

AMSER Rad Path Case of the Month:

48 year old male with alcoholic cirrhosis, liver mass, and new-onset diabetes



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Patient Presentation

- **HPI:** 48 year old male followed up for cirrhosis and hypoechoic mass found on complete abdominal ultrasound, originally indicated by hepatic failure, ascites, and secondary thrombocytopenia. MRI subsequently recommended for further evaluation of liver mass.
- **PMH:** Alcoholic cirrhosis, CHF, HTN
- **Physical Exam:** Cachectic appearance. Scleral icterus. Soft, nontender abdomen without rebound or palpable mass. 3+ pitting peripheral edema. Speech is sub-fluent

Pertinent Labs and Imaging

- **Lab** results from admission, several months ago, indicated hepatic failure and new-onset diabetes
 - **CBC:** Hb 10.3; MCV 123.5; Plt 80k; INR 2.2
 - **CMP:** glucose 455; AST 147
 - **Urine:** glucose >500; negative for ketones
- **Complete abdominal US** revealed cirrhosis and a hypoechoic mass

Radiology Images (Not Labeled)

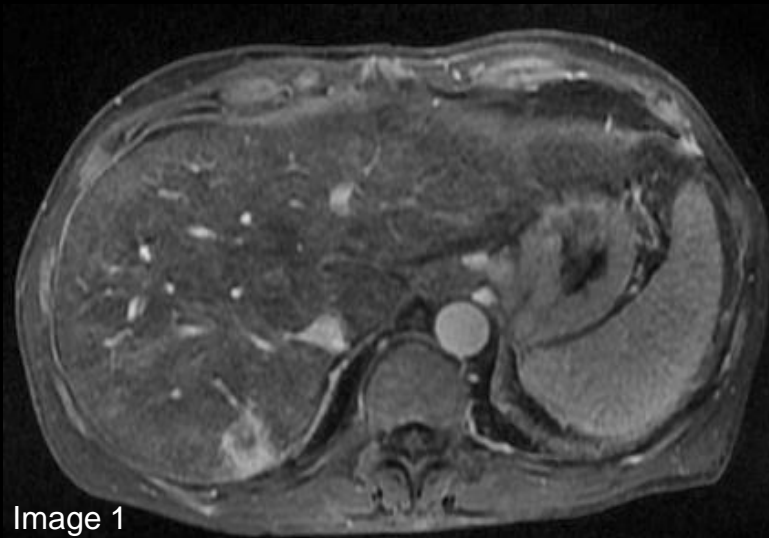


Image 1

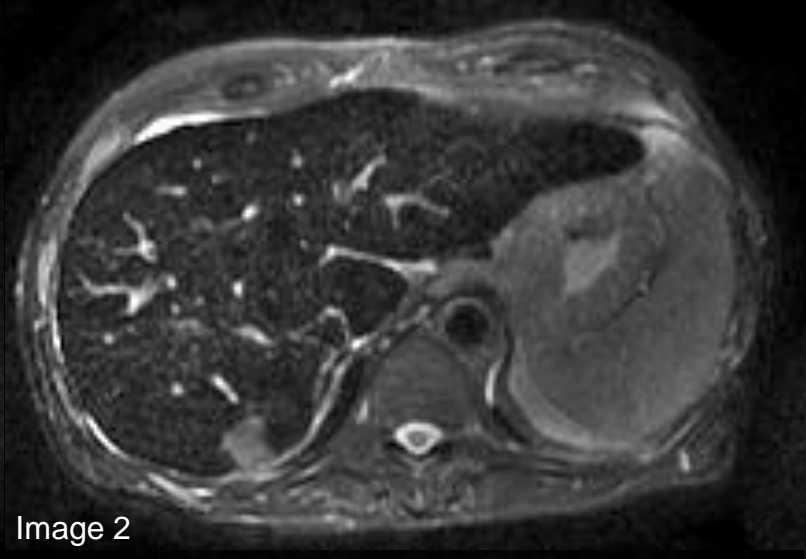


Image 2

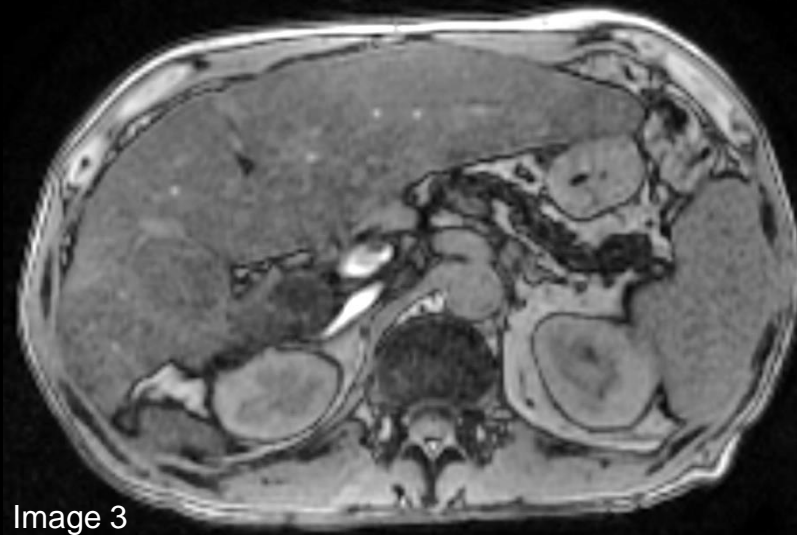


Image 3

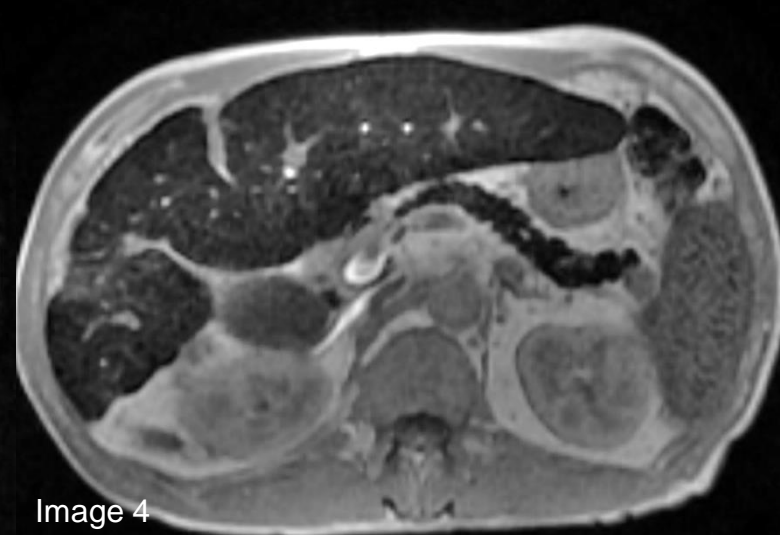
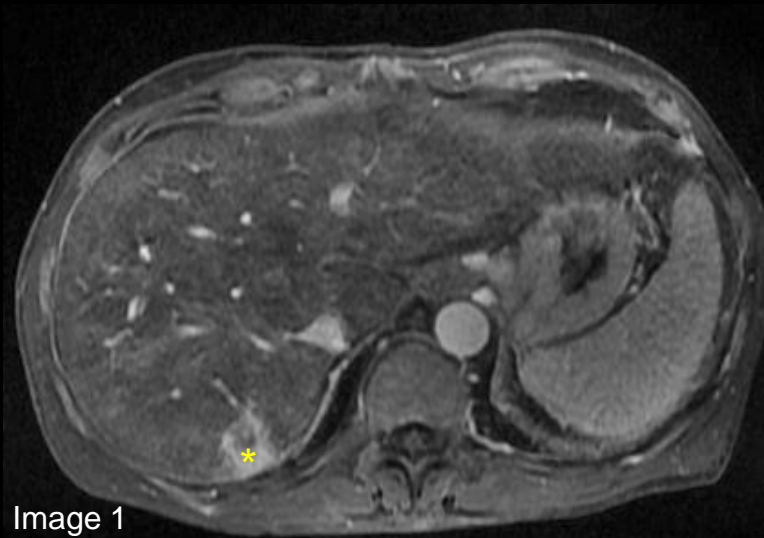


