

# World Journal of *Hepatology*

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## Primary hepatic leiomyosarcoma: A case report and literature review

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### Abstract

#### BACKGROUND

Primary hepatic leiomyosarcoma (PHL) is a rare tumor with a very low incidence of about 0.2%.

#### CASE SUMMARY

A 48-year-old diabetic, hypertensive, and morbidly obese female patient presented with a history of abdominal pain and weight loss for 2 mo. She had no history of fever, jaundice, or other liver disease(s). Clinical examination revealed a palpable mass in the epigastrium. Imaging evaluation with a contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis revealed an ill-defined enhancing hyper vascular hepatic mass of 9.9 cm × 7.8 cm occupying the left hepatic lobe with evidence of central necrosis, compression effect on the left hepatic vein, and partial wash-out on delayed images. On further workup, the maximum standardized uptake value on positron emission computed tomography scan was 6.4, which was suggestive of malignancy. The remaining part of the liver was normal without any evidence of cirrhosis. Ultrasound-guided biopsy of the mass showed smooth muscle neoplasm suggestive of leiomyosarcoma. After optimization for co-morbidities, an extended left hepatectomy was planned in a multidisciplinary team meeting. On intraoperative ultrasound, the left hepatic lobe was entirely replaced by a large tumor extending to the caudate lobe with a compression effect on the middle and left hepatic veins. Final histopathology showed nodular and whorled white tumor comprised of spindle/fascicular cells with moderate to severe pleomorphism and focal necrosis. The mitotic index was greater than 20 mitoses per 10 high-power fields. The resection margins were free of tumor. Immunohistochemistry (IHC) depicted a desmin-positive/caldesmon-negative/discovered on gastrointestinal stromal tumor 1-negative/cluster of differentiation 117-negative profile, confirming the definitive diagnosis as PHL.

#### CONCLUSION

This case report highlights the rare malignant mesenchymal hepatic tumor. To confirm PHL diagnosis, one requires peculiar histopathological findings with ancillary IHC confirmation. Management options include adequate/complete surgical resection followed by chemotherapy and/or radiotherapy.

**Key Words:** Leiomyosarcoma; Immunohistochemistry; Hepatectomy; Surgical resection; Case Report

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**Core Tip:** Primary hepatic leiomyosarcoma is an extremely rare tumor among all primary hepatic malignancies, with approximately 70 cases worldwide, including our case. The rare nature of the disease has precluded its underlying pathogenesis. Clinical manifestations are usually nonspecific, and tumors are generally asymptomatic until they significantly increase in size. They have a relatively poor prognosis and aggressive metastatic potential. The preferred type of treatment is surgical excision, which is sometimes combined with adjuvant chemotherapy and radiotherapy; however, little is known about their effectiveness because of the disease rarity. In-depth studies are needed to shed light on this uncommon clinical entity.

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## INTRODUCTION

Primary hepatic leiomyosarcoma (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. This manuscript presents a case report of a 48-year-old woman with PHL.

## CASE PRESENTATION

### Chief complaints

A 48-year-old woman presented to the outpatient clinic with complaints of abdominal pain associated with weight loss.

### History of present illness

The patient's symptoms started 2 mo prior with recurrent episodes of abdominal pain without any specific aggravating or relieving factor(s).

### History of past illness

She had no documented history of fever, jaundice, liver disease(s), blood transfusion, tattooing, or alcohol abuse.

### Personal and family history

Her co-morbidities included diabetes, hypertension, and obesity. She had no family history of carcinoma.

### Physical examination

On examination, there was a palpable mass in the epigastrium. The rest of the clinical examination was unremarkable, without any signs of disease elsewhere in the body.

### Laboratory examinations

Complete blood count, serum biochemical profile, coagulation profile, and carbohydrate antigen 19-9 were normal. Viral serology was nonreactive.

