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CASE REPORT | LIVER

An Infiltrative Case of Angiosarcoma Causing Portal Hypertension

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ABSTRACT

Hepatic angiosarcoma is a rare and aggressive liver tumor. We report a case study of an 82-year-old elderly gentleman who presented with failure to thrive and ascites. Initially suspected to be cirrhosis, biopsy results eventually concluded angiosarcoma of the liver. Our patient presented with an infiltrative form, rather than distinct masses, which led to portal hypertension and ascites. The variance in symptomatology and radiology presentations make a diagnosis of hepatic angiosarcoma challenging and require a high index of suspicion.

KEYWORDS: hepatic angiosarcoma; hepatic tumor; portal hypertension

INTRODUCTION

Hepatic angiosarcoma (HAS) is a rare and aggressive malignancy, accounting for only 2% of primary liver malignancy with majority of untreated patients dying within 6 months.[1,2] A review of the National Cancer Institute's epidemiology database identified only 207 patients with primary HAS over a period of 30+ years.[3] Certain exposure agents—such as vinyl chloride, Thorotrast, arsenic, and anabolic steroid use—have been linked with HAS, although 75% of etiology still remains unknown.[4,5] The largely idiopathic nature, low incidence rate, and variable presentation hinder the development of efficient diagnostic protocols and effective treatment options. Here, we present a case of an infiltrative presentation of this exceedingly rare disease.

CASE REPORT

An 82-year-old man was admitted for failure to thrive. He had large ascites and splenomegaly. Laboratory workup revealed thrombocytopenia, aspartate aminotransferase 166 U/L, alanine aminotransferase 60 U/L, total bilirubin 2.8 mg/dL, and alkaline phosphatase 500 U/L. Albumin was 3.4 g/dL. Serum ascites albumin gradient >1.1 suggested ascites secondary to portal hypertension. The total protein of the ascitic fluid was 1.0 g/dL with negative cytology. Abdominal computed tomography demonstrated splenic lesions (Figure 1). Owing to concern for infectious etiology, the inpatient medical team ordered serology for Coxiella, Bartonella, and Brucellosis, all of which were negative. Positron emission tomography-computed tomography scan showed fluorodeoxyglucose uptake in the liver, spleen, and bone marrow (Figure 2). Flow cytometry and bone marrow aspirates were unrevealing. A detailed history and physical examination did not provide further concerns for alcohol-associated liver disease, metabolic-associated liver disease, or drug-induced liver injury. Chronic hepatitis A, B, and D were negative. Genetic causes including α-1 antitrypsin deficiency and hereditary hemochromatosis were excluded. No features of autoimmune liver disease were found on liver biopsy. Imaging did not show concern for chronic Budd-Chiari syndrome. Liver biopsy was diagnostic for HAS (Figures 3 and 4: Hematoxylin and eosin stain, magnification, 200×; immunohistochemical stain for ERG showing nuclear tumor positivity, magnification 200×), with atypical spindle cell proliferation, hyperchromatic nuclei, and scant pale eosinophilic cytoplasm growing along sinusoids causing destruction of liver cell plates. Detailed medical and surgical history following the diagnosis did not elucidate

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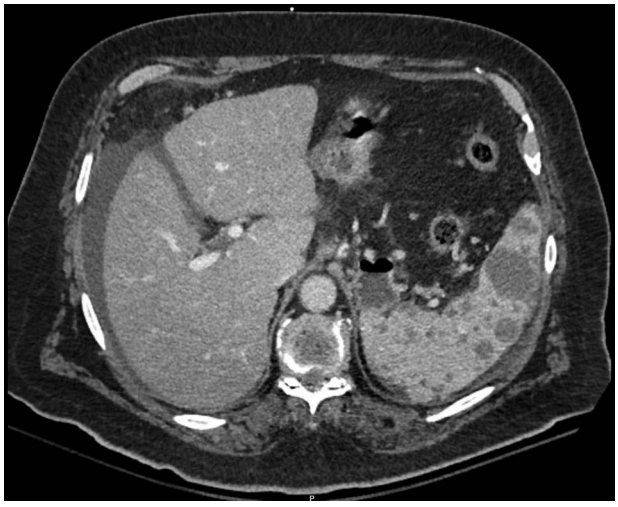

Figure 1. Abdominal computed tomography demonstrating splenic lesions.

clear underlying risk of development of this liver tumor. Owing to our patient's age and comorbidities, his family elected to transition him to comfort care. He died a few weeks later.

DISCUSSION

HAS is a rare malignant tumor which arises from vascular endothelium with poor prognosis and rapid clinical decline.[6] Clinical presentation can be nonspecific with hepatomegaly, pain, anorexia, fatigue, and weight loss. Most cases present as hypervascular foci, allowing for detection as distinct masses through contrast imaging.[7] This patient presented with an infiltrative form, highlighting the difficulty with early diagnosis due to the variance in symptomatology and radiologic features. Moreover, extrahepatic involvement is common, and, in this case, the splenic lesions found on the CT scan may present as metastasis. Interestingly, this patient also presented with features of noncirrhotic portal hypertension. A study by Wu X et al suggested that HAS and noncirrhotic portal hypertension may share similar etiologies although no clear associations have been made yet.[8]

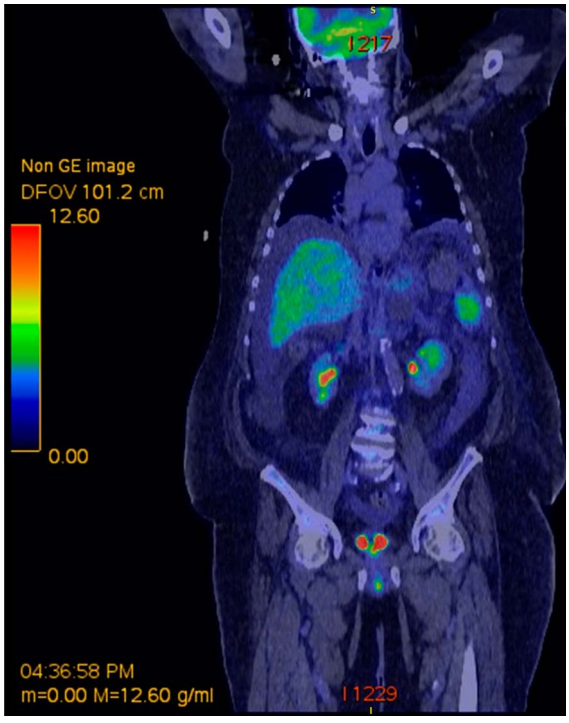

Figure 2. Positron emission tomography-computed tomography scan showed fluorodeoxyglucose uptake in the liver, spleen, and bone marrow.

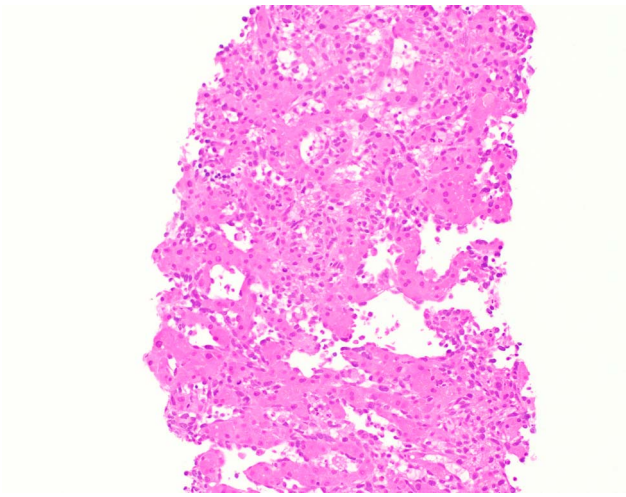

Figure 3. Liver biopsy was diagnostic for hepatic angiosarcoma (Hematoxylin and eosin stain; magnification, 200×) with atypical spindle cell proliferation, hyperchromatic nuclei, and scant pale eosinophilic cytoplasm growing along sinusoids causing destruction of liver cell plates.

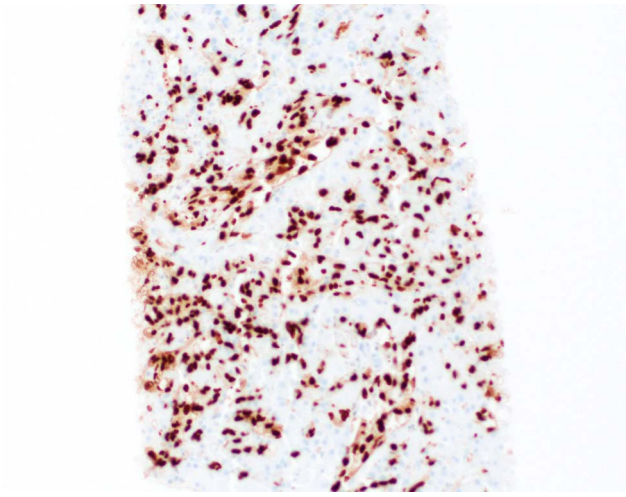

Figure 4. Liver biopsy was diagnostic for hepatic angiosarcoma (immunohistochemical stain for ERG showing nuclear tumor positivity; magnification, 200×) with atypical spindle cell proliferation, hyperchromatic nuclei, and scant pale eosinophilic cytoplasm growing along sinusoids causing destruction of liver cell plates.

The number of patients with primary HAS is low, and furthermore, the proportion of those with the infiltrative form is even more rare. Primary angiosarcoma of the liver disproportionately occur in men with peak incidence in the sixth or seventh decade of life.[5] Unfortunately, prognosis is poor with a rapid clinical decline 6–12 months following diagnosis. Complications include hepatic failure or intra-abdominal bleeding secondary to rupture. The poor prognosis is mostly attributed to the aggressive nature of the tumor, early metastasis, and the relatively low awareness among physicians, which further delay the diagnostic process and optimal treatment administration.[9] Currently, there are no established guidelines for management and treatment. In patients with good functional status, radical resection can be considered. Liver transplantation is contraindicated due to high rate of recurrence. Transcatheter arterial chemoembolization can be used to control bleeding or to palliate a dominant mass.

This case presented a diagnostic challenge because our patients' workup for failure to thrive initially revealed anemia, thrombocytopenia, elevated liver biochemistries, and splenic lesions. He was found to have an infiltrative form of HAS, rather than distinct masses, which may have contributed to the portal hypertension and ascites. Without the presence of the splenic lesions, it is likely he would have been diagnosed with cirrhosis. As infiltrative angiosarcoma is a rare finding on workup for liver disease, this case helps highlight keeping the differential diagnosis broad as you evaluate for nonspecific symptoms of pain, fatigue, and weight loss in the setting of hepatomegaly.

DISCLOSURES

Author contributions: G. Bae and KA Dunleavy wrote and revised the manuscript. C. Hagen provided the radiographic and histologic images. DA Simonetto and MF Abdelmalek supervised and reviewed the manuscript for intellectual content. KA Dunleavy is the article guarantor.

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