Have you seen it all? Then take a stab at these unusual liver lesions!

Natally Horvat, MD $^{1,2}$
Maria LaGratta, MD$^1$
Richard Kinh Gian Do, MD, PhD$^1$

Memorial Sloan Kettering Cancer Center, New York, NY
Hospital Sírio-Libanês, São Paulo, Brazil

The authors have nothing to disclose
OBJECTIVES

✓ To review the appearance of rare neoplastic and non-neoplastic hepatic lesions.
✓ To develop a diagnostic approach to rare hepatic lesions using imaging findings and clinical features.

CONTENTS

Infectious diseases
- Larva migrans
- Echinococcosis
- Schistosomiasis
- Fungal abscesses

Inflammatory disorders
- Amyloidosis
- Sarcoidosis

Cystic Tumors
- Mucinous cystadenoma
- Cystic metastasis
- Ciliated hepatic cyst

Solid Tumors
- Hepatic metastasis
- Schwannoma
- Angiosarcoma
- Solitary fibrous tumor
- Lymphoma
- Angiomyolipoma
- Post transplant - lymphoproliferative disease
- Fibrolamellar carcinoma
- Epithelioid Hemangioendothelioma

Miscellaneous
- Hepatic splenosis
- Ectopic liver tissue

All white boxes provide important information for radiological interpretation.
Visceral *Larva migrans*

- Worldwide distribution
- Labs: eosinophilia and hypergammaglobulinemia
- Imaging findings:
  - Multiple
  - Elongated
  - Ill-defined
  - Migratory
  - Small dimensions (< 2 cm)

46-year-old woman treating cervical cancer, presented an interval development of migratory elongated liver lesion (arrows) and eosinophilia. Serology confirmed the diagnosis.

- Peripheral distribution
- Migratory lesions
- Eosinophilia
Echinococcosis

- Four subtypes: cystic, alveolar, polycystic, unicystic
- Imaging findings:
  - ~ simple cyst (early stages)
  - Multiseptate wheel-like with daughter cysts
  - Wall calcifications (after parasite death)
- Complications: bile duct compression, biliary communication, peritoneal seeding and rupture → fever, pruritus and anaphylaxis

45-year-old man with abdominal pain. Non-contrast enhanced CT (a) and T2 WI MRI (b) demonstrate cystic lesions with multiple peripheral “daughter” cysts and calcification (arrow). Serology confirmed echinococcosis.
Schistosomiasis

- Sub-Saharan Africa, Middle East, South America, Caribbean
- Imaging findings:
  - Irregular liver surface
  - Atrophy of the right hepatic lobe
  - Periportal and peribiliary fibrosis ("clay-pipestem" pattern)
  - Severe portal hypertension
  - Splenic siderotic nodules

Patients with hepatosplenic schistosomiasis. US demonstrate periportal hyperechogenicity (a, red arrow), marked thickening and echogenicity of the gallbladder bed (b, white arrows), multiple hypoechoic splenic nodules (c), and CECT shows lobulated liver surface, atrophy of the right hepatic lobe and portal hypertension (d).
Fungal Abscess – Dark Ring Sign

- Immunocompromised patients
- Patients with transfusional hemosiderosis may present perilesional low signal on T1 and T2WI → confirm the diagnosis of fungal abscess

42-year-old male with acute myeloid leukemia and fever, with a history of fungal pneumonia and transfusional hemosiderosis. (a,c) T1WI in-phase and (b,d) CE T1WI. In the admission (a,b) MRI demonstrated iron deposition in the spleen and multiple lesions in the liver. After an empirical antifungal treatment (c,d) the hepatic lesions reduced and appeared dark ring halo surrounding them.

- Dark ring halo on MRI surrounding hepatic lesions after antifungal treatment confirms fungal infection
Hepatic Amyloidosis

- Primary – worse prognosis
- Imaging findings (non-specific):
  - Hepatomegaly
  - Diffuse or focal regions of decreased parenchymal attenuation on CT
  - Increased SI on T1WI
  - Very high liver stiffness values on US or MR elastography

Systemic amyloidosis
- Intact surrounding vascular architecture
- Very high liver stiffness values on US or MR elastography

57-year-old woman with recently diagnosed systemic amyloidosis. T2WI (a) and T1WI (b) demonstrate hepatomegaly, peripheral bandlike areas of low T1 and high T2 SI in the left lobe (arrows). The liver parenchyma presented slightly high SI on T1WI (b) and on portal venous phase (c) reticular pattern and areas with reduced enhancement in the left lobe (asterisk). Liver biopsy confirmed the diagnosis.
Hepatic Sarcoidosis

