Metastatic Ocular Melanoma Mimicking Budd-Chiari Syndrome

Krutika Lakhoo[1], Grace Guzman[2], Adam Mikolajczyk[1]

[1]Division of Gastroenterology and Hepatology, University of Illinois Medical Center, Chicago IL; [2]Department of Pathology, University of Illinois Medical Center, Chicago IL

UI Health UIC

Introduction

- Budd-Chiari syndrome (BCS) is hepatic venous outflow obstruction at the hepatic venules, hepatic veins or inferior vena cava (IVC).
- Classically, it causes right-upper quadrant (RUQ) abdominal pain, hepatomegaly, and ascites.
- It can present with a wide spectrum of imaging findings.
- We present a novel case of metastatic ocular melanoma that mimicked several of these clinical and radiographic features of BCS.

Case Presentation

A 78-year-old man with a history of uveal melanoma treated with radiation therapy (8 years ago) presented with abdominal pain, distention, and jaundice.

- He had no history of liver disease and denied any new medications or heavy alcohol use.
- Physical examination revealed scleral icterus, jaundice, RUQ abdominal pain, and abdominal distention with bulging flanks and a fluid wave.
- Labs showed elevations in total bilirubin (7.8 mg/dL), alkaline phosphatase (131 U/L), alanine aminotransferase (158 U/L), aspartate aminotransferase (272 U/L), and prothrombin time (15.6 seconds).
- Doppler ultrasound showed hepatomegaly, ascites and absent blood flow in the right hepatic vein with patency of the remaining hepatic and portal vein branches.
- MRI of the abdomen revealed hepatomegaly with heterogeneous enhancement of the liver parenchyma, several hyper-enhancing hepatic nodules, and evidence of thrombosis in the posterior right lobe hepatic veins (arrow) (**Figure 1**).

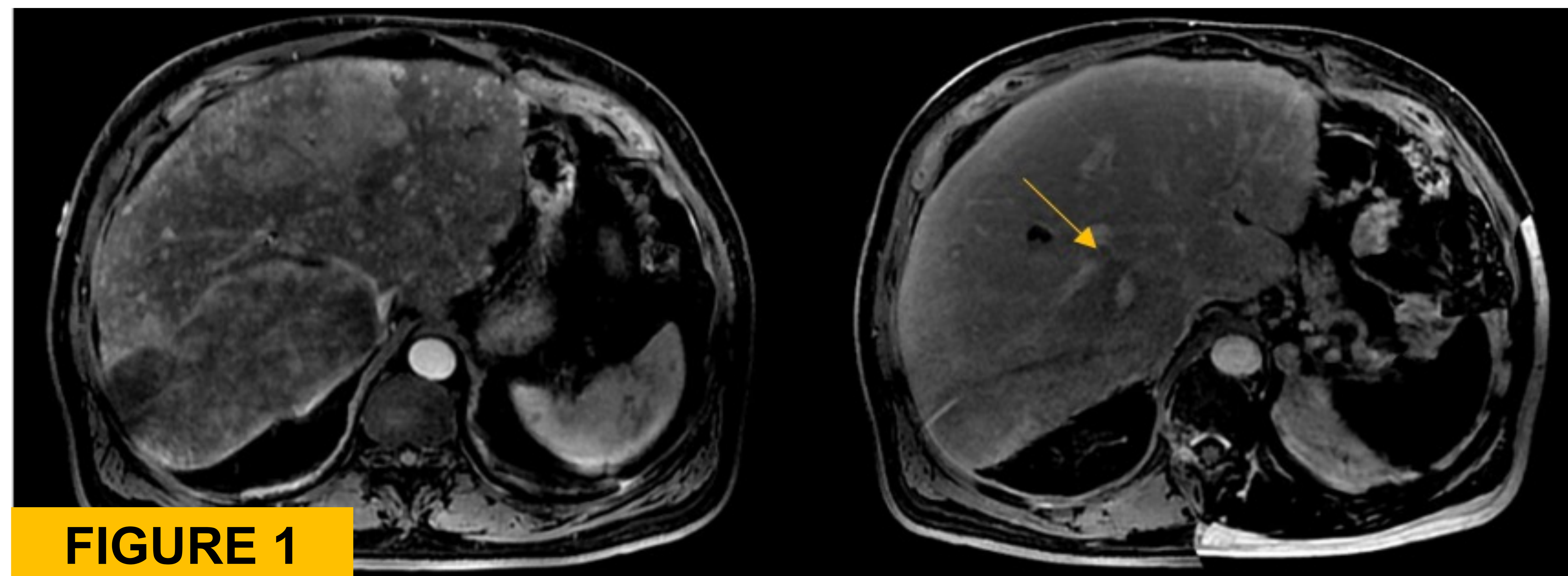

FIGURE 1

- Given a high suspicion for BCS, venography was done, showing nonocclusive thrombus in the peripheral branches of the right hepatic vein with patent central portion and patent central and left hepatic veins.
- The calculated sinusoidal pressure gradient was 12 mmHg.
- Liver biopsy was performed and showed an effaced liver architecture due to nests of multinodular malignant pigmented cell infiltrates preferentially distributed in the centrilobular sinusoids compressing adjacent hepatocytes and displaying histopathological features indicative of metastatic melanoma (**Figure 2**).

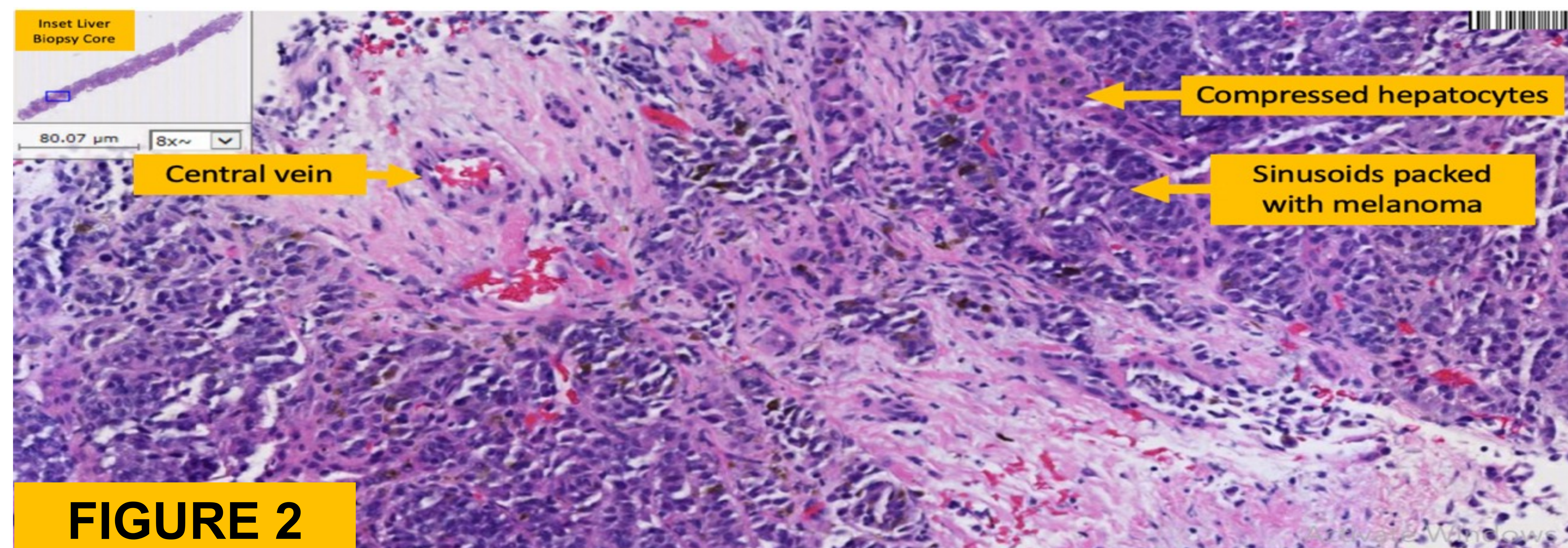

FIGURE 2

- There was strong immunoexpression of biomarker SOX10 which confirmed metastatic melanoma.
- The patient unfortunately developed progressive liver failure and was discharged home to hospice care.

Discussion

- Imaging findings in conjunction with clinical presentation are often essential to definitively diagnose BCS.
- Typical imaging findings include occlusion of the hepatic veins and IVC, caudate lobe hypertrophy, inhomogeneous liver enhancement, and the presence of intrahepatic collateral vessels and hypervascular nodules.
- Other etiologies, such as invasive intrahepatic malignancy, can present with similar radiographic findings and venography (the gold standard test) may be needed to definitively diagnose BCS.
- Uveal melanoma, albeit rare, commonly metastasizes to the sinusoids in the liver and can mimic clinical and radiographic features of BCS, especially when the metastases preference the centrilobular sinusoids as in this case.

REFERENCES:

1. DeLeve L.D et al, Vascular Disorders of the Liver, AASLD Practice Guidelines; Hepatology: 2009
2. Simonetto D.A et al, ACG Clinical Guideline: Disorders of the Hepatic and Mesenteric Circulation; American Journal of Gastroenterology: 2019
3. Narayanan Menon K.V et al, The Budd-Chiari Syndrome, NEJM 2004
4. Brancatelli G, Vilgrain V, Federle MP, Hakime A, Lagalla R, Iannaccone R, Valla D. Budd-Chiari syndrome: spectrum of imaging findings. AJR Am J Roentgenol. 2007