### Imaging examinations

An initial imaging evaluation with a contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis showed an ill-defined heterogeneous contrast-enhancing and hyper vascular mass

in the left lobe of the liver with extension up to the middle hepatic vein and compression effect on the left hepatic vein. The approximate measurement of the mass was 9.9 cm × 7.8 cm with central necrosis in the mass and partial wash-out on delayed images. The tumor encased the left portal vein branch at its point of bifurcation; however, the main portal vein and its right branch were not involved. No signs of hepatic cirrhosis were identified on imaging studies (Figure 1).

Positron emission tomography-CT (PET-CT) scan showed a maximum standardized uptake value (SUV) of the left hepatic mass of 6.4. The remaining liver had a standard baseline hepatic SUV (hepatic parenchymal baseline reference activity was up to 2.96 SUV).

No significant abnormality was seen above or below the diaphragm. In addition, the lungs, gastrointestinal (GI) tract, adrenal glands, kidneys, ovaries, and uterus did not show any evidence of primary disease or metastasis. No metastasis was found elsewhere in the body.

Ultrasound-guided biopsy was done, which showed cores of liver tissue with a proliferation of tumor cells having spindle-shaped nuclei with features of pleomorphism, up to 10 mitosis/10 high-power fields (HPFs), and necrosis. These findings suggested a spindle cell neoplasm raising a differential diagnosis of GI stromal tumor (GIST) and leiomyosarcoma. A panel of Immunohistochemical stains were applied that depicted a desmin-positive/caldesmon-negative/discovered on GIST1-negative (DOG1-)/cluster of differentiation 117-negative (CD117-) profile, thereby supporting the definitive diagnosis of PHL.

## MULTIDISCIPLINARY EXPERT CONSULTATION

The case was discussed for management discussion in GI and hepatobiliary multidisciplinary team (MDT) meetings. According to MDT recommendations, surgical resection of the liver mass was planned, as there was no clinical and radiological evidence of metastatic disease or unknown primary disease elsewhere in the body.

## FINAL DIAGNOSIS

The definitive preoperative diagnosis of the presented case was PHL in the absence of clinical and radiological evidence of metastatic disease or unknown primary disease elsewhere in the body.

## TREATMENT

A formal extended left hepatectomy was performed. Staging laparoscopy was negative for metastasis, followed by a traditional laparotomy using a Mercedes Benz incision. The total operating time was 540 min with a blood loss of 300 milliliters.

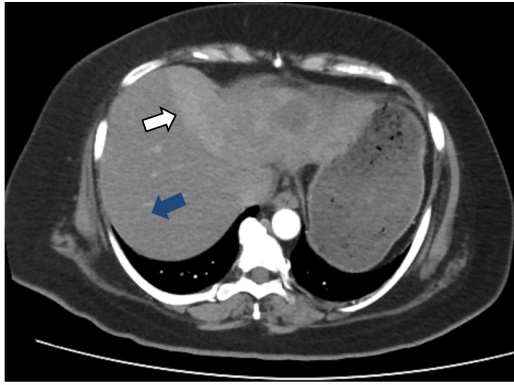
Intraoperatively, the left lobe of the liver was involved by a large tumor extending inferiorly to the caudate lobe (left side only) and superiorly up to the middle hepatic vein (MHV). MHV and its branches were sacrificed. An en bloc resection was achieved with an intact capsule (Figure 2). Resection margins were confirmed to be free of the tumor with the help of intraoperative ultrasound. Enlarged hilar nodes (hepatic artery lymph node) and periportal lymph node were also removed separately. No distant metastasis or aortocaval nodes were found per operatively.

### *Final diagnosis and histopathology*

Grossly, the left hepatectomy specimen measured 18 cm × 12.5 cm × 9.5 cm. Serial slicing of the specimen revealed a tumor (10.5 cm × 7.5 cm) with a nodular and whorled white appearance. The hepatic capsule was intact without any evident perforation. The microscopic evaluation revealed a cellular tumor with a fascicular array of tumor cells having spindle-shaped nuclei with a moderate-to-severe degree of cellular pleomorphism and more than 20 mitoses per 10 HPFs. Necrosis was focally seen. Lympho vascular invasion was not identified (Figures 3 and 4). All three regional lymph nodes (hepatic artery, periportal, and gallbladder neck) were negative for metastasis.

