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Primary Hepatic Leiomyosarcoma Presenting as a Budd-Chiari-like Syndrome

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INTRODUCTION

Primary Hepatic Leiomyosarcoma (PHL) is a rare malignant tumor arising from smooth muscle cells of either vascular, biliary, or ligamentous origin. Underlying etiologic factors are unknown, with few associations described in the literature. The median age of diagnosis is 58 years. Diagnosis is often delayed due to the tumor causing nonspecific symptoms and often remaining asymptomatic until mass effect is produced. Once PHL presents itself, it is still challenging to diagnose due to the non-specific nature of symptoms and lack of serological or tumor markers. This case represents one of thirty cases reported in the literature and is consistent with the presentation and disease course described by previous case reports in the literature, yet with an interesting difference in the patient’s history.

LAB DATA AND IMAGING

A repeat CT scan and an MRI were performed at our institution (Figure 1 and Figure 2). Due to the suspicion for hepatic abscess, CT guided drainage was ordered. However, given the solid nature of the lesion, a CT guided liver biopsy demonstrated nonspecific fibrotic and infarcted liver parenchyma. As infectious workup and tumor markers were unrevealing, a repeat biopsy was obtained. This revealed a poorly differentiated carcinoma without a definitive primary site. Further imaging studies to assess for additional lesions and metastatic disease were unrevealing.

DISCUSSION

PHL is a rare malignant tumor. Most studies are limited to case reports or case series. In our patient, with an extensive travel history, yet low risk for cirrhosis or hepatocellular carcinoma, the differential diagnosis initially pointed to infectious disease. Hepatocellular carcinoma and hydatid disease are often initial diagnoses in other cases eventually diagnosed with PHL. PHL is especially difficult to diagnose because of the non-specific symptoms or lab work, including lack of established tumor makers or serological markers.

The rarity of the disease makes clinical judgment and standard of care a challenge. Until recently, surgeries involving the IVC in primary or metastatic liver tumors were considered too complex due to high-risk morbidity and mortality. Tumors arising from the hepatic veins are more prone to develop Budd-Chiari syndrome and have a worse prognosis. Yet, surgery offers the only chance for cure. Unfortunately, surgical removal does not guarantee long-term survival. Alternative therapies, such as radiotherapy and chemotherapy often proved insufficient.

CASE

A 60-year-old male with extensive travel history to developing countries initially presented to a medical facility in Micronesia with bilateral lower extremity edema, dyspnea, fever, and fatigue attributed to an upper respiratory infection. He ultimately underwent CT of the abdomen and pelvis in Guam, which revealed a 13.6 x 16.8cm right hepatic lobe mass with obstruction of the IVC. He left Guam without a definitive diagnosis and presented to our institution two days later.

On arrival, he complained of non-specific symptoms such as dyspnea, malaise, weight loss, fever and lower extremity edema. Physical exam was notable for abdominal distention and palpable organomegaly, bilateral pitting lower extremity edema to the knee with a desquamating rash on all extremities. Initial vitals: BP: 115/78  Pulse: 110 Temp: 100.5F Respiration Rate: 20

Due to leukocytosis, fever, and recent travel to developing areas with known exposures to unclean water, animals, and work as a plumber, the initial leading diagnoses were largely infectious etiologies. Hepatocellular carcinoma remained lower on the differential as he had no history of cirrhosis, hepatitis C, or alcohol abuse.

The patient underwent open right hepatic lobectomy with resection of tumor and repair of the IVC with Surgical Oncology and Cardiothoracic Surgery. The patient tolerated surgery remarkably well and was subsequently discharged home with plans for potential adjuvant treatment. Surgical pathology, reviewed by an outside institution, demonstrated epithelioid elements with regions of smooth muscle morphology supporting the impression of high-grade PHL (Figure 3). The patient underwent a trial of chemotherapy, but ultimately succumbed to the malignancy within a year.

CONCLUSIONS

- Primary hepatic leiomyosarcoma is an exceedingly rare liver tumor with a poor prognosis. The literature on this condition is limited to mostly case reports and series.
- The characteristically delayed presentation due to nonspecific symptoms and the challenges with diagnostic and therapeutic modalities all contribute to difficulties with diagnosis and treatment.
- Most PHL cases are thought to arise from the wall of major blood vessels. It is likely that our patient’s tumor originated from a venous structure.
- Surgical resection is the management of choice with the role of chemotherapy and radiation not well defined.
- Although rare, primary hepatic leiomyosarcoma should remain on the differential of hepatic masses in patients without the classic risk factors for hepatocellular carcinoma.

References:


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