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Case Report

Primary Leiomyosarcoma of the Inferior Cava Vena: Case Report and Literature Review

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Abstract

Primary leiomyosarcoma of the inferior cava vena is an exceptionally rare malignant tumor arising from the smooth muscle of the vessel wall, with only a few hundred cases that have been documented in medical literature. There are no well-defined treatment protocols; however, surgical removal currently stands as the sole curative option. Further research is essential to gain a deeper understanding of the disease and provide guidance for its management. We present a case involving a 50-year-old man who was admitted with a six-month history of vague abdominal pain and weight loss. Abdominal computed tomography revealed a mass araising from with the inferior vena cava associated with hepatic nodules. Histological examination confirmed the diagnosis of leiomyosarcoma of the inferior vena cava with liver metastases.

Keywords: *Leiomyosarcoma; Liver; Primary; Imaging; Inferior Vena Cava*

Introduction

Primary hepatic leiomyosarcomas (PHL) account for 6 - 16% of primary hepatic sarcomas, constituting a rare type of malignant tumor originating from smooth muscles. They represent only 0.2 - 2% of all primary hepatic cancers [1].

PHL is challenging to diagnose due to its nonspecific clinical presentation and imaging features, often resembling more common liver tumors like hepatocellular carcinoma and intrahepatic cholangiocarcinoma.

This tumor exhibits aggressive metastatic potential and is typically detected at an advanced or metastatic stage. Radical hepatectomy is the only curative treatment for PHL, but a significant number of patients have unresectable disease and/or metastases [2].

In this report, we present one case of PHL, contributing to an updated understanding of the disease's incidence, diagnosis, and management.

Case Presentation

A 50-year-old man, with no significant medical history, presented to the hospital with a six-month history of abdominal pain and weight loss. His clinical examination revealed hepatomegaly, without ascites, fever, or jaundice. Laboratory tests showed hepatic cytolysis. An abdominal ultrasound revealed a large tissular mass in the right lobe of the liver. Then, the patient underwent an abdominal CT scan

which confirmed the presence of a tissular lesion in the right lobe of the liver, hypodense, showing heterogeneous enhancement after contrast agent injection, and causing invasion with tumor thrombosis of the inferior vena cava (Figure 1). The diagnosis of an atypical hepatocellular carcinoma on a non-cirrhotic liver was suspected.

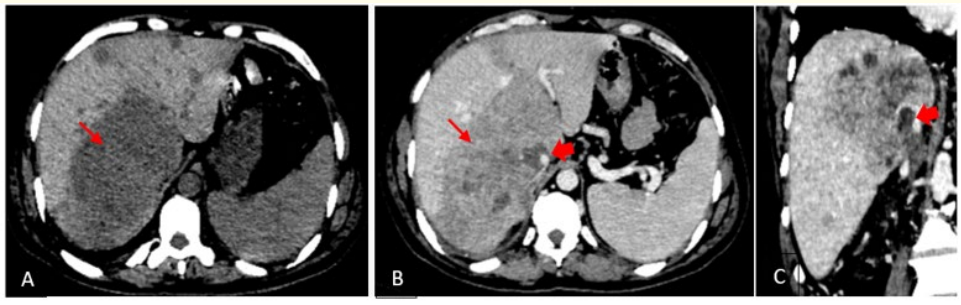

Figure 1: *A: Non enhanced computed tomography in axial section showing a large hypodense tumor of the right liver (arrow). B/C: Contrast enhanced CT scan in axial and sagittal sections indicated significant heterogeneous enhancement of the mass (arrow) with tumor thrombosis of the inferior vena cava (arrowhead).*

A percutaneous-guided biopsy was performed, and the histopathological examination revealed the presence of a malignant spindle cell tumor, suggesting the possibility of a sarcomatous process or a sarcomatoid carcinoma. Immunohistochemical analysis confirmed the diagnosis of a hepatic origin leiomyosarcoma.

Subsequently, the patient underwent a staging assessment, revealing the presence of secondary hepatic and pulmonary metastases (Figure 2). The case was discussed in a multidisciplinary meeting, during which the diagnosis of metastatic primary hepatic leiomyosarcoma was confirmed, and the decision was made to initiate the patient on palliative chemotherapy.

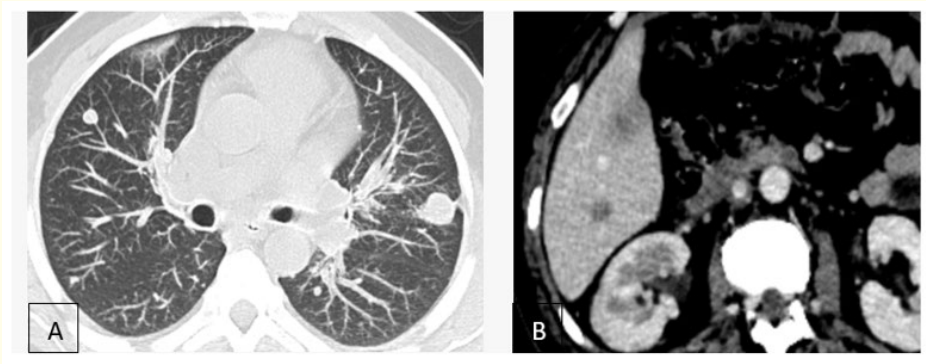

Figure 2: *Contrast enhanced CT scan in axial section showing pulmonary (A) and hepatic (B) metastatic.*

The patient received 3 cycles of doxorubicin-based chemotherapy. A follow-up CT scan showed progression of the primary and metastatic tumor pathology (Figure 3).

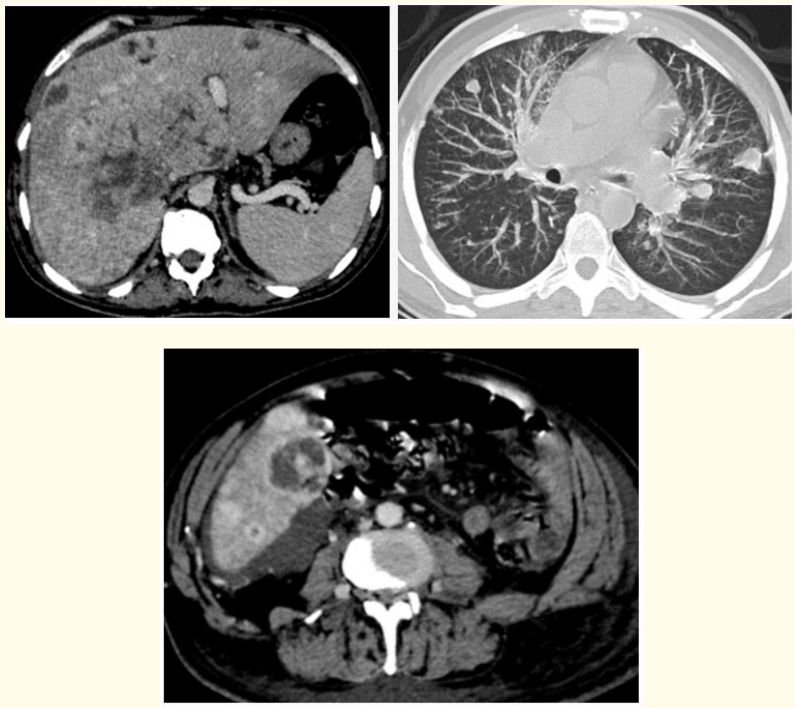

Figure 3: *A 6 month follow CT scan showing a progression of the leiomyosarcoma and the pulmonary and hepatic metastasis.*

Discussion

PHLs, originating from smooth muscle, can arise from intrahepatic vascular structures, bile ducts, or the round ligament [3]. In our case, the presence of a portal vein tumor thrombosis suggests a portal vein origin, which is exceptionally rare with only few reported cases. It's important to note that when a portal vein tumor thrombosis is observed in a previously healthy liver, the initial suspicion is often hepatocellular carcinoma, as was the case with our patient [4].

Diagnosing PHL can be challenging as the clinical presentation and cross-imaging are nonspecific. Patients with hepatic leiomyosarcoma commonly experience symptoms such as abdominal pain, weight loss, loss of appetite, and weakness [2]. Less frequently, they may exhibit symptoms like fever, abdominal distension, back pain, and jaundice [5]. However, small tumors might not cause noticeable symptoms [6].

The age at which the tumor is detected varies widely, ranging from 5 months to 86 years, with an average age of around 58 years. There is no significant gender difference in tumor incidence, although slightly more males are affected [2].

The cause of hepatic leiomyosarcoma remains unknown and has been associated with factors such as viral hepatitis (B and C), advanced HIV/AIDS, Hodgkin lymphoma, autosomal dominant polycystic kidney disease, kidney transplantation, or other concurrent tumors. Additionally, there was a previous hypothesis about the oncogenic effect of hepatitis C, but no evidence has supported the presence of the hepatitis C virus genome in these tumors [7,8].

In our patient's case, a middle-aged man who was previously healthy, none of the mentioned known risk factors were present.

Hepatic leiomyosarcoma commonly occurs in the right lobe and is often large in size, with average diameters reaching up to 14 cm. CT scans of hepatic leiomyosarcomas can reveal either well-defined or poorly defined hypodense masses, often necrotic and occasionally exhibiting bleeding or heterogeneous enhancement [9]. Magnetic resonance imaging (MRI) of the tumor may depict lobulated masses with hypo-intensity in T1-weighted images, heterogeneous hyperintensities in T2-weighted images, high signal on diffusion-weighted imaging, heterogeneous enhancement, and occasional capsules. Hepatic leiomyosarcomas typically exhibit a high metabolic activity on positron emission tomography.

The main differential diagnoses are hepatocellular carcinoma and cholangiocarcinoma. Distinguishing hepatic leiomyosarcoma from hepatocellular carcinoma (HCC), the latter is frequently linked with cirrhosis and elevated AFP levels, which serve as distinguishing features between the two tumors. Nevertheless, hepatic leiomyosarcoma, like HCC, can also manifest with a wash-out enhancement pattern [1].

Tumor biopsy is the definitive means of diagnosis. Immunohistochemistry can facilitate the correct diagnosis and staging [6].

Radical R0 hepatectomy is the primary approach for managing PHL. But was unfortunately non feasible for our patient since he was diagnosed at a metastatic stage. A systematic review by Chi., *et al*. analyzed 111 cases, of which 64% were operable. Most of the unoperated patients had technically unresectable disease or extra-hepatic metastases [10].

The median overall survival was 19 months, with 1-, 2-, and 5-year survival rates of 61.2%, 41.1%, and 14.5%, respectively. Smaller lesion size and, more importantly, tumor-free resection margins were associated with improved survival [11].

The role of chemotherapy for PHL, whether as a neoadjuvant, adjuvant, or palliative treatment, remains uncertain. The potential role of liver transplantation in PHL management is also a topic of debate.

Conclusion

In summary, primary hepatic leiomyosarcomas are rare, and their preoperative diagnosis requires a high degree of suspicion due to atypical clinical presentation and nonspecific imaging features. Surgery, with a focus on achieving R0 resection, is the cornerstone of PHL management, with tumor size and margin status being key predictors of survival. The role of chemotherapy and liver transplantation in PHL treatment is still a subject of ongoing discussion.

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