



Laparoscopically Assisted Resection of a Primary Hepatic Leiomyosarcoma: A Safe Approach in a Rare Malignancy[☆]

Resección laparoscópica asistida de leiomiosarcoma hepático primario: un abordaje seguro en una tumoración infrecuente

Primary hepatic leiomyosarcomas (PHL) are malignant mesenchymal tumors derived from smooth muscle cells that are quite rare and difficult to diagnose preoperatively. Their poor prognosis requires R0 resection, which allows us to achieve long-term survival. Here, we present the case of a PHL that was completely resected by hand-assisted laparoscopic surgery (HALS).¹

A 63-year-old woman with no prior history of interest reported abdominal pain. Upon examination, a mass was palpated in the epigastrium. Abdominal ultrasound and contrast-enhanced CT scan detected a 7 cm mass in the left hepatic lobe, with no evidence of metastatic disease. FNA reported a mesenchymal tumor suggestive of leiomyosarcoma. On PET-CT, the mass was seen to have malignant metabolic characteristics with a maximum SUV of 4.5. We performed HALS and found a 7 cm mass in segment III of the left hepatic lobe; segment IV and the rest of the liver parenchyma were normal on intraoperative ultrasound. A laparoscopic left lateral sectionectomy was performed without portal occlusion. The surgical time was 120 min, and no blood transfusion was required. The patient was discharged 3 days post-op. Microscopically, the hepatic parenchyma was observed to be infiltrated by a malignant mesenchymal neoplasm derived from smooth muscle cells, consisting of bands that crossed in different directions of atypical ovoid or fusiform cells with occasional multinucleated cells (Fig. 1). In addition, areas of necrosis and up to 12 mitotic figures were observed. In the wall of an intrahepatic vessel, these malignant cells were observed to be "emerging" from the wall. The neoplastic cells showed intense cytoplasmic positivity for vimentin, smooth muscle actin and desmin, and negativity for cytokeratin AE1/AE3, CD-117 (C-Kit), DOG-1 and S-100. The Ki-67 cell proliferation index was 20%-30%. The patient did not receive adjuvant treatment and is disease-free 132 months after the intervention.

Hepatic sarcomas are exceptional; they represent 0.1%-1% of primary hepatic malignancies.² Although angiosarcoma is most frequent, PHL constitutes around 8%-10%.³ Leiomyosarcomas can arise from the muscular walls of intrahepatic vascular structures, bile ducts, or the round ligament. Most hepatic leiomyosarcomas are metastases of leiomyosarcomas from other locations (uterus, retroperitoneum, gastrointestinal tract⁴); therefore, their exclusion is essential for an accurate diagnosis.

Symptoms are non-specific and frequently include pain in the upper hemiabdomen accompanied by weight loss, low-grade fever, asthenia, jaundice, and even acute intra-abdominal bleeding secondary to tumor rupture. Physical examination may reveal hepatomegaly or an abdominal mass, whereas laboratory data (bilirubin, alkaline phosphatase, transaminases, alpha-fetoprotein or other tumor markers) are usually not useful for diagnosis.⁵

CT usually describes a large heterogeneous, hypodense mass with internal and peripheral enhancement with occasional central necrosis, similar to the findings in our patient.⁶ MRI showed homogeneous hypointensity in T1 and homogeneous hyperintensity in T2.⁷

Given the non-specific physical, analytical and radiological data, preoperative histology study (by cytology or ultrasound-guided percutaneous biopsy) and postoperative histological study are key factors in the diagnosis. Macroscopically, the surface of the cut is usually whitish-pink and yellow, with areas of necrosis or dark red, caused by hemorrhagic foci, and the neoplasm shows a good delimitation from the adjacent liver parenchyma, separated by a capsule-like fibrocollagenous tissue. Histologically, the tumor usually consists of interconnected fascicles of cells with abundant cytoplasm of elongated or ovoid nuclei, with variable atypia; multinucleated cells and cells with vacuolated or eosinophilic cytoplasm can be observed. Mitotic figures are frequent and there is usually necrosis. The histological finding of presence of these neoplastic cells that emerge from the wall of intrahepatic vessels supports their origin in these structures. They express immunohistochemical markers that support the smooth muscle origin, such as smooth muscle actin, desmin and H-caldesmon, and they are negative for other markers, such as CD-117 (C-Kit), DOG-1, S-100, and CKAE1-AE3, which helps exclude other types of neoplasms with which the differential diagnosis is established.⁸

Treatment consists of hepatic resection with intended R0 resection as the most widely accepted indication. There is no survival of more than 3 years with R1 resections.⁹ Adjuvant chemotherapy in PHL is not accepted because of the low rate of response, although in advanced stages tumorectomy associated with chemotherapy with doxorubicin and ifosfamide, or especially irinotecan, are the most accepted options.¹⁰ Another alternative is liver transplantation, although it is not recommended because of high rates of recurrence.

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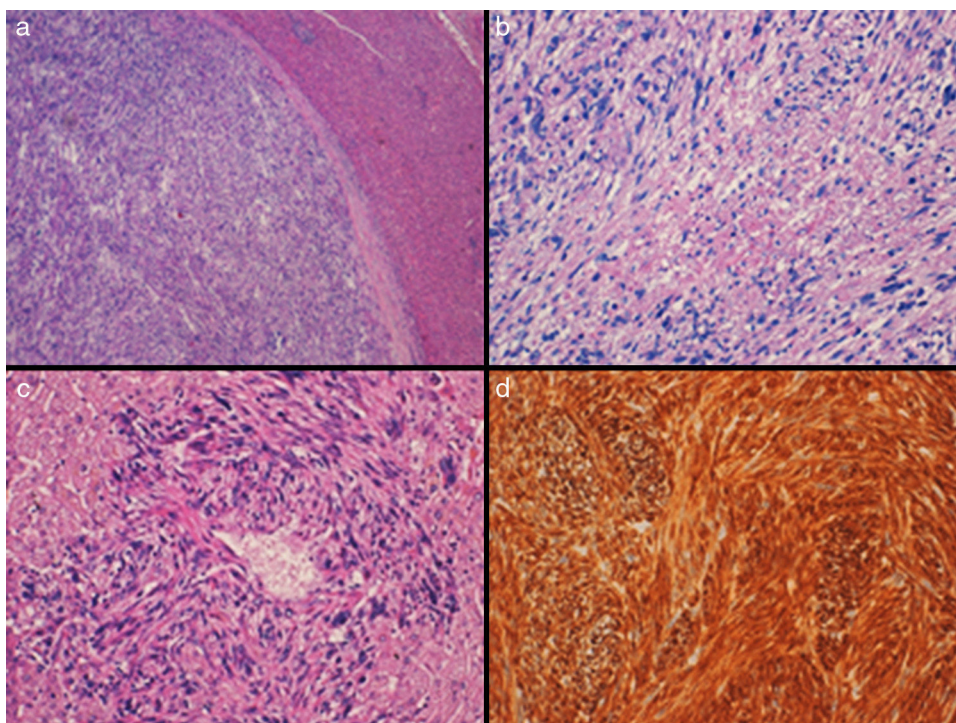


Fig. 1 – (a) Solid encapsulated neoplasm that is well outlined from the adjacent liver parenchyma (H-E, 4×); (b) It is constituted by intertwining bundles of spindle cells with marked nuclear atypia, atypical mitotic figures and a focus of necrosis (H-E, 10×); (c) Neoplastic cells emerging from the wall of a small hepatic vessel (H-E, 20×); (d) Intense positivity of tumor cells (desmin 20×).

HALS and totally laparoscopic surgery are presented as the most appropriate approaches in this type of tumors. In our case, the tumor was 7 cm, which is why we were more in favor of using HALS, since the introduction of the left hand allows for manual and ultrasound examination to locate preoperatively undetected lesions in the abdominal cavity, while also providing for the distinction between a primary tumor or the presence of metastatic disease or tumor dissemination throughout the abdominal cavity.

In conclusion, HALS is a safe approach in the treatment of PHL that enables R0 resection with good long-term survival results.

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Víctor López-López^{a,*}, Ricardo Robles^a, Belén Ferri^b,
Roberto Brusadin^a, Pascual Parrilla^a

*Corresponding author.

E-mail address: victorlopez@gmail.com
(V. López-López).

^aServicio de Cirugía General y del Aparato Digestivo, Hospital Clínico
Universitario Virgen de la Arrixaca, Instituto Murciano de
Investigación Biosanitaria (IMIB), Murcia, Spain

^bServicio de Anatomía Patológica, Hospital Clínico Universitario
Virgen de la Arrixaca, Murcia, Spain

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Autoimmune Pancreatitis: Differential Diagnosis With Pancreatic Adenocarcinoma[☆]



Pancreatitis autoinmune: diagnóstico diferencial con adenocarcinoma de páncreas

Autoimmune pancreatitis (AIP) is an unusual etiology of chronic pancreatitis, involved in approximately 6% of all cases.¹ It is a systemic disease that affects the pancreas as well as other organs, such as the bile duct, salivary glands, lymph nodes, kidneys and retroperitoneum.

It was first described in 1961 by Sarles et al.² Since then, our understanding of this entity has increased immensely, although some controversy still remains regarding the diagnostic and therapeutic criteria, as demonstrated by the 2010 Consensus Conference held in Honolulu.³

AIP is divided into 2 clinical-pathological forms: type I, or lymphoplasmocytic sclerosing pancreatitis (LPSP); and type II, or idiopathic duct-centric pancreatitis (IDCP). The basic histological difference is the presence of infiltration by IgG4 cells over 10 cells/field in type I, which is very low or null in

type II. Another important difference is the presence of granulocytic epithelial lesions in type II (GEL). Clinically, both types usually present as obstructive jaundice associated with a mass in the head of the pancreas (Table 1). It is therefore essential to perform a differential diagnosis with adenocarcinoma of the pancreatic head. According to Johns Hopkins Hospital data, the percentage of patients with AIP defined by the pathology study of a specimen from a resection due to suspected pancreatic adenocarcinoma is 2.4%.⁴

AIP is a disease that responds to treatment with corticosteroids, and accurate diagnosis is crucial to avoid unnecessary major surgeries. The diagnosis is based on five factors: histology, imaging, serology, involvement of other organs and response to corticoid treatment (HISORT criteria).⁶ Based on these criteria, the Mayo Clinic has proposed an algorithm to

Table 1 – Histological Clinical Aspects of Type I and II AIP.

	Type I AIP (LPSP)	Type II AIP (IDCP)
Clinical aspects⁵		
Sex	More frequent in males	More frequent in males
IgG4	Elevated in some 80%	17%
Extra pancreatic involvement	Biliary/retroperitoneal stenosis, retroperitoneal fibrosis, etc.	No
Inflammatory bowel disease	6%	16%
Histological aspects		
Infiltrate	Predominantly lymphoplasmocytic with eosinophils, rarely neutrophils	Predominantly lymphoplasmocytic with neutrophils in medium and small ducts
Pancreatic ducts	Periductal inflammation with no epithelial lesions	Periductal inflammation with damage to the duct epithelium due to granulocytes (GEL+ lesions)
Vein	Obliterative phlebitis	Rarely presents phlebitis
IgG4 immunostain	Abundant with more than 10 IgG4-positive cells per field	Limited or none
Peripancreatic fat	May be affected	Process usually limited to the pancreas
IDCP: idiopathic duct-centric pancreatitis; LPSP: lymphoplasmocytic sclerosing pancreatitis; AIP: autoimmune pancreatitis.		

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