Magnetic Resonance (MR) Imaging
CASE 3.1

CLINICAL HISTORY 65-year-old female with liver lesion noted as an incidental finding on CT performed for abdominal pain; MR obtained for further evaluation.
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Morphologic changes of chronic liver disease (example, a nodular liver as in this case) favor a diagnosis of hepatocellular cancer. By comparison, a hyperenhancing lesion with intracellular lipid and washout in an otherwise healthy young woman taking oral contraceptive pills is most likely a hepatocellular adenoma.

Questions for Further Thought
1. Should this lesion be biopsied?
2. What are treatment options for this lesion?

Reporting Responsibilities
1. Describe the size and location of the hepatocellular carcinoma.
2. Notify the ordering physician of this finding.

What the Treating Physician Needs to Know
1. This patient’s liver lesion is compatible with a hepatocellular carcinoma.
2. The patient has mild morphologic changes of chronic liver disease.

Answers
1. No. The lesion should not be biopsied. When a lesion is compatible with hepatocellular carcinoma based on imaging, biopsy should not be performed because of risk of tract seeding.
2. Because of her mild morphologic changes of chronic liver disease, this patient underwent a left hepatectomy. For individuals with more advanced chronic liver disease and who are within Milan criteria (up to 3 hepatocellular carcinomas ≤3 cm or 1 hepatocellular carcinoma ≤5 cm), liver transplant is the preferred treatment. Patients who are outside of Milan criteria often benefit from locoregional therapy (for example, transcatheter chemoembolization).

REFERENCES
CASE 3.2

CLINICAL HISTORY 39-year-old woman presenting with right upper quadrant pain.

FINDINGS Coronal T2-weighted (A), T1-weighted pre- (B) and postcontrast (C) images obtained through the liver. Edema is present as evidenced by increased T2 signal and decreased T1 signal with relative sparing of the caudate lobe. Postcontrast image demonstrates heterogeneous enhancement of the liver with more homogeneous enhancement of the caudate lobe (contrast agent: gadobenate dimeglumine [Gd-BOPTA, MultiHance; Bracco Diagnostics, Milan, Italy]). Venogram (D) performed via a right internal jugular vein approach demonstrates nonfilling of the central hepatic veins.
**DIFFERENTIAL DIAGNOSIS**  Budd-Chiari syndrome, primary sclerosing cholangitis (PSC).

**DIAGNOSIS**  Budd-Chiari syndrome.

**DISCUSSION**  Budd-Chiari syndrome is defined as hepatic venous outflow obstruction and may be due to hypercoagulable states (e.g., antiphospholipid syndrome, protein C and S deficiencies, and pregnancy), membranous webs, or direct invasion of the hepatic veins or inferior vena cava by neoplasm (e.g., hepatocellular, adrenal, or renal cell carcinoma). Budd-Chiari syndrome most commonly occurs in women and young adults.

With acute Budd-Chiari syndrome, the liver is enlarged with heterogeneously decreased signal on T1-weighted pre-contrast images and increased signal on T2-weighted images indicating edema with relative sparing of the caudate lobe as in the above case. A central versus peripheral enhancement pattern also is seen with decreased and heterogeneous peripheral enhancement and relatively increased and homogeneous central enhancement. This peripheral heterogeneous enhancement occurs because of decreased blood flow due to increased tissue pressure. Relatively increased and homogeneous caudate lobe enhancement is thought to reflect alternate venous drainage of the caudate lobe.

With chronic Budd-Chiari syndrome, caudate lobe hypertrophy, irregularities of liver contour due to cirrhosis, and regenerative nodules are seen. PSC can also result in caudate lobe hypertrophy due to a separate biliary drainage pathway, but with PSC irregular intrahepatic biliary ductal dilatation is seen.

**Question for Further Thought**

1. What treatment options are available for Budd-Chiari syndrome?

**Reporting Responsibilities**

1. Identify occlusion of the hepatic veins.
2. Determine etiology of hepatic vein thrombosis (bland thrombus versus tumor thrombus).

**What the Treating Physician Needs to Know**

1. The extent of hepatic vein occlusion (complete versus incomplete thrombosis; are right, middle, and left hepatic veins involved?).
2. Is the thrombus bland thrombus or tumor thrombus?

**Answer**

1. Treatment options depend on disease severity and include medical management with anticoagulation therapy, mechanical recanalization of hepatic veins, transjugular intrahepatic portosystemic shunt (TIPS) creation, and liver transplantation. For patients with less severe disease, medical management options include managing underlying conditions, preventing further venous thrombosis with anticoagulant therapy, and management of ascites. More severe disease may necessitate attempts at hepatic vein recanalization (e.g., thrombolysis with stent placement). If attempts at recanalization fail, the patient may benefit from a TIPS or may ultimately require a liver transplant.

**REFERENCES**

**CASE 3.3**

**CLINICAL HISTORY** 41-year-old woman with liver lesion seen at ultrasound performed because of elevated liver function tests. MR performed for further evaluation.

**Figure 3.3A**

**Figure 3.3B**

**Figure 3.3C**

**Figure 3.3D**

**Figure 3.3E**

**Figure 3.3F**
**FINDINGS** Axial T2-weighted (A), T1-weighted precontrast (B), T1-weighted arterial phase (C), T1-weighted portal venous phase (D), and T1-weighted delayed phase (E) MR images through the upper abdomen demonstrate a 6-cm, lobulated left hepatic lobe lesion. A T2 bright central scar is visible (A). The lesion hyperenhances in the late arterial phase (C, 20-second delay), but is “stealth” or isointense to the surrounding liver parenchyma at T2-weighted (A), T1-weighted precontrast (B), venous phase (D, 70-second delay), and delayed (E, 3-minute delay) images. Delayed enhancement of the central scar is visible (E) (contrast agent: Gd-BOPTA [MultiHance; Bracco Diagnostics, Milan, Italy]). Color Doppler ultrasound image (F) demonstrates a hypoechoic mass with a central blood vessel.

**DIFFERENTIAL DIAGNOSIS** Focal nodular hyperplasia (FNH), hepatocellular adenoma, hepatocellular carcinoma, hemangioma, hypervascular metastasis.

**DIAGNOSIS** FNH.

**DISCUSSION** FNH is a benign hepatic tumor and is the second most common liver tumor following hemangioma. FNH is usually asymptomatic and has no malignant potential. Though the etiology of FNH is uncertain, a vascular malformation or vascular injury has been posited as the underlying event leading to the development of FNH.

At gross pathology, malformed vascular structures are visible within the central scar. Lobules of hepatic parenchyma surround the central scar and are separated by radiating fibrous bands. FNH does not have a capsule.

At MR imaging, FNH is most commonly iso- or mildly hyperintense on T2-weighted images and iso- or mildly hypointense on T1-weighted precontrast images. The central scar is frequently bright on T2-weighted images due to the presence of blood vessels, myxomatous elements, and biliary ductules. FNH avidly and homogeneously enhances in the arterial phase and becomes more isointense in the portal venous phase. The fibrous component of the central scar frequently demonstrates delayed contrast uptake. At 1- to 3-hour delayed images performed after administration of Gd-BOPTA, FNH appears iso- to hyperintense to the surrounding liver in more than 96% of cases.