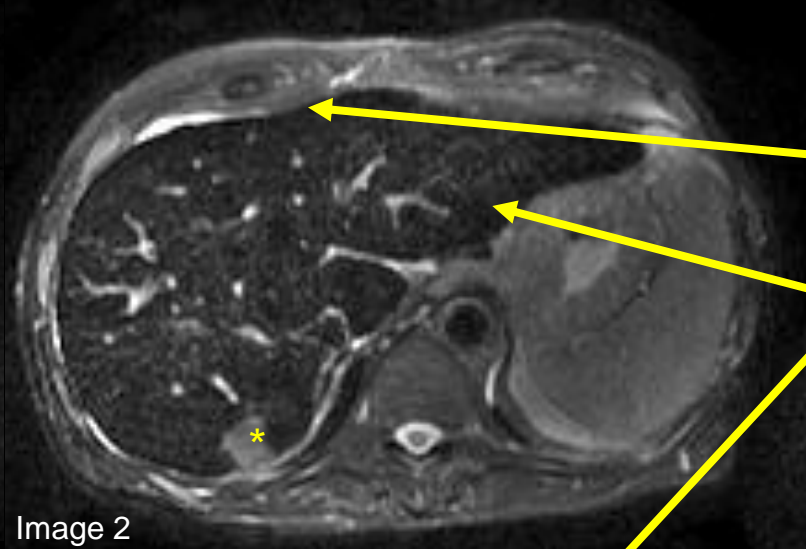
Image 4

Radiology Images (Labeled)



Axial, T1, through liver and spleen with contrast

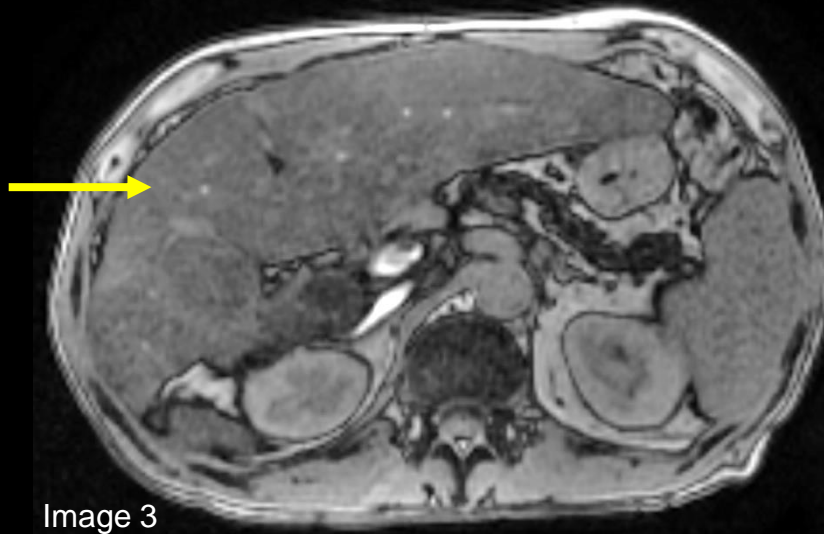
Segment 7 lesion (*) is suspicious for malignancy (LI-RADS-M). The T2 hyperintense mass demonstrates targetoid enhancement



Axial, T2, through liver and spleen

Nodular hepatic contour. Cirrhotic morphology.

Diffuse low signal on T2 weighted sequences with signal loss on in-phase (Image 4) sequence consistent with iron deposition.



Axial, out-of-phase GRE, through pancreas

Liver loses signal on in-phased sequence. This finding indicates iron deposition and is the opposite of what is seen in hepatic steatosis.

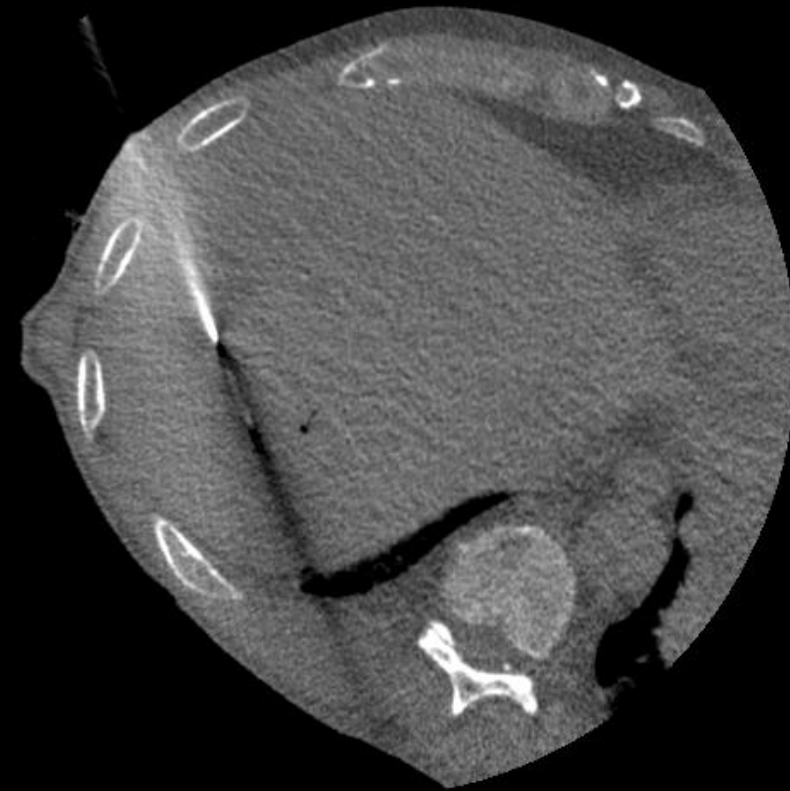


Axial, in-phase GRE, through pancreas

Atrophic pancreas with marked hypointense signal with signal loss on in-phased sequence, indicating iron deposition.

DDX and Next Steps

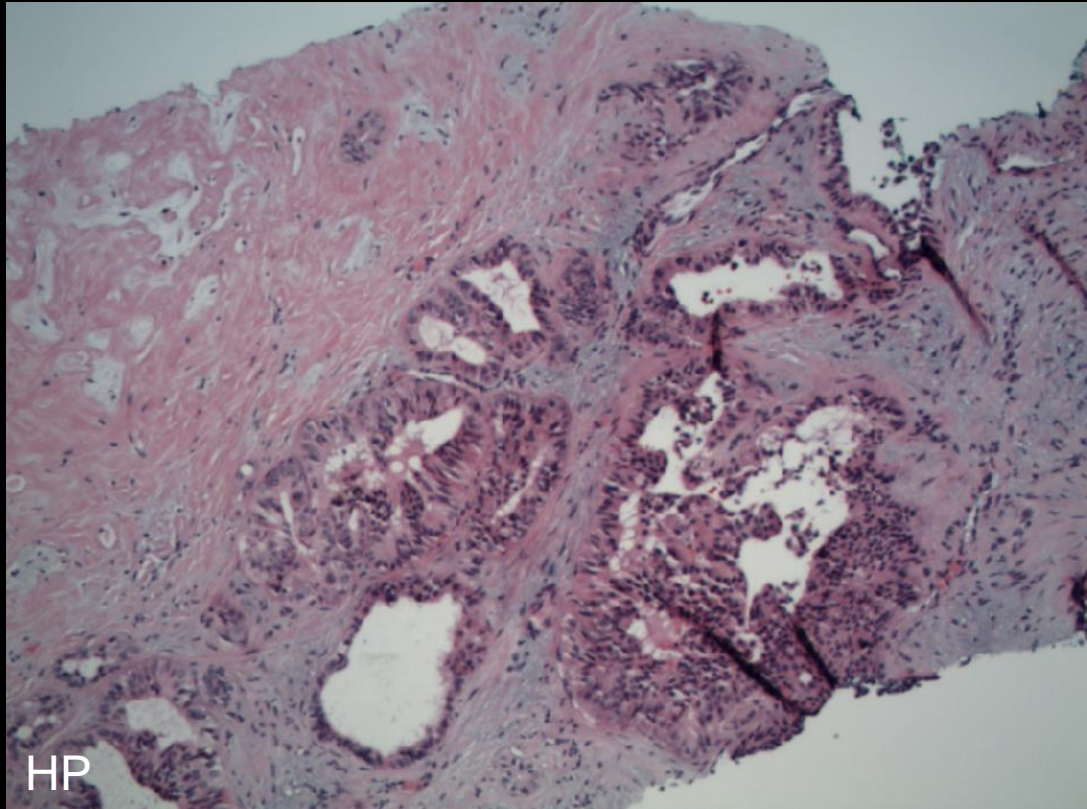
- CT Guided Biopsy of targetoid mass in segment 7 consistent with malignancy (LI-RADS-M).
- **Differential:**
 - Intrahepatic cholangiocarcinoma
 - Hepatocellular carcinoma with atypical features
 - Metastasis



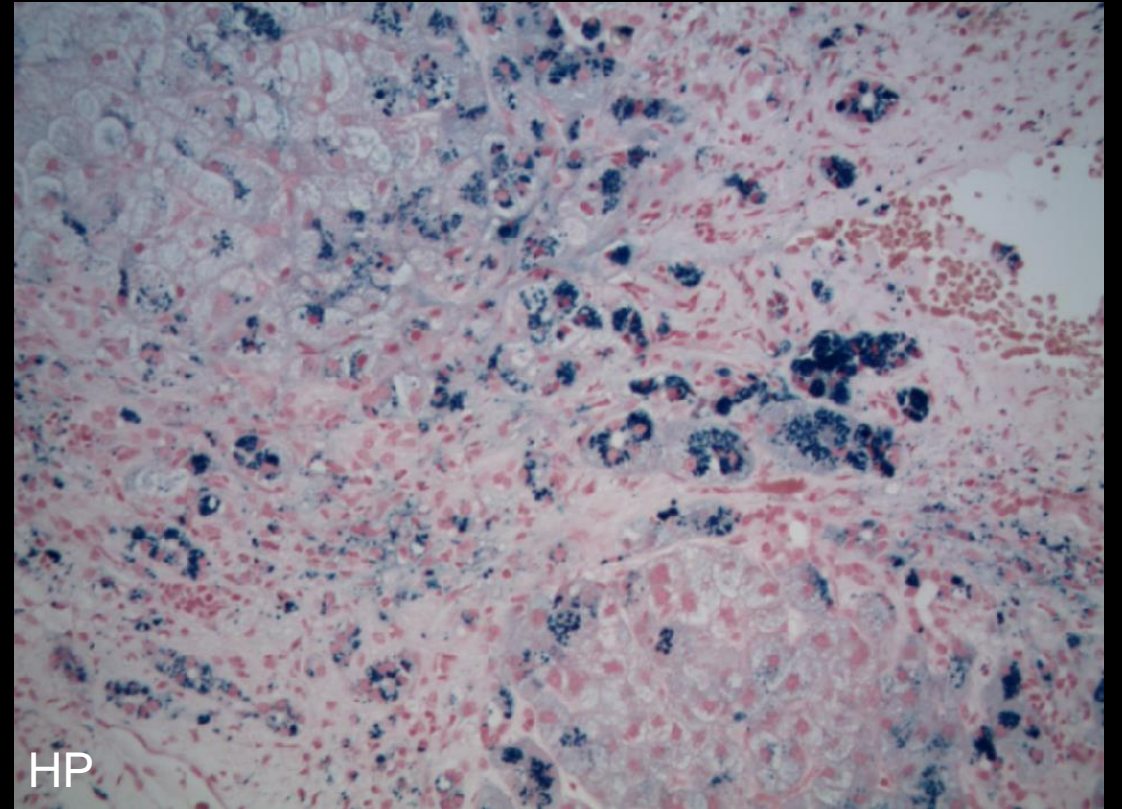
DDX and Next Steps

- CT-guided needle core biopsy – Liver Mass
- Final Pathologic Diagnosis:
 - Well-differentiated adenocarcinoma suggestive of an **intrahepatic cholangiocarcinoma**
 - Adjacent background liver with advanced fibrosis/cirrhosis

Micro Path (Labeled)



Cholangiocarcinoma. Invasive malignant glands in a prominent desmoplastic stroma. Liver biopsy showed a well-differentiated adenocarcinoma. Overall findings are suggestive of an intrahepatic cholangiocarcinoma.



Hereditary hemochromatosis. In this Prussian blue stained section, hepatocellular iron appears blue.

Final Dx:

Intrahepatic Cholangiocarcinoma
Primary Hemochromatosis of liver and pancreas

Case Discussion (1/3) - Hemochromatosis

- Primary Hemochromatosis
 - Autosomal recessive disorder (HFE1 mutation) – disrupts iron regulation leading to overload²
 - Most common genetic disease in Caucasian populations^{2,3}
 - Classic triad: cirrhosis, diabetes mellitus, skin pigmentation
 - Other classic signs: restrictive cardiomyopathy, hypogonadism, arthropathy
 - Alcohol consumption and associated genes can vary phenotypic expression²
 - Increased hepatocellular carcinoma (HCC) risk^{2,3}
 - Hereditary hemochromatosis not classically associated with cholangiocarcinoma.⁸

Case Discussion (2/3) - Hemochromatosis

- Imaging Hemochromatosis
 - CT, although widely available, is not very sensitive for hemochromatosis diagnosis⁴
 - However, dual-energy CT can be used to quantify iron deposition
 - MRI is the most sensitive modality for diagnosis of hemochromatosis and can estimate iron concentration within liver, preventing the need for repeated biopsies⁴

Case Discussion (3/3) - Hemochromatosis

- Imaging Hemochromatosis (continued)
 - Visceral iron results in susceptibility artifact. Leads to T2* signal loss⁵
 - Low signal can be seen on all sequences but particularly on gradient echo out of phase imaging and T2⁵
 - Lower organ signal compared to skeletal muscle indicates the presence of iron⁵
 - Spleen and bone marrow signal is typically normal in primary hemochromatosis and low pancreatic signal is usually seen with cirrhotic liver⁵

Case Discussion (1/3) - Cholangiocarcinoma

- Cholangiocarcinoma

- Malignancy of the cholangiocytes in the biliary tree⁷
- 2nd most common primary hepatic malignancy (~15%) after HCC⁷
- Typical presentation: Painless jaundice
- Common associations and risk factors: ^{6,7}
 - Cirrhosis
 - Primary sclerosing cholangitis
 - Choledochal cysts
 - Clonorchis sinensis
 - Toxins – thorotrast, polyvinylchloride, alcohol
 - Viral – HIV, hepatitis B & C, EBV

Case Discussion (2/3) - Cholangiocarcinoma

- Imaging Cholangiocarcinoma⁷
 - Findings will differ depending on location and growth pattern

| | Mass forming | Periductal infiltrating | Intraductal |
|------------|---|--|---|
| Ultrasound | Homogenous mass of intermediate echogenicity with peripheral hypoechoic halo of compressed liver parenchyma. | Altered bile duct caliber (narrow or dilated) without a well-defined mass. | Altered bile duct caliber, usually duct ectasia with or without visible mass. |
| CT | Low attenuation on non-contrast scans. Heterogeneous minor peripheral enhancement with gradual centripetal enhancement. | Regions of duct wall thickening or of the periductal parenchyma. Altered bile duct caliber (usually narrowed). Contrast enhancement. | Altered bile duct caliber, usually duct ectasia with or without visible mass. |

Case Discussion (3/3) - Cholangiocarcinoma

- Imaging Cholangiocarcinoma

- MRI is the imaging modality of choice as it best visualizes the tumor, biliary ducts, and blood vessels which are essential for determining resectability
- Appearance on MRI are similar to the findings on CT but with better visualization of contrast enhancement and bile duct visualization
- Diffusion Weighted Imaging (DWI/ADC)
 - Demonstrates a peripherally hyperintense “target” appearance on DWI favors cholangiocarcinoma over HCC

LIRADS

- Liver Imaging Reporting and Data System
 - Standardized terminology and classification system for liver lesions in patients that have risk factors for HCC (i.e. cirrhotic livers or HBV without cirrhosis)
 - Scoring is beyond the scope of this presentation but takes into consideration a lesion's size, enhancement pattern with washout, and capsule
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- LR-1 (100% benign such as a cyst or hemangioma)
 - LR-2 (probably benign)
 - LR-3 (intermediate probability for HCC)
 - LR-4 (probably HCC)
 - LR-5 (100% HCC)
 - LR-M (lesions that are probably malignant but whose appearance is not compatible with HCC such as our case)

References:

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7. Gaillard, F., & Di Muzio, B. (n.d.). Cholangiocarcinoma. Retrieved September 23, 2020, from Radiopaedia website: <https://radiopaedia.org/articles/cholangiocarcinoma?lang=us>
8. Morcos M, Dubois S, Bralet MP, Belghiti J, Degott C, Terris B. Primary liver carcinoma in genetic hemochromatosis reveals a broad histologic spectrum. *Am J Clin Pathol*. 2001;116(5):738-743. doi:10.1309/2RFK-HD06-A788-1FJH