- Liver, spleen, heart and bone marrow – most common extrapulmonary sites
- Imaging findings:
  - Hepatomegaly +/- splenomegaly +/- lymph node enlargement
  - Hepatic granulomas: multiple low-attenuating lesions with variable sizes

CE CT in a patient with known systemic sarcoidosis demonstrated hepatomegaly, multiple small low-attenuating hepatic lesions and lymph node enlargement (arrows).
Mucinous Cystadenoma

- Middle-age female patients
- Risk of malignant transformation → Surgical resection is required
- Imaging findings:
  - Solitary cystic lesion in the left lobe
  - Enhancement along the wall and internal septa
  - May contain calcification

56-year-old woman. CT images before (a) and after (b) contrast injection show a solitary cystic lesion in the left lobe with enhancing septa and calcification (arrow). Surgical resection confirmed the diagnosis.

42-year-old woman. T2WI MRI (a) and CE T1WI demonstrate a solitary cystic lesion in the left lobe with thin septations. Surgical resection confirmed the diagnosis.
Cystic Metastasis

✓ Cause by:
  ✓ Necrosis due to rapid growth (i.e., neuroendocrine, melanoma, GIST)
  ✓ Mucin (i.e., colorectal or ovarian carcinoma)
  ✓ Systemic of locoregional treatment

- Metastatic lesions can be paradoxically larger after treatment due to cystic degeneration
- Mucinous mets can be cystic

70-year-old woman with metastatic mucinous rectal cancer. CECT show the mucinous metastasis in the liver and in the porta hepatic lymph nodes (a), and the rectal lesion (b, arrow).

Cystic transformation of a hepatic metastasis of gastric GIST (arrowhead) after 4 months of imatinib (b).
Rare cases of malignant transformation in squamous cell carcinoma

Imaging findings:
- Solitary
- Unilocular
- Subcapsular
- IV segment of the liver
- Variable density on CT and signal on MRI

MRI demonstrate a subcapsular cyst in segment IV with hypointense content on T2WI (a, arrow), marked restriction on DWI (b,c), without enhancement (d). Abdominal US shows low-level echos within the cyst.
Liver Metastasis of Immature Teratoma

- Immature teratoma of the ovary is a rare tumor
- First 2 decades of life
- Imaging findings:
  - May contain foci of fat and calcification

Hepatic metastasis (a) of an immature ovarian teratoma (b), with foci of fat and calcification.
Liver Metastasis within a Focal Nodular Hyperplasia

- FNH is the second most common benign liver tumor
- Rare case reports of metastasis to a FNH
- High blood flow to HNF may predisposes the development metastasis

Liver metastasis of breast cancer within FNH. CECT in October 2009 (a), December 2009 (b) and March 2010 (c) show the lesion increased in size (arrow), as well as other new metastasis.
Porta Hepatic Schwannoma

- Asymptomatic or obstructive jaundice
- Pre-operative diagnosis is difficult
- DDx: lymphadenopathy, mesenchymal tumors, cholangiocarcinoma
- Imaging findings (non-specific):
  - Homogeneous
  - Well-demarcated

CECT images (a,b) demonstrate a homogeneous solid elongated mass in the porta hepatis (arrows) causing mild biliary tree dilatation (asterixis). Porta hepatic schwannoma was confirmed after surgery.
Angiosarcoma

- Aggressive, multifocal, hypervascular tumor
- Rapid and early clinical deterioration

6th-7th decades, M > F
Prognosis is very poor
DDx: hemangioma, HCC, hypervascular metastases
Imaging findings (most frequent):
  - Multifocal, hypervascular, variable necrosis
  - Hemorrhagic areas, fluid-fluid levels
  - Spleen may be affected

Biopsy-proven angiosarcoma in a 72-year-old woman with rapid deterioration in her functional status. Contrast-enhanced CT images demonstrates multifocal hypervascular masses in the liver and in the spleen.
Solitary Fibrous Tumor

- Slowly growing mass
- Malignant potential
- Imaging findings (non-specific):
  - Single
  - Large
  - Well-circumscribed
  - Heterogeneous enhancement

Surgically proven solitary fibrous tumor in a 59-year-old woman. T2WI (a) and T1WI (b) MR demonstrates a large well-circumscribed, heterogeneous mass in the right hepatic lobe. Contrast-enhanced MR obtained during arterial (c), portal venous (d), and delayed (e) phases show a progressive peripheral enhancement with curvilinear and patchy pattern.
26-year-old woman treating Hodgkin Lymphoma with a new hepatic lesion. The hepatic nodule presented high SI on T2WI (a), marked restriction on DWI (b,c), low SI on T1WI (d), and progressive enhancement (e,f). Liver biopsy confirmed the diagnosis of lymphoma.

- History of lymphoma
- Hypovascular lesions
- Marked restriction on DWI

- Secondary >>> Primary
- Risk factors: immunosuppression
- Imaging findings (variable and non-specific):
  - Multiple >> Single
  - Infiltrative
  - Hypovascular
  - MR: hyper T2WI, hypo T1WI, marked restriction of DWI
Post Transplant Lymphoproliferative Disease

- Extranodal >> nodal involvement
- DDx: Opportunistic infections and recurrent / new malignancy
- Imaging features (non-specific):
  - Single or multiple hypovascular masses
  - Mass in the portal hepatis
  - Diffuse infiltration of the liver

21-year-old male post bone marrow transplantation. CECT show hypovascular lesions in the liver and spleen, as well as hepatic hilar lymphadenopathy. Liver biopsy confirmed the diagnosis of monomorphic post-transplant lymphoproliferative disorder.
Epithelioid Hemangioendothelioma

- M < F, 25-58 years
- Survival 5-10 years after the diagnosis, even metastatic
- Imaging findings:
  - Coalescent peripheral / subcapsular nodules
  - Target-like appearance
  - Capsular retraction
  - May present with calcification

36-year-old woman with liver EH metastatic to the lung. CT and MRI (a,b,c) show subcapsular confluent nodules, with target appearance. 6 years later (d,e) the lesions were calcified and the uninvolved parenchyma were hypertrophic.
18-year-old female with palpable mass. CT demonstrate a large exophytic hepatic lesion, with central calcification (a, arrow) and heterogeneous enhancement (b). The patient presented a lymph node metastasis in the porta hepatis (c, red arrow).

- Adolescents and young adults
- Low alpha-fetoprotein
- DDx: FNH (scar on T2WI: low and heterogeneous SI in FC vs high SI in FNH)
- Imaging findings:
  - Heterogeneous enhancement
  - May have central calcified scar, large nodal metastases

- Young patients
- Central scar
Angiomyolipoma

- Benign tumor
- Varying proportions of smooth muscle, blood vessels and adipose tissue
- DDx: adenoma, HCC
- Imaging findings (highly variable):
  - Well-defined lesion
  - Fat component (~60%)
  - Hyperenhancement in arterial phase
  - Washout

51-year-old woman previously healthy with an incidentally found hepatic nodule. MRI demonstrate a hepatic nodule (arrow) without obvious fat content (a,b), hypervascular on arterial phase (d) and with washout on delayed phase (e). The patient underwent partial hepatectomy and angiomyolipoma was diagnosed.
Hepatic Splenosis

- Ectopic splenic tissue
- After trauma or splenectomy
- Imaging findings:
  - Well-defined nodules with density and signal similar to spleen
  - Scintigraphy
    - Tc 99m sulphur colloid
    - Tc 99m tagged heat-damaged RBC with autologous erythrocytes

47-year-old male with hepatitis C, liver mass on ultrasound and prior traumatic splenectomy. CECT images show hypervascular masses in the liver (a, arrow), perigastric (a, arrowhead) and pelvis (b, arrow). Splenosis was surgically confirmed, due to suspicion of HCC.
Ectopic Liver Tissue

- Common areas: next to gallbladder, abdominal and thoracic wall
- Risk of complications: bleeding, obstruct surrounding structures, malignant transformation to HCC
- Imaging findings:
  - Well-defined nodules with density and signal similar to liver
  - Enhancement on hepatobiliary phase (HBP)

Ectopic liver tissue incidentally detected on endoscopy in an asymptomatic 35-year-old woman. MRI show a nodule next to the gastric greater curvature (a,d, arrow) hypervascular on arterial phase (b, arrow) and presenting enhancement on hepatobiliary phase (c, arrow). Hepatic tissue was confirmed on biopsy.
✓ The liver is commonly involved in many primary and secondary neoplastic diseases as well as infectious and inflammatory diseases.

✓ The rare and atypical liver lesions may have a wide spectrum of morphological and enhancement patterns on CT and MRI.

✓ The clinical background of the patient and some imaging findings may help in narrowing the differential diagnosis.

✓ Radiologists are familiar with common benign and malignant liver lesions. However, lack of exposure to rare liver neoplasms and infectious/inflammatory entities may limit their utility to referring physicians.
REFERENCES


AUTHOR INFORMATION

Corresponding Author
Natally Horvat, MD
1275 York Ave, New York, NY, 10065, USA
Tel. +1 (212) 639-3418
E-mail: natallymhorvat@gmail.com