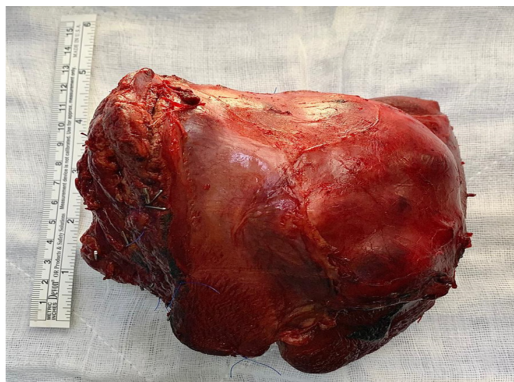
### *Immunohistochemical stain*

Desmin (clone DE-R11, Ventana; Roche Diagnostics, Indianapolis, IN, USA), a marker of smooth muscle differentiation, was positive in tumor cells; and caldesmon (clone E-89; Cell Marque Co., Rocklin, CA, USA) was negative in tumor cells. In addition, CD117 (clone EP-10; Leica, Wetzlar, Germany) and DOG1 (clone SP31; Cell Marque) were also negative in tumor cells, ruling out the possibility of the closest differential diagnosis of GIST (Figure 5).



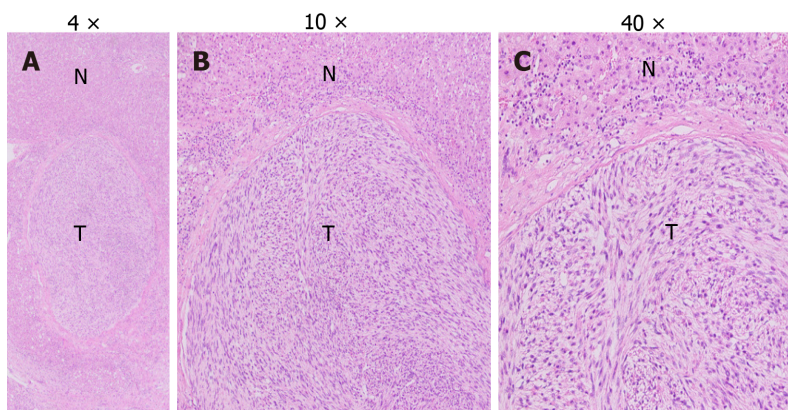
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**Figure 1** Contrast enhanced tomography scan image demonstrating a large enhancing heterogeneous mass in the left lobe of the liver (white arrow), surrounding normal the liver tissue (blue arrow).



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**Figure 2** Resected specimen.



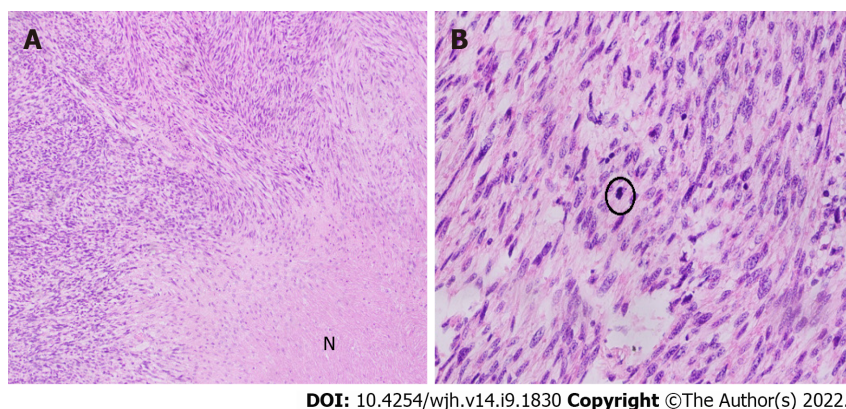
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**Figure 3** Leiomyosarcoma, subsequent resection specimen. A: Original magnification: 4 ×, scale bar: 100 μm; B: The above figures show liver parenchyma with a central nodule/tumor fascicular array of cells having spindle-shaped nuclei, moderate degree nuclear atypia (original magnification: 10 ×; scale bar: 100 μm); C: Mitoses marked as T, and normal liver hepatocytes marked as N (original magnification: 40 ×; scale bar: 100 μm).

