), hepatosplenomegaly (90%), lymphadenopathy (42%), rashes, and neurologic abnormalities (47%)



38-year-old man who presented with weight loss and flu-like illness. (b) Axial CT image shows marked hepatosplenomegaly, with numerous hypoenhancing masses (arrow). (c) Coronal contrast-enhanced MR image shows numerous hypoenhancing lesions in the liver (arrowhead) and spleen (arrow), with hepatosplenomegaly. The findings are nonspecific, and the differential diagnosis includes metastases and lymphoma. Biopsy demonstrated HLH.

Combining the imaging features with clinical manifestations and laboratory findings can facilitate correct diagnosis. Clinical features that suggest a hematologic neoplasm as the cause of liver lesions include a young patient (<40 years of age), no known history of cancer, abnormal bone marrow biopsy results, fever of unknown origin, and night sweats

Imaging features that suggest hematologic malignancy include hepatosplenomegaly or splenic lesions, vascular encasement by a tumor without occlusion or thrombosis, an infiltrating mass at the hepatic hilum with no biliary obstruction, and widespread adenopathy above and below the diaphragm

Biopsy need performed, the predominant treatment of hematologic malignancies is chemotherapy or radiation therapy rather than surgery.

THANK YOU!

Sun Yat-sen University Cancer Center

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