At CT imaging, FNH is most commonly iso- to hypodense relative to the surrounding liver at precontrast imaging, avidly and homogeneously enhances in the arterial phase, and becomes more “stealth” or isoattenuating to the surrounding liver in the portal venous phase. As with MR, the fibrous component of the central scar may demonstrate delayed contrast uptake.

Ultrasound imaging features of FNH are nonspecific. At ultrasound, FNH is usually hypoechoic and may demonstrate a central blood vessel.

**Questions for Further Thought**
1. How can you differentiate this tumor from hepatocellular adenoma, hemangioma, and hepatocellular carcinoma?
2. What is the usual management of FNH?

**Reporting Responsibilities**
1. Describe the location and size of the lesion.
2. Attempt to confidently characterize the lesion as FNH. When the diagnosis of FNH can be made with confidence at imaging, it is a “don’t touch” lesion that does not warrant biopsy or surgical resection.

**What the Treating Physician Needs to Know**
1. Whether you are confident that the lesion is an FNH.
2. The size and location of the lesion.

**Answers**
1. Helpful discriminators include the above-described and illustrated enhancement pattern, absence of a capsule (both hepatocellular carcinoma and adenoma frequently demonstrate capsules), and normal background liver parenchyma (hepatocellular carcinoma most frequently occurs in cirrhotic livers). The presence of a central scar is a nonspecific finding as T2 bright central scars can be seen in hemangiomas (but hemangiomas classically demonstrate peripheral nodular enhancement with centripetal fill-in rather than homogeneous enhancement) and fibrolamellar hepatocellular carcinomas (scar is often dark on T1 and T2).
2. As these tumors have no malignant potential, they are not resected. If the diagnosis of FNH can be made with confidence at imaging, biopsy is not required.

**REFERENCES**
CLINICAL HISTORY  80-year-old man with hepatitis B and serum α-fetoprotein (AFP) level of 710 ng/mL.
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with hemangiomas. FNH can be eliminated as the mass is not “stealth” in the T2-weighted image. Both hepatocellular adenoma and hepatocellular carcinoma may contain visible intracellular lipid though the patient demographics are usually quite different. Hepatic adenomas are rare tumors occurring usually in young women taking oral contraceptive pills, individuals with a glycogen storage disease, or individuals using anabolic steroids. Hepatocellular carcinomas tend to occur in patients with chronic liver disease like this patient.

Hepatocellular cancers are often bright at T2-weighted imaging. Fat-containing hepatocellular cancers will lose signal at out-of-phase imaging. About 80% to 90% of hepatocellular carcinomas are hypervascular and hyperenhancing relative to the adjacent hepatic parenchyma in the arterial phase of imaging. About 10% to 20% of hepatocellular carcinomas are relatively hypoenhancing at arterial phase imaging. Hepatocellular carcinomas demonstrate a capsule at histologic evaluation in 65% to 82% of cases, and washout with a capsule is often seen at imaging.

Questions for Further Thought
1. What conditions can result in an elevated AFP level?
2. Can the AFP level be normal in patients with hepatocellular cancer?
Reporting Responsibilities

1. Describe the size, number, and location of lesions that are compatible with hepatocellular cancer.
2. Notify the ordering clinician if the finding is unexpected.

What the Treating Physician Needs to Know

1. In patients with imaging features compatible with hepatocellular cancer (as in this case), biopsy is NOT recommended to confirm the diagnosis as biopsy may result in track seeding.
2. If the patient is suitable for liver transplant, the patient should be referred to a transplant center as liver transplant is the preferred treatment for patients with cirrhosis and hepatocellular cancer who satisfy transplant criteria (a single tumor ≤5 cm in size or not more than three tumors ≤3 cm in size).

Answers

1. A variety of tumors that can produce elevated serum AFP levels including cancers of the pancreas, stomach, and biliary tree. Liver inflammation due to chronic liver disease can also result in an elevated AFP that increases with increasing inflammation.
2. Up to 30% of patients with hepatocellular cancer may have a normal AFP at the time of diagnosis. An AFP level >400 to 500 ng/mL is thought to be diagnostic of hepatocellular cancer.

REFERENCES

CASE 3.5

CLINICAL HISTORY 56-year-old man with abdominal pain.

FINDINGS T1- (A) and T2-weighted (B) MR images demonstrate a T1 dark mass in the right hepatic lobe with intermediate-to-bright, heterogeneous T2 signal. Postcontrast MR images in the arterial (C) and portal venous (D) phase as well as 3-minute delayed (E) MR image demonstrate progressive enhancement of the mass (contrast agent: Gd-BOPTA [MultiHance; Bracco Diagnostics, Milan, Italy]).

DIFFERENTIAL DIAGNOSIS Metastatic disease, hepatocellular carcinoma, intrahepatic cholangiocarcinoma.

DIAGNOSIS Intrahepatic cholangiocarcinoma.

DISCUSSION Cholangiocarcinoma is the second most common primary liver cancer following hepatocellular carcinoma. Cholangiocarcinomas are categorized based on location as extrahepatic, intrahepatic hilar, and intrahepatic peripheral.
At MR and CT imaging, intrahepatic cholangiocarcinomas demonstrate progressive enhancement at delayed imaging as seen in this case due to their fibrous nature. Progressive enhancement at delayed image is thought to reflect contrast material diffusing into the tumor interstitium.

By comparison, hepatocellular carcinomas classically demonstrate hyperenhancement in the arterial phase and washout at delayed imaging. An additional helpful discriminator is that cholangiocarcinomas tend to constrict adjacent vessels, whereas hepatocellular carcinomas more commonly invade adjacent vessels. Also, intrahepatic cholangiocarcinomas are often associated with more peripheral biliary ductal dilatation, whereas hepatocellular carcinomas are not. Metastatic disease could have a similar appearance to the above case, and percutaneous tissue sampling may be required to establish a diagnosis.

Questions for Further Thought
1. What are risk factors for cholangiocarcinoma?
2. What tumor markers are elevated in patients with cholangiocarcinoma?

Reporting Responsibility
1. Suggest cholangiocarcinoma in the differential diagnosis of an intrahepatic solid mass that demonstrates progressive enhancement at delayed imaging.

What the Treating Physician Needs to Know
1. Tumor size, location, and involvement of any major vascular structures.

Answers
1. Risk factors for cholangiocarcinoma include PSC, choledochal cysts, and liver fluke infestation.
2. Carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA 19-9) are elevated in patients with cholangiocarcinomas but can also be elevated due to other etiologies. For example, CEA can be elevated in patients with colorectal cancer or cancers of the pancreas, lung, thyroid, or breast. CEA may also be elevated in the setting of infection and inflammatory bowel disease. CA 19-9 may also be elevated in the setting of pancreatic cancer, colorectal cancer, and gastric cancer. Nonmalignant obstructive jaundice can also result in an elevated CA 19-9.

REFERENCES
CASE 3.6

CLINICAL HISTORY 51-year-old man with pain.

Figure 3.6A

Figure 3.6B

Figure 3.6C

Figure 3.6D

Figure 3.6E

Figure 3.6F
Questions for Further Thought
1. What is the usual treatment for non-Hodgkin lymphoma?
2. How is the diagnosis of primary hepatic lymphoma usually made?

Reporting Responsibilities
1. Describe the size and location of the liver lesion(s).
2. Describe whether there are findings of extrahepatic disease.

What the Treating Physician Needs to Know
1. Whether the liver lesion(s) is an isolated finding or if there is other extranodal disease.
2. If the liver lesion is an isolated finding and demonstrates nonspecific imaging features, tissue sampling will likely be needed to make the diagnosis.

Answers
1. Chemotherapy is the primary treatment for non-Hodgkin lymphoma.
2. As the imaging appearance of primary hepatic lymphoma is nonspecific, tissue sampling is usually required to make the diagnosis.

REFERENCES
CASE 3.7

CLINICAL HISTORY 45-year-old woman with abdominal pain.

Figure 3.7A

Figure 3.7B

Figure 3.7C

Figure 3.7D

Figure 3.7E

Figure 3.7F
FINDINGS Numerous (>20) well-circumscribed T2 bright (A) and T1 dark (B) structures are present in the liver and replace more than 50% of the hepatic parenchyma. No enhancement was seen in these structures after administration of intravenous contrast material (C). The patient also has numerous renal cysts, best seen in the coronal T2-weighted image (D). Note that some of the renal and liver lesions contain intrinsic T1 bright material reflecting small-volume blood products or proteinaceous debris (B). Numerous nonenhancing structures are also present at contrast-enhanced CT (E). Ultrasound (F) demonstrates anechoic structures with increased through transmission and no internal blood flow.

DIFFERENTIAL DIAGNOSIS Abscesses, cystic liver metastases, polycystic liver disease.

DIAGNOSIS Polycystic liver disease associated with autosomal-dominant polycystic kidney disease (ADPCKD).

DISCUSSION Polycystic liver disease most commonly occurs in association with ADPCKD but may also occur in isolation due to a separate genetic mutation that results in autosomal-dominant polycystic liver disease. Polycystic liver disease occurs in approximately 30% to 70% of patients with ADPCKD due to mutations on chromosomes 4 and 16. The genetic mutations that result in isolated polycystic liver disease have been isolated to chromosomes 6 and 19.

The cysts in polycystic liver disease are bile duct hamartomas that are lined with biliary epithelium but have no connection to the biliary system. Over time, the epithelial lining secretes fluid and the cysts progressively enlarge.

Most patients with polycystic liver disease are asymptomatic, and liver function tests are usually normal. However, a minority of patients will develop massive hepatomegaly and secondary symptoms of abdominal pain, distention, and early satiety or dyspnea due to mass effect from the massively enlarged liver. Rarely, liver cysts may hemorrhage or become infected.

At MR, the cysts associated with polycystic liver disease appear as well-circumscribed T2 bright structures. T1 signal will vary depending on cyst content. Cysts are usually dark on T1-weighted imaging but may demonstrate intrinsic T1 bright signal if they contain blood products or proteinaceous debris. If a cyst becomes infected, it may demonstrate perilesional edema and perilesional enhancement. At CT, polycystic liver disease appears as multiple well-circumscribed and non-enhancing cysts. At ultrasound, cysts associated with polycystic liver disease appear as anechoic (black) structures with increased through transmission and no internal blood flow.

Abscesses are not as homogeneous in signal intensity, may demonstrate adjacent edema, and often demonstrate abnormal enhancement such as internal septations or peripheral enhancement. Cystic liver metastases usually can be distinguished from polycystic liver disease as cystic metastases usually also have a soft-tissue component and will demonstrate areas of enhancement after administration of intravenous contrast material.

Questions for Further Thought
1. What are treatment options for patients with polycystic liver disease?
2. How many cysts are necessary for a diagnosis of polycystic liver disease?

REPORTING REQUIREMENTS
1. Describe the size and distribution of cysts with reference measurements of some of the larger cysts.
2. Report if the enlarged liver appears to be causing mass effect on adjacent structures such as bowel and diaphragm.
3. Report if cysts contain internal blood products or evidence of infection.

What the Treating Physician Needs to Know
1. If the patient has massive hepatomegaly with mass effect on adjacent structures.
2. If a cyst appears to contain new hemorrhage or be infected as either scenario could be a cause of patient symptoms.

Answers
1. No known medical therapy will halt cyst enlargement. Aspiration of a dominant cyst may provide brief partial symptomatic relief, but fluid within the cyst will reaccumulate. Surgical unroofing (also known as fenestration) of a cyst may be performed whereby the operating surgeon will attempt to remove as much of the cyst wall as possible. In rare cases, liver transplant may be performed for patients with severe refractory symptoms.
2. Different sources suggest different definitions of the number of cysts necessary for a diagnosis of polycystic liver disease. In general, more than 20 cysts that replace a substantial portion of the liver parenchyma should be present before a diagnosis of polycystic liver disease is invoked. Patients with fewer scattered liver cysts that occupy a small percentage of total hepatic parenchyma should not be diagnosed with polycystic liver disease based on imaging.

REFERENCES
CASE 3.8

CLINICAL HISTORY 38-year-old woman with hyperechoic liver lesion seen at ultrasound.

Figure 3.8A

Figure 3.8B

Figure 3.8C

Figure 3.8D

Figure 3.8E

Figure 3.8F
At ultrasound, hemangiomas sometimes appear hyper-echoic with increased through transmission and sometimes appear hypoechoic. However, malignant tumors also can appear hyperechoic at ultrasound. CT or MR should be recommended to further characterize indeterminate liver lesions identified at ultrasound. The blood flow in hemangiomas is usually too slow to be detectable at color Doppler or power Doppler ultrasound. If blood flow is detected at ultrasound, the lesion in question is most likely not a hemangioma.

Questions for Further Thought
1. What is Kasabach-Merritt syndrome?
2. What is the characteristic enhancement pattern of flash-filling hemangiomas.

Reporting Responsibilities
1. When hemangiomas demonstrate a classic appearance as above, a hemangioma can be confidently diagnosed at imaging.
2. In the setting of extremely large hemangiomas, report if there is mass effect on adjacent organs.

What the Treating Physician Needs to Know
1. Hemangiomas are benign lesions and generally require no further follow-up or treatment.
2. Hemangiomas can be confidently diagnosed at contrast-enhanced cross-sectional imaging and do not require biopsy.

Answers
1. Kasabach-Merritt syndrome is also known as hemangioma thrombocytopenia syndrome. This syndrome occurs when platelets become sequestered in a vascular tumor such as a hemangioma resulting in a consumptive coagulopathy that can ultimately progress to disseminated intravascular coagulation. Deaths have been reported. Kasabach-Merritt syndrome more commonly occurs in pediatric patients with hemangioendotheliomas but has been described in association with hemangiomas.
2. Flash-filling hemangiomas are small lesions that demonstrate marked arterial hyperenhancement in the arterial phase and are usually isointense to liver in the portal venous phase of enhancement.

REFERENCES
**CASE 3.9**

**CLINICAL HISTORY** 43-year-old woman with abnormal liver function tests.

**FINDINGS** Grayscale ultrasound (A) demonstrates an echogenic liver with a geographic hypoechoic area along the porta hepatis. No definite blood flow is visible in this area at color Doppler imaging (B, calipers). In-phase (C) and out-of-phase (D) MR demonstrate diffuse hepatic signal loss in the out-of-phase image with the exception of the hepatic parenchyma around the porta hepatis.

**DIFFERENTIAL DIAGNOSIS** Hepatic steatosis with fat sparing; hepatic steatosis with focal fat deposition.

**DIAGNOSIS** Hepatic steatosis with fat sparing.

**DISCUSSION** Histologically, fatty liver is characterized by the accumulation of triglycerides within hepatocyte cells. At
Questions for Further Thought
1. List conditions associated with NASH.
2. How can you identify the out-of-phase image?

Reporting Requirements
1. This patient has diffuse hepatic steatosis.
2. The geographic hypoechoic area seen at ultrasound reflects an area of fat sparing.

What the Treating Physician Needs to Know
1. Further evaluation to determine the etiology of hepatic steatosis is warranted.
2. If untreated, steatosis can progress to steatohepatitis and ultimately hepatic fibrosis.

Answers
1. Obesity, type 2 diabetes, high cholesterol, elevated triglycerides, and metabolic syndrome are associated with NASH.
2. The out-of-phase images can be identified by noting the “India ink artifact” (also known as chemical shift artifact of the second kind). The India ink artifact describes the solid black line at all fat–water interfaces that looks as if it were drawn with India ink.

REFERENCE
CLINICAL HISTORY 53-year-old man with chronic liver disease undergoing screening for hepatocellular cancer.
**FINDINGS** T1-weighted MR (A) demonstrates a small, nodular liver in keeping with changes of advanced chronic liver disease. Notice also the geographic area of low signal with linear margins involving primarily segments 4A and 8 near the inferior vena cava. T2-weighted MR demonstrates corresponding T2 bright signal abnormality in segments 4A (B) and 8 (not shown). Dynamic contrast-enhanced MR demonstrates this area to be hypoenhancing relative to adjacent hepatic parenchyma in the arterial phase (C) of enhancement, hyperenhancing in the portal venous phase (D), and extremely hyperenhancing in the 3-minute delayed image (E) (contrast agent: Gd-BOPTA [MultiHance; Bracco Diagnostics, Milan, Italy]). Noncontrast CT (F) demonstrates a corresponding area of low attenuation.

**DIFFERENTIAL DIAGNOSIS** Confluent hepatic fibrosis.

**DIAGNOSIS** Confluent hepatic fibrosis.

**DISCUSSION** Hepatic fibrosis occurs in response to liver injury such as chronic inflammation due to hepatitis. Liver fibrosis is histologically defined as the deposition of substances including collagen and proteoglycans in the extracellular matrix.

The phrase “confluent fibrosis” refers to large, mass-like areas of fibrosis. Confluent fibrosis most commonly involves the anterior segment right hepatic lobe and medial segment left hepatic lobe.

At MR, areas of fibrosis usually demonstrate low T1 signal and high T2 signal due to the presence of water within areas of fibrosis. Areas of fibrosis exhibit delayed hyperenhancement at both CT and MR due to accumulation of gadolinium in the extracellular compartment. Areas of fibrosis can be thought of as hanging on to contrast material while contrast material washes out from the remainder of the liver. Delayed hyperenhancement in areas of confluent hepatic fibrosis is usually relatively homogeneous.

Confluent hepatic fibrosis can be distinguished from a neoplasm by its geographic morphology often with linear borders, associated capsular retraction, relatively homogeneous delayed hyperenhancement, and progressive volume loss over time.

**Questions for Further Thought**
1. What intrahepatic malignancy contains fibrous tissue and may demonstrate delayed hyperenhancement?
2. List other fibrotic conditions elsewhere in the body that demonstrate delayed hyperenhancement.

**Reporting Requirements**
1. Describe the presence of confluent hepatic fibrosis.
2. Describe any sequelae of portal hypertension such as splenomegaly, varices, and ascites.
3. Careful attention should be paid to evaluate for hepatocellular cancer in patients with findings of chronic liver disease.

**What the Treating Physician Needs to Know**
1. That the patient has areas of confluent hepatic fibrosis.
2. If the patient has findings of portal hypertension or evidence of hepatocellular cancer.

**Answers**
1. Intrahepatic cholangiocarcinomas often demonstrate delayed hyperenhancement at contrast-enhanced MR due to the presence of fibrous tissue. Intrahepatic cholangiocarcinomas can be distinguished from areas of confluent fibrosis based on morphology. For example, intrahepatic cholangiocarcinomas usually have infiltrating or rounded borders, whereas confluent hepatic fibrosis is frequently geographic in shape with linear borders. Though intrahepatic cholangiocarcinomas often show delayed hyperenhancement, the enhancement is more heterogeneous than is seen with confluent hepatic fibrosis. Additionally, intrahepatic cholangiocarcinomas are often associated with biliary ductal dilatation more peripherally, whereas areas of confluent hepatic fibrosis are not.
2. Retroperitoneal fibrosis and mesenteric fibrosis can also demonstrate delayed hyperenhancement due to the presence of fibrous tissue.

**REFERENCES**
**CASE 3.11**

**CLINICAL HISTORY** 68-year-old man with abnormal liver function tests 3 months after liver transplant.

**FINDINGS** Color Doppler ultrasound demonstrates heterogeneous hepatic parenchyma (A) and patent transplant vasculature (not shown). Patient next underwent MR imaging to better evaluate the liver parenchyma. T2-weighted MR (B) demonstrates innumerable 1- to 2-cm T2 bright liver lesions that are T1 dark (C) and hypoenhancing (D) (contrast agent: Gd-BOPTA [MultiHance; Bracco Diagnostics, Milan, Italy]).

**DIFFERENTIAL DIAGNOSIS** Abscesses, post-transplant lymphoproliferative disorder (PTLD), metastatic disease.

**DIAGNOSIS** PTLD.

**DISCUSSION** PTLD occurs as a complication of organ transplantation due to immunosuppression. A majority of cases appear to be related to Epstein-Barr virus. PTLD is a general term that describes a spectrum of illnesses. At the milder end of the spectrum, some patients will manifest an acute mononucleosis-type illness. At the more severe end of the spectrum, patients develop a monoclonal proliferation of lymphoid cells that meet diagnostic criteria for lymphoma. Liver biopsy was performed in the above patient, and pathology revealed B-cell lymphoma.

PTLD is uncommon occurring in approximately 1% to 2% of liver transplant patients. PTLD is slightly more common in other solid organ transplant patients (e.g., kidney and...
lung) and occurs in up to a third of patients who undergo bowel transplant or multiple organ transplants.

Of patients who develop PTLD, a majority of patients will develop PTLD in the first year after transplant when immunosuppression is most intense. At imaging, patients with PTLD may present with solid organ lesions or lymphadenopathy with extranodal disease being more common. PTLD preferentially involves the transplanted organ. Central nervous system and gastrointestinal tract involvement also may occur. The mainstay of treatment of PTLD is cessation or decrease in immunosuppressive medications.

Abscesses could have a similar appearance at imaging, though fever and elevated white blood cell count would be expected. Diffuse metastatic disease could have a similar appearance. Tissue sampling is usually required to make the diagnosis of PTLD.

Questions for Further Thought
1. What is the typical clinical presentation of PTLD?
2. What are the most common locations of gastrointestinal involvement with PTLD?

Reporting Requirement
1. If solid organ lesions or marked lymphadenopathy are seen in a transplant recipient, suggest the possibility of PTLD.

What the Treating Physician Needs to Know
1. Solid organ lesions or lymphadenopathy could reflect PTLD in a transplant recipient.
2. Tissue sampling will likely be needed to confirm the diagnosis.

Answers
1. Clinical presentation of PTLD can be quite variable. Many patients are initially asymptomatic. However, given the propensity of PTLD to involve the transplanted organ or the gastrointestinal tract, evidence of graft dysfunction or diarrhea could reflect PTLD.
2. Distal small bowel and proximal colon are the most commonly affected bowel segments. PTLD appears similar to nontransplant-related lymphoma and may manifest as segmental bowel wall thickening with aneurysmal dilatation.

REFERENCES
CLINICAL HISTORY 67-year-old woman with right upper quadrant pain.

Figure 3.12A

Figure 3.12B

Figure 3.12C

Figure 3.12D

Figure 3.12E

Figure 3.12F
Case 3.12

**FINDINGS** Slab image from MRCP (A) demonstrates innumerable T2 dark-filling defects within the gallbladder, central intrahepatic biliary ducts, and in the extrahepatic bile duct. These filling defects are confirmed in the coronal (B) and axial T2-weighted images (C) and demonstrate low signal in the T1 precontrast (D) and True FISP images (E). At CT, these structures appear as well-circumscribed areas of increased attenuation (F).

**DIFFERENTIAL DIAGNOSIS** Air bubbles, gallstones, flow voids.

**DIAGNOSIS** Gallstones.

**DISCUSSION** The differential diagnosis of multiple areas of low T2 signal within the biliary system includes air bubbles, gallstones, and flow voids. The rounded shape and dependent location of the above intraluminal structures are diagnostic of gallstones.

Gallstones may contain cholesterol, glycoproteins, and/or calcium bilirubinate. All stones usually appear dark at T2-weighted imaging. Cholesterol stones also appear dark at T1-weighted imaging, whereas pigment stones appear bright at T1-weighted imaging.

A way to evaluate for the presence of air in the biliary system is to compare signal intensity with in- and out-of-phase images. Air will “bloom” or appear darker and larger in the in-phase images if in-phase images are obtained with a longer TE. The reason for this finding is that air produces magnetic susceptibility artifact, and this artifact increases with longer TE.

A way to distinguish flow voids from gallstones is to look at the True FISP images. True FISP images are relatively insensitive to motion. Flow voids are usually not visible at True FISP imaging. Therefore, if focal low signal is visible within the biliary system on the True FISP images it is most likely a gallstone.

MRCP is a heavily T2-weighted MR technique whereby fluid-containing structures demonstrate high signal (appear bright). Note in the above case how fluid in the biliary system and pancreatic duct is bright. Note also the bright fluid in the lumen of the duodenum located just inferior to the biliary system. Fluid is also visible within jejunal loops in the left abdomen as well as ileal loops in the right lower quadrant.

**Question for Further Thought**
1. How sensitive are radiographs, ultrasound, CT, and MR for the detection of gallstones?

**Reporting Requirements**
1. Report the presence of innumerable stones filling the gallbladder and biliary system.
2. Evaluate for complications such as cholecystitis or pancreatitis.

**What the Treating Physician Needs to Know**
1. This patient has innumerable gallstones filling the biliary system.
2. There is no evidence of acute cholecystitis or pancreatitis.

**Answer**
1. Abdominal radiographs are approximately 15% to 20% sensitive for the detection of gallstones. Ultrasound is 95% sensitive for the detection of gallstones larger than 2 mm in size. CT is 75% sensitive for the detection of gallstones and is better able to detect calcium-containing stones when compared with cholesterol-containing stones. MR is >95% sensitive for the detection of gallstones.

**REFERENCES**
CASE 3.13

CLINICAL HISTORY 62-year-old man with right upper quadrant pain.

FINDINGS  MRCP slab image (A) demonstrates mild-to-moderate intrahepatic biliary ductal dilatation with abrupt termination just beyond the confluence of the right and left hepatic ducts. Note also the irregularity of the intrahepatic biliary system with areas of relative dilatation and stricturing.

Axial T1 MR (B) demonstrates circumferential low signal surrounding the extrahepatic bile duct just below the confluence of the right and left hepatic ducts. Corresponding T2 dark signal is present surrounding the T2 bright bile duct (C).

Postcontrast images demonstrate this material to be hypoenhancing in the arterial phase (D) and relatively hyperenhancing...
in the 3-minute delayed images (E). Note that this mass-like area is located along the right posterior aspect of the right hepatic artery (D) and along the right aspect of the main portal vein (E).

**DIFFERENTIAL DIAGNOSIS**
Stricture, cholangiocarcinoma.

**DIAGNOSIS**
Cholangiocarcinoma.

**DISCUSSION**
When measurable soft tissue is present at the site of abrupt bile duct termination or tapering as in the above case, cholangiocarcinoma is present until proven otherwise. Cholangiocarcinomas may be intrahepatic, perihilar, or extrahepatic in location. Perihilar is the most common location as in the above case.

The Bismuth classification is used to characterize perihilar cholangiocarcinomas. Type 1 tumors involve the common hepatic duct below the confluence of the right and left hepatic ducts. Type 2 tumors involve the confluence of the right and left hepatic ducts. Type 3a tumors are type 2 tumors that also extend to the bifurcation of the right hepatic duct. Type 3b tumors are type 2 tumors that extend to the bifurcation of the left hepatic duct. Type 4 tumors extend to the bifurcation of the right and left hepatic ducts. Type 5 tumors are located at the junction of the common bile duct and the cystic duct.

The above case was characterized as a Bismuth 2 lesion. The irregularity of the intrahepatic biliary system visible in the slab MRCP image reflects the patient’s underlying diagnosis of primary sclerosis cholangitis, a risk factor for cholangiocarcinoma.

Historically, surgical resection has been the treatment of choice for cholangiocarcinomas. The relationship of the tumor to major vascular structures such as the hepatic artery and portal vein strongly influences surgical decisions regarding resectability. More recently, liver transplants are being performed at some centers to treat hilar cholangiocarcinomas with positive outcomes.

**Questions for Further Thought**
1. What are Klatskin tumors?
2. How could tissue sampling be performed in the above case?

**Reporting Requirements**
1. Describe the presence of a hilar soft-tissue mass resulting in intrahepatic biliary ductal dilatation.
2. Describe the relationship of the mass to major vascular structures such as the hepatic artery and portal vein.
3. Evaluate for distant metastatic disease such as liver metastases.

**What the Treating Physician Needs to Know**
1. This patient has an obstructing hilar cholangiocarcinoma on a background of PSC.
2. The tumor abuts the right hepatic artery and the main portal vein.
3. No distant metastatic disease is visible.

**Answers**
1. Klatskin tumors are cholangiocarcinomas that occur at the confluence of the right and left hepatic ducts. Gerald Klatskin (1910 to 1986) was an American pathologist and hepatologist.
2. Tissue sampling could be performed via brushings with endoscopic retrograde cholangiopancreatography (ERCP). Brushings can also be performed during transhepatic cholangiopancreatography.

**REFERENCES**
CLINICAL HISTORY  53-year-old woman 3 years status post-liver transplant now with pruritus and elevated bilirubin.
FINDINGS Slab MRCP (A) demonstrates severe intrahepatic biliary ductal dilatation with peripheral pruning of the biliary tree. Multiple filling defects are identified within the intrahepatic biliary system. The common bile duct is decompressed. ERCP (B) performed for stent placement confirms marked irregularity of the intrahepatic biliary system with numerous rounded filling defects. These filling defects are dark at T2-weighted MR (C) and bright at T1-weighted MR (D). Arterial phase postcontrast images demonstrate the hepatic artery jump graft to be patent at its aortic origin (E). The graft occludes just beyond its origin (F).

DIFFERENTIAL DIAGNOSIS Anastomotic stricture, ischemic cholangiopathy.

DIAGNOSIS Ischemic cholangiopathy; also intraductal stones and sludge due to stasis.

DISCUSSION Biliary ductal irregularity and stricturing with an occluded hepatic artery or hepatic artery jump graft is diagnostic of ischemic cholangiopathy. The hepatic artery provides the sole blood supply to the biliary system of the transplanted liver. Hepatic artery occlusion consequently results in bile duct ischemia, necrosis, and eventual stricturing.

Biliary complications are the second most common cause of liver transplant failure and occur in approximately 15% of liver transplant recipients. When evaluating a post-liver transplant patient at ultrasound or cross-sectional imaging, it is critically important to evaluate the patency of the hepatic artery, portal vein, and hepatic veins. Failure to identify and address an occluded hepatic artery can result in ischemic cholangiopathy that may necessitate retransplantation.

Question for Further Thought
1. What is another potential biliary complication of liver transplant?

Reporting Requirements
1. Describe the presence of bile duct stricture and dilatation.
2. Evaluate the patency of the hepatic artery

What the Treating Physician Needs to Know
1. This patient has ischemic cholangiopathy due to an occluded arterial jump graft.

Answer
1. Bile duct leak is a potential acute complication of liver transplant. Chronically, liver transplant patients may also develop focal strictures at the biliary anastomosis. Anastomotic strictures are often due to localized ischemia or scarring rather than proximal hepatic arterial occlusion.

REFERENCES
CASE 3.15

CLINICAL HISTORY 48-year-old woman with right upper quadrant pain.

Adenomyomatosis is characterized by gallbladder wall thickening, intramural diverticula, and deposition of cholesterol crystals in these diverticula. Fluid-filled diverticula correspond to the cystic spaces seen at MR. When multiple cystic spaces are visible in the gallbladder wall as in the above case, the appearance has been referred to as the “pearl necklace sign.”

The comet tail artifact seen at ultrasound is reverberation artifact seen posterior to cholesterol crystals in Rokitansky-Aschoff sinuses and is characterized by multiple parallel echogenic bands posterior to an object.

Adenomyomatosis may be diffuse, segmental, or focal. Segmental adenomyomatosis most commonly occurs in the gallbladder body and results in wasting with an hourglass appearance. Focal adenomyomatosis most commonly occurs at the gallbladder fundus. Focal or segmental adenomyomatosis can sometimes be difficult to distinguish from gallbladder cancer at ultrasound. If MR is not definitive, fluorine 18 fluorodeoxyglucose positron emission tomography can be used to distinguish the two entities as gallbladder cancer will appear hypermetabolic while focal adenomyomatosis will not.

Questions for Further Thought
1. What is cholesterolosis?
2. What symptoms may be associated with adenomyomatosis?
3. What is the usual treatment of adenomyomatosis?

Reporting Requirements
1. Describe the presence of diffuse adenomyomatosis.

FINDINGS Grayscale ultrasound images (A, B) demonstrate diffuse gallbladder wall thickening. Note the echogenic material with parallel bands of increased echogenicity posteriorly (“comet tail” artifact). Note the area of cystic change in the gallbladder wall as seen at MRCP (C).

DIFFERENTIAL DIAGNOSIS Adenomyomatosis, emphysematous cholecystitis.

DIAGNOSIS Adenomyomatosis.

DISCUSSION The comet tail artifact seen at ultrasound and cystic spaces seen in the gallbladder wall at MRCP are characteristics of adenomyomatosis.
What the Treating Physician Needs to Know

1. Adenomyomatosis alone is usually not considered an indication for cholecystectomy.

Answers

1. Though the terms “adenomyomatosis” and “cholesterosis” are sometimes used interchangeably, technically these terms refer to two different and distinct conditions. Adenomyomatosis refers to deposition of cholesterol crystals within Rokitansky-Aschoff sinuses in the gallbladder wall with corresponding wall thickening seen at ultrasound. By comparison, cholesterosis refers to cholesterol crystal deposition along the lamina propria of the gallbladder and is usually not associated with gallbladder wall thickening.
2. Patients with adenomyomatosis may be asymptomatic or have vague abdominal pain.
3. Adenomyomatosis is thought to have no malignant potential and therefore requires no further treatment.

REFERENCES

**CLINICAL HISTORY** 62-year-old woman with vague right upper quadrant pain.

**FINDINGS** Color Doppler ultrasound (A) demonstrates a 20-cm well-circumscribed hypoechoic structure with layering debris but no definite internal blood flow. T1-weighted MR image obtained 60 seconds after administration of intravenous contrast material (B) demonstrates rim enhancement around the area of fluid. Note that fluid is contiguous with the gallbladder lumen through an approximately 3-cm gallbladder wall defect. A 1-cm T1 dark stone is present in the gallbladder lumen. T2-weighted MR without (C) and with (D) fat saturation demonstrate primarily T2 bright fluid with layering-dependent relatively lower signal debris. Note also T2 dark gallstone. (Incidentally noted 3.5-cm cyst exophytic off the upper pole right kidney.)

**DIFFERENTIAL DIAGNOSIS** Perforated acute cholecystitis, remote prior gallbladder perforation.

**DIAGNOSIS** Contained remote prior gallbladder perforation.

**DISCUSSION** T2-weighted images with fat saturation are crucial when assessing for the presence or absence of acute inflammation. Acute inflammation is indicated by T2 bright signal reflecting fluid. Such signal is much more conspicuous against a background of dark fat in T2-weighted images obtained with fat saturation. The absence of T2 bright inflammatory changes around the above fluid collection in the T2 with fat saturation image indicates that the abnormality is chronic rather than acute.

The above patient had a history of acute cholecystitis approximately 6 weeks prior to the above study and was pain free at the time of the above study. The patient underwent
percutaneous drainage of the above fluid collection and later underwent elective cholecystectomy.

Gallbladder perforation is thought to result when intraluminal pressure increases due to outflow obstruction. This increased intraluminal pressure results in impaired blood flow, tissue necrosis, and eventual tissue breakdown.

Question for Further Thought
1. What portion of the gallbladder most commonly perforates?

Reporting Requirements
1. Describe the presence of a large fluid collection contiguous with the gallbladder lumen.
2. Assess for acute inflammatory changes.

What the Treating Physician Needs to Know
1. This patient has a large fluid collection likely reflecting a remote prior gallbladder perforation.
2. No significant acute inflammatory changes are visible about the collection.

Answer
1. The gallbladder fundus is the most common site of perforation, possibly due to a tenuous blood supply in this location.

REFERENCE
**CASE 3.17**

**CLINICAL HISTORY** 26-year-old woman with history of abdominal pain.

**DISCUSSION** SPT of the pancreas is a rare neoplasm that accounts for 1% to 2% of pancreatic exocrine malignancies. These tumors most commonly occur in non-Caucasian women in the second or third decade of life.

SPTs most frequently occur in the pancreatic head or tail and appear as large well-circumscribed cystic masses with peripheral solid components as in the above case. However, when small these tumors may appear entirely solid.

These tumors usually exhibit benign behavior. However, since up to 15% of SPTs undergo malignant degeneration all tumors thought to be SPTs are usually resected.

The tumor gets its name from its distinct appearance at microscopy where cells are arranged in solid and papillary configurations.

The well-circumscribed nature of SPTs distinguishes them from the more ill-defined, infiltrating appearance typical of pancreatic ductal adenocarcinomas. The imaging appearance of SPTs can be similar to acinar cell carcinomas as acinar cell tumors are also well-circumscribed tumors that may appear solid when small and contain large cystic components when large. However, SPTs most commonly occur in young women, whereas pancreatic acinar carcinomas most commonly occur in patients in the fifth through seventh decades of life. Small SPTs can be difficult to distinguish from neuroendocrine tumors. The peripheral papillary solid projections seen in large primarily cystic SPTs is atypical for neuroendocrine tumors.

**FINDINGS** MR image demonstrates a 15-cm primarily cystic mass with small, peripheral solid components in the region of the pancreatic head. The cystic components demonstrate T1 dark signal (A) and T2 bright signal (C). The peripheral solid components enhance at postcontrast imaging (B). No enlarged lymph nodes or distant metastatic disease were seen.

**DIFFERENTIAL DIAGNOSIS** Pancreatic adenocarcinoma, neuroendocrine tumor, acinar cell carcinoma, solid pseudo-papillary tumor (SPT).

**DIAGNOSIS** SPT of the pancreas.
Questions for Further Thought
1. What lab abnormalities are associated with SPT?
2. What is the usual treatment for SPT?

Reporting Responsibilities
1. Describe the size and location of the mass.
2. Evaluate for nodal or other metastatic disease.

What the Treating Physician Needs to Know
1. That an SPT is favored rather than a ductal adenocarcinoma as this information may alter treatment planning (see Question 2).

Answers
1. None. Pancreatic cancer markers (CA19-9, AFP, CEA) are normal in patients with SPT.

2. Surgical resection is usually curative. At the present time, there is no role for neoadjuvant chemotherapy in these patients. By comparison, neoadjuvant chemotherapy may be administered to patients with pancreatic ductal adenocarcinomas. Therefore, tissue sampling may be performed prior to resection to determine if neoadjuvant chemotherapy is warranted in questionable cases.

REFERENCES
44-year-old woman with abdominal pain.
IgG4 sclerosing disease can also involve a variety of other structures including the biliary system, kidneys, and retroperitoneum. Specifically, segmental biliary strictures may be visible along with hypoenhancing areas in the kidneys (E, F) and retroperitoneal fibrosis. Though not present in every case, identification of these other findings can help in distinguishing focal autoimmune pancreatitis from pancreatic adenocarcinoma.

Questions for Further Thought
1. What laboratory abnormalities may be present in patients with autoimmune pancreatitis?
2. What is the usual treatment of autoimmune pancreatitis?

Reporting Requirements
1. Report that the appearance of the pancreas is most suggestive of autoimmune pancreatitis.
2. Evaluate for involvement of other organs such as hypoenhancing renal lesions and biliary strictures.

What the Treating Physician Needs to Know
1. This patient most likely has autoimmune pancreatitis.

Answers
1. Patients with autoimmune pancreatitis often have elevated IgG4 antibody levels. However, some patients with pancreatic adenocarcinoma may also have elevated IgG4 antibody levels.
2. Patients with autoimmune pancreatitis are usually treated with steroids. A pancreatic mass due to autoimmune pancreatitis would be expected to decrease in size during the course of steroid treatment.

REFERENCES
**CASE 3.19**

**CLINICAL HISTORY** 31-year-old pregnant woman with right lower quadrant pain.

**FINDINGS** Axial (A) and coronal (C) T2-weighted MR demonstrate the appendix located in the right lower quadrant arising from the base of cecum. Axial (B) and coronal (D) T2-weighted MR with fat saturation demonstrate marked T2 bright signal surrounding the appendix.

**DIFFERENTIAL DIAGNOSIS** Normal appendix, acute appendicitis.

**DIAGNOSIS** Acute appendicitis.

**DISCUSSION** Noncontrast MR is the preferred modality to evaluate for acute appendicitis in pregnant women. MR is preferred over CT because MR does not expose the fetus to ionizing radiation. Contrast material should not be administered to pregnant women as gadolinium-based contrast agents are excreted into the amniotic fluid thereby subjecting the fetus to prolonged exposure.

A satisfactory MR protocol to evaluate for acute appendicitis in pregnant women includes coronal, sagittal, and axial...
T2-weighted images; coronal and axial T2-weighted images with fat saturation; axial True FISP images; axial in- and out-of-phase images; and axial and coronal T1-weighted images.

Multiplanar T2-weighted images are used to identify the appendix. T2-weighted images with fat saturation are key to evaluate for periappendiceal inflammation. The periappendiceal inflammation visible in the above case (B, D) is diagnostic of acute appendicitis. Without fat saturation (A, C), bright signal indicating fluid and inflammation can be difficult or impossible to identify on a background of T2 bright fat. Note how periappendiceal inflammation is much more obvious in the T2-weighted images with fat saturation (B, D) when compared with the T2-weighted images without fat saturation (A, C).

True FISP images serve as a backup sequence to identify the appendix as True FISP images are often the least sensitive to motion. T1-weighted images are most helpful to evaluate for other causes of right lower quadrant pain such as a hemorrhagic cyst that would contain T1 bright material. In- and out-of-phase images can be used to evaluate for extraluminal air which would be expected to “bloom” (appear darker and larger) in the in-phase images with longer TE due to increased susceptibility artifacts with longer TE.

As with CT, findings of perforation at MR include periappendiceal fluid collections and extraluminal air.

**Question for Further Thought**
1. Are there any risks to a fetus undergoing MR imaging?

**Reporting Requirements**
1. Report that the patient has acute appendicitis.
2. Report whether or not the patient has evidence of perforation.

**What the Treating Physician Needs to Know**
1. This patient has acute appendicitis.
2. There are no findings to indicate perforation.

**Answer**
1. The major risk of noncontrast MR is heat deposition in the fetus. No current literature indicates that this minimal heat deposition has deleterious effects on the fetus. The risks of performing MR in a pregnant patient should always be weighed against the potential benefit of the study and the potential risk of not performing the study. In general, the benefits of performing noncontrast MR to evaluate for acute appendicitis are thought to outweigh the risks at all stages of pregnancy.

**REFERENCE**
CASE 3.20

CLINICAL HISTORY 51-year-old man with generalized abdominal pain.

FINDINGS T2-weighted MR without (A) and with (B) fat saturation demonstrate a 2.7 cm in diameter T2 bright structure in the right lower quadrant. This structure was determined to be arising from the base of cecum in other images of the study. Pre- (C) and postcontrast (D) MR images demonstrate a small 3-mm-enhancing nodule along the medial wall of the structure. This enhancing nodule demonstrates low signal in the T2-weighted images (A, B). CT image from a different patient (E) with the same diagnosis demonstrates a distended appendix filled with low-attenuation material and no surrounding inflammatory changes.

DIFFERENTIAL DIAGNOSIS Normal fluid-containing appendix, appendiceal mucocele.

DIAGNOSIS Appendiceal mucocele due to mucinous cystadenoma.
**DISCUSSION**

Appendiceal mucocele is defined as an appendix distended with mucin. At CT, mucoceles appear as homogeneous low-attenuation material distending the appendix. Periappendiceal inflammatory changes usually are absent. At MR, mucoceles generally demonstrate homogeneously bright T2 signal. T1 signal will vary depending on whether appendiceal contents are mostly fluid (T1 dark) or whether blood products or proteinaceous debris are present (T1 bright signal).

Mucin may accumulate in the appendix due to outflow obstruction or due to a mucin-producing tumor such as a benign cystadenoma or a malignant cystadenocarcinoma. Both cystadenomas and cystadenocarcinomas can perforate resulting in mucinous material filling the abdomen and pelvis, a condition known as pseudomyxoma peritonei.

The enhancing small nodule in the above MR images is somewhat unusual as most often no enhancing solid component is seen. If an enhancing solid component is visible, the patient has a cystadenoma or cystadenocarcinoma.

**Question for Further Thought**

1. What is the usual management of an appendiceal mucocele?

**Reporting Requirements**

1. Report the presence of an appendiceal mucocele.
2. Report that the small soft-tissue nodule most likely indicates a cystadenoma or cystadenocarcinoma.

**What the Treating Physician Needs to Know**

1. This patient has an appendiceal mucocele most likely due to a cystadenoma or cystadenocarcinoma.

**Answer**

1. Appendiceal mucoceles are virtually always removed due to the risk of malignant transformation. If no soft-tissue component is visible, the mucocele will likely be removed via appendectomy. If there is suspicion for malignancy, a right hemicolectomy will be performed with removal of regional lymph nodes.

**REFERENCE**

CLINICAL HISTORY  61-year-old man with rectal cancer diagnosed at colonoscopy. MR performed for staging.

FINDINGS  High-resolution T2-weighted MR image through the pelvis demonstrates rectal wall thickening extending from approximately 7 o’clock to 1 o’clock. The mass extends beyond the serosal surface of the rectum as well as the mesorectal fascia from approximately 9 o’clock to 1 o’clock. Tumor invades the right posterior bladder wall.

DIFFERENTIAL DIAGNOSIS  T1–T4 rectal cancer.

DIAGNOSIS  T4 rectal cancer.

DISCUSSION  T staging of known rectal cancer is an increasingly common indication for MR imaging. T1 disease is defined as tumor that invades the submucosa. T2 disease invades the muscularis propria. T3 disease invades the perirectal fat. T4 disease extends beyond the mesorectal fascia to directly invade other structures.

T1 and T2 diseases can be difficult to distinguish at MR. The hallmark of T3 disease is extension beyond the serosal surface of the rectum. Normally, at T2 MR the rectum is outlined by a thin black line. Such a line is seen from approximately 1 o’clock to 6 o’clock in the above case. Disruption of this line indicates tumor extension beyond the serosal surface of the rectum.

The distance between the tumor and the mesorectal fascia is an important data point for the surgeon as patients whose tumors extend to within 5 mm of the mesorectal fascia may benefit from neoadjuvant chemoradiation therapy prior to tumor removal.

The mesorectal fascia appears as a thin low-signal band at T2 MR which completely encircles the perirectal fat and is located approximately 2 to 3 cm away from the rectum. In the above image, normal mesorectal fascia is visible from approximately 1 o’clock to 8 o’clock.

Tumors that do not extend beyond the mesorectal fascia are usually removed via total mesorectal excision which includes resection of the tumor as well as the mesorectal fat. On the other hand, tumors that extend to within 5 mm of the mesorectal fascia are usually treated with chemoradiation first.

Question for Further Thought
1. In addition to measuring the distance between the tumor and the mesorectal fascia, what other measurement is helpful for preoperative planning?

Reporting Requirements
1. Describe if the tumor extends beyond the serosal surface of the rectum.
2. Report the distance between the tumor and the mesorectal fascia.
3. Report if the tumor invades adjacent organs.

What the Treating Physician Needs to Know
1. The distance between the tumor and the mesorectal fascia.

Answer
1. The distance between the tumor and the levator ani musculature as this distance can impact preoperative planning. The amount of normal rectum between the tumor and the levator ani musculature informs the surgeon’s decision regarding whether a primary colonic anastomosis should be performed. For very low tumors, there may not be enough normal distal rectum left to perform a primary anastomosis and the patient may be left with a colostomy.

REFERENCE
CASE 3.22

CLINICAL HISTORY  25-year-old woman with history of familial adenomatous polyposis (FAP) syndrome.

FINDINGS  CT image (A) demonstrates a large soft-tissue mass centered in the small bowel mesentery resulting in tethering and distortion of small bowel loops as well as left-sided hydrenephrosis with a stent visible in the left ureter. T1-weighted MR imaging (B, precontrast; C 20-second delay [arterial phase]; and D, 3-minute delay) demonstrates heterogeneous, delayed hyperenhancement. The mass is primarily dark at T2-weighted imaging (E) (contrast agent: Gd-BOPTA [MultiHance; Bracco Diagnostics, Milan, Italy]).

DIFFERENTIAL DIAGNOSIS  Carcinoid tumor, desmoid tumor, lymphoma.

DIAGNOSIS  Desmoid tumor.

DISCUSSION  Desmoid tumors are composed of fibroblasts. These tumors most frequently involve the small bowel mesentery or the anterior abdominal wall where they may occur...
at surgical scar sites. Though histologically benign, these infiltrative tumors may result in significant morbidity or mortality due to involvement of bowel loops, ureters, and/or blood vessels.

Desmoid tumors are most commonly sporadic. A minority of desmoid tumors occur in individuals with FAP. About 9% to 18% of patients with FAP develop desmoid tumors.

At CT, these tumors often appear as an enhancing, soft-tissue mesenteric mass. At MR, the appearance of these tumors is variable with, for example, T2 signal ranging from hypo- to hyperintense depending on the cellularity of the tumor. T1 signal may be hypo- to isointense with respect to muscle. A variety of enhancement patterns have been described ranging from no enhancement to homogeneous or heterogeneous enhancement.

Clinical history was key to making the correct diagnosis in the above case given the known association of FAP with desmoid tumors. Carcinoid tumors, unlike desmoid tumors, frequently calcify and may be associated with hepatic metastases. Additional sites of lymphadenopathy would be expected in a patient with mesenteric lymphoma.

Questions for Further Thought

1. What is the usual management of desmoid tumors?
2. Name other tumors associated with FAP.

Reporting Responsibilities

1. Describe the size and location of the mass.
2. Describe any complications such as bowel or ureteral obstruction.

What the Treating Physician Needs to Know

1. That a mesenteric or abdominal wall solid mass is most likely a desmoid tumor in a patient with FAP.

Answers

1. Management may include nonsteroidal anti-inflammatory drugs and antiestrogen medications. These tumors have a high rate of recurrence following resection.
2. FAP is an autosomal-dominant colorectal cancer syndrome. Given the near 100% likelihood of developing colon cancer by age 35 to 40, FAP patients usually undergo prophylactic colectomy. Patients may also develop upper gastrointestinal tract polyps. Gardner syndrome is a subdivision of FAP and includes osteomas, desmoid tumors, and soft-tissue tumors. Turcot syndrome is another subdivision of FAP and is associated with central nervous system tumors and desmoid tumors.

REFERENCES