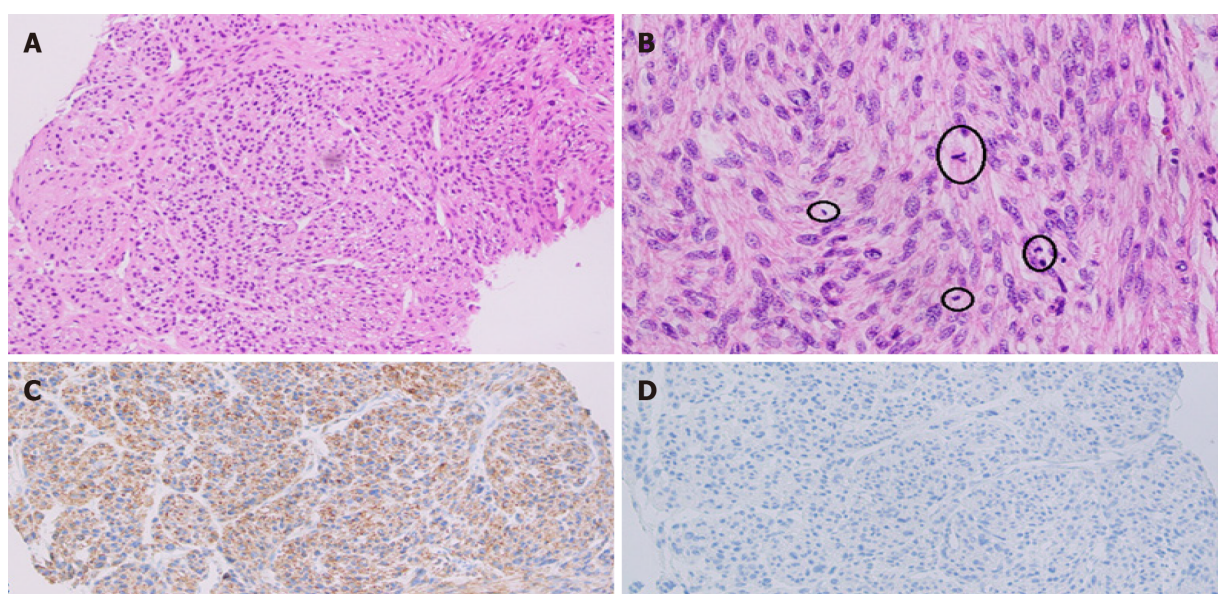
## OUTCOME AND FOLLOW-UP

The postoperative recovery of the patient was uneventful. She was discharged on the 6<sup>th</sup> postoperative day. The patient remained asymptomatic until the follow-up visit after 6 mo. No short- or long-term postoperative complications were reported. CT and PET-CT scans were performed during this follow-up visit, which showed no evidence of recurrence and metastases. The patient was later presented in the MDT of Sarcoma, where it was decided to keep the patient under bi-annual surveillance, and no





**Figure 4 Hematoxylin and eosin stain of leiomyosarcoma.** A: Hematoxylin and eosin stain of leiomyosarcoma, resected specimen, a section of the liver shows fascicular array of cells with spindle-shaped nuclei (Original magnification: 100 ×; scale bar: 100 μm); B: A moderate degree nuclear atypia, and mitoses (encircled) along with areas of necrosis marked as N (original magnification: 400 ×; scale bar: 100 μm).



**Figure 5 Fascicular array of cells with spindle-shaped nuclei.** A: Moderate degree nuclear atypia (original magnification: 100 ×; scale bar: 100 μm); B: Mitoses (encircled) (original magnification: 100 ×; scale bar: 100 μm); C: Immunostaining of desmin highlighting spindle-shaped cells (original magnification: 100 ×; scale bar: 100 μm); D: Caldesmon-negative (original magnification: 100 ×; scale bar: 100 μm).

adjuvant treatment was recommended.

## DISCUSSION

Sarcomas from the liver are rare, constituting only 1%-2% of all primary hepatic malignant tumors[1,2]. Leiomyosarcoma accounts for 8%-10% of all hepatic sarcomas, whereas other hepatic sarcomas including angiosarcoma, fibrosarcoma, liposarcoma, embryonal sarcoma, malignant fibrous histiocytoma, carcinosarcoma, and epithelioid hemangioendothelioma collectively comprise nearly 70%-80% of hepatic sarcomas[3]. Hepatic leiomyosarcomas are generally metastatic, arising from the GI tract, uterus, retroperitoneum, or lungs, requiring careful staging[2]. To the best of our knowledge, only about 70 cases, including case series, have been published internationally, including our patient (Tables 1 and 2)[1].

Leiomyosarcoma potentially originates from the smooth muscle cells in the round ligament, intrahepatic blood vessels, and bile ducts. Tumors arising from intrahepatic veins have a worse prognosis because they tend to progress to Budd-Chiari syndrome. The differentiation between benign leiomyoma and low-grade leiomyosarcoma is based on mitotic figures per HPF, although variation occurs according to the site of origin[3,4].

**Table 1 Management outcomes of published case reports of primary hepatic leiomyosarcoma**

Ref.	Patient age in yr	Sex	Risk factor/s	Management	Follow-up/ outcome
Jeong <i>et al</i> [12], 2008	49	F		Wedge resection	Died at 18 mo
Cioffi <i>et al</i> [2], 1996	62	M			Died at 20 mo
O'Leary <i>et al</i> [27], 1982	69	M			Alive at 24 mo
Baur <i>et al</i> [28], 1993	69	F		Surgery	Recurrence after 10 yr
Sato <i>et al</i> [29], 2000	62	F			Diagnosis at autopsy
Iordanidis <i>et al</i> [30], 2002	25	M		Surgery	Death at three mo
Lee <i>et al</i> [31], 2002	64	F		Right lobectomy + wedge resection of the left lobe	No evidence of disease at 24 mo
Shivathirthan <i>et al</i> [32], 2011	78	M		Chemotherapy, radiation	Death at 10 mo
Muranushi <i>et al</i> [33], 2019	62	M	Gastric gist post resection	Left hepatectomy+ resection of 3 other lesions	No evidence of disease
Watanabe <i>et al</i> [34], 1991	62	M		Lobectomy	No evidence of disease at 5 mo
Esposito <i>et al</i> [35], 2020	78	M		Left hepatectomy	No evidence of disease at 18 mo
Yoshikawa <i>et al</i> [36], 1977	58	F		Wedge resection	Death, 11 <sup>th</sup> postoperative day
Bloustein[37], 1978	12	F		Trisegmentectomy, chemotherapy	No evidence of disease at 6 yr
Maki <i>et al</i> [6], 1987	86	F		Surgery	No evidence of disease at 5 mo
Holloway <i>et al</i> [15], 1996	63	M		Conservative	
Soyer <i>et al</i> [19], 1996	67	F		Surgery	
Tsuji <i>et al</i> [10], 2000	68	M	Hepatitis c		Diagnosis at autopsy
Fujita <i>et al</i> [9], 2002	33	F	Prior renal transplant	Right posterior segmentectomy	No evidence of disease at 24 mo
Giuliente <i>et al</i> [11], 2009	26	M	Hodgkin's lymphoma	Right lobectomy + wedge resection of segment id	Death at 25 mo
Liang <i>et al</i> [21], 2009	44	F		Liver transplant	Death at 34 mo
Shivathirthan <i>et al</i> [32], 2011	67	M		Extended left hepatectomy + partial resection of segment 6	No evidence of disease at 9 mo
Tsai <i>et al</i> [5], 2013	Five mo	M		Chemotherapy + partial hepatectomy (segment vi) + adjuvant chemotherapy	No evidence of disease at 48 mo
Feritis <i>et al</i> [24], 2019	68	F	Hepatitis b	Left hepatectomy+ cholecystectomy, chemotherapy, target therapy, redo surgery for recurrence	Recurrence at 18 mo 2 <sup>nd</sup> recurrence at 21 mo died at 37 mo
Zhu <i>et al</i> [25], 2019	63	M		Unresectable tumor TACE (2011)	No evidence of disease at 82 mo
Vella <i>et al</i> [1], 2020	77	F		Right hepatectomy	No evidence of disease at 8 mo

F: Female; M: Male; TACE: Transcatheter arterial chemoembolization.

There is no apparent sex predilection with an approximate male-to-female ratio of 1:1 in the literature review of cases reported to date including our patient. Age at the time of presentation is variably reported with a range of 5 mo to 86 years (mean age of 51.3 years)[5,6].

No specific pathological causes of PHL have been identified to date, although the literature review shows an association with multiple etiological factors, including immunosuppression due to acquired immunodeficiency syndrome. Epstein-Barr virus infection is reported in two cases and a previous history of immunosuppression postrenal transplant in 1 case. The other reported associations include hepatitis C virus infection, exposure to thorotrast, and previously treated Hodgkin's lymphoma[7-11].



**Table 2 Management outcomes of published case series of primary hepatic leiomyosarcoma**

Ref.	Patient age in yr	Sex	Treatment	Follow up
Almoghy <i>et al</i> [22], 2004	58	F	Surgery + chemotherapy	Death at 4 mo
	63	F	Surgery + chemotherapy	Death at 12 mo
Watanabe <i>et al</i> [34], 2008	63	F		Diagnosis at autopsy
	49	F		Diagnosis at autopsy
Matthaei <i>et al</i> [14], 2009	19	F	Liver transplant	Death at 73 mo
	64	F	Surgery	No evidence of disease 181 mo
	53	F	Surgery	Death at 21 mo
	55	M	Surgery	No evidence of disease at 133 mo
	51	M	Liver transplant	No evidence of disease at 144 mo
	59	M	Surgery	Death at 45 mo
	63	F	Surgery	No evidence of disease at 133 mo
Shamseddine <i>et al</i> [23], 2010	25	F	Right lobectomy	Death at 22 mo
	39	M	Extended right lobectomy	Death at 19 d
	30	M	Right lobectomy + chemotherapy	No evidence of disease at 12 mo
Esposito <i>et al</i> [35], 2020	78	M	Left hepatectomy	No evidence of disease at 18 mo
	53	M	Right extended hepatectomy	Lung metastasis at seven mo death at 14 mo

F: Female; M: Male.

PHL usually pursues an indolent course being asymptomatic initially until they enlarge in size causing non-specific symptoms such as abdominal pain or discomfort, a palpable mass, fever, jaundice, anorexia, nausea or vomiting, and weight loss. The literature review revealed a single patient presenting as an emergency due to acute intraabdominal hemorrhage following tumor rupture. There is usually a single mass, although there are cases with multiple masses. Tumor size varies significantly ranging from 0.6 cm to 30 cm in the largest dimension (mean and median diameters of 10.3 cm and 9.1 cm, respectively). In our case, the tumor size was 10 cm in its largest dimension. The distribution of PHL within the liver segments and lobes also differs, with 2/3<sup>rd</sup> cases involving the right lobe and 1/3<sup>rd</sup> cases involving the left lobe. The case presented here involved the left lobe of the liver[6,11-16].

Histological examination usually shows intersecting bundles of spindle-shaped cells with hyperchromatic nuclei and mitotic figures. Immunohistochemistry shows positive results with smooth muscle actin, desmin, and vimentin, whereas cytokeratins, neuron-specific enolase, S-100 protein,  $\alpha$ -fetoprotein, CD34, CD117, and DOG1 were not expressed.

Ultrasonography usually shows hