Primary Hepatic Leiomyosarcoma (PHL) with pulmonary metastatic activity. Case report and literature review

Monserrat M. Munguía-Guízar[1], Jocelyn Serrano-Casas[1], Víctor H. García-y-García[1], Guadalupe C. Che-Cantun[1], Eduardo Fragoso-Pérez[2]

[1] *Gastroenterology department Hospital de Especialidades CMN, Puebla*
[2] *Pathology department, Hospital de Especialidades CMN, Puebla*

Introduction and Objectives: Sarcomas from the liver are rare, constituting only 1%-2% of all primary hepatic malignant tumors. Leiomyosarcoma accounts for 8%-10% of all hepatic sarcomas. However, these are generally metastatic tumors, arising from the GI tract, uterus, retroperitoneum, or lungs. So far, less than 100 cases of primary hepatic leiomyosarcoma (PHL) have been reported in the literature. PHL is a rare mesenchymal hepatic tumor whose clinical manifestations are often nonspecific and remain asymptomatic until there is a significant increase in tumor size, causing a mass effect. Furthermore, alpha-fetoprotein and other serological markers are usually normal. Thus, histological examination is the only way to achieve the diagnosis. The pathological features of leiomyosarcoma include spindle-shaped cells, abundant cytoplasm, nuclear atypia, and the presence of mitotic figures. Immunohistochemical staining of tumors is positive for vimentin, desmin and actin, and negative for a-fetoprotein, CD34, CD117, cytokeratin, and hepatocytes.

No risk factors have yet been identified. Interestingly, many of these tumors occur in immunocompromised individuals, such as post-transplantation and AIDS patients. This is postulated to be secondary to the effect of the uninhibited Epstein-Barr virus (EBV) on smooth muscle proliferation and previously treated Hodgkin's lymphoma. However, patients without any predisposing condition have also been described in the literature.

Surgical resection is considered to be the only potentially curative treatment. Standard chemotherapies (doxorubicin, ifosfamide; gemcitabine and docetaxel) have modest activity with single−agent and combination response rates of 10%-20% and 17%-40%, respectively.

Materials and Patients: A 43-year-old male with a history of alcoholism consuming 135 g/week for three years, smoking index of 1.8 packs/year, and six years diagnosis with type 2 diabetes mellitus. The clinical presentation started with pain in the upper right quadrant, nocturnal diaphoresis, and weight loss, progressing to hiccups, nausea, and vomiting.

Results: Dynamic CT revealed an irregular heterogeneous liver tumor involving the entire right lobe, with attenuation ranging from 7-45 HU in the non-contrast phase, showing heterogeneous enhancement in arterial phase up to 138 HU, with hypodense areas suggestive of necrosis, isodense to the parenchyma in the portal and delayed phases, measuring 174 × 138 × 170 mm. There were adenopathies and pulmonary nodules, the largest measuring 7.3 × 6.2 mm. Liver biochemistry and tumor markers AFP, Ca 19-9, and CEA were within normal limits. Liver biopsy revealed a malignant mesenchymal neoplasm composed of spindle cells with nuclear pleomorphism and atypical mitotic figures within the hepatic parenchyma. Immunohistochemistry showed positive staining for smooth muscle actin, desmin, calponin, DOG1, and negative for CD34, cytokeratin, CD117, ACE, consistent with leiomyosarcoma.

Conclusions: Palliative chemotherapy with doxorubicin and ifosfamide was initiated in 2021. A year later, clinical evolution ECOG 1, but follow-up CT showed a lesion measuring 314 × 159 × 155mm, leading to the decision for supportive care.

Ethical statement

The identity of the patients is protected. Consentment was obtained.

Declaration of interests

None

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

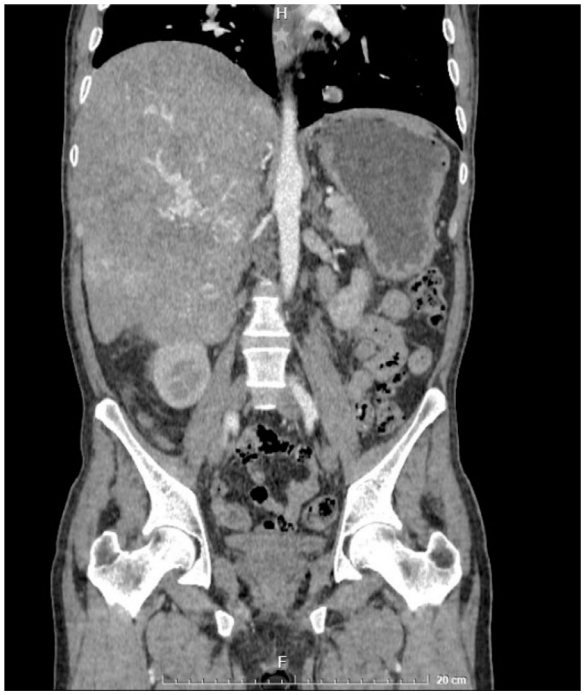

Figure 1. Triphasic CT in arterial phase, coronal section, showing an irregular and heterogeneous liver tumor, which fully involves the right hepatic lobe, dimensions 174 × 138 × 170 mm, with heterogeneous enhancement in the arterial phase of up to 138 HU, and hypodense areas in relation to necrosis.

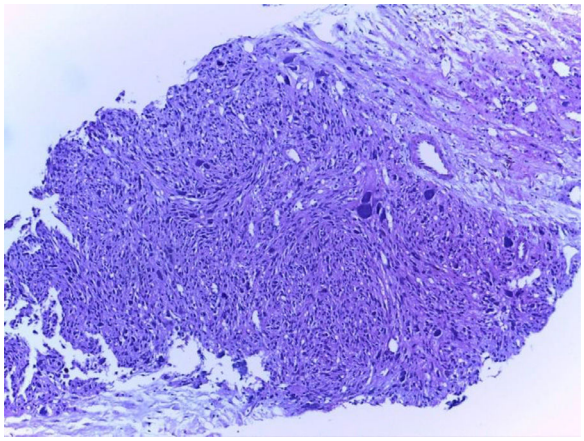

Figure 2. Hematoxylin-eosin stained liver biopsy showing spindle cell neoplasm with eosinophilic cytoplasm, nuclear pleomorphism and atypical mitotic figures.

https://doi.org/10.1016/j.aohep.2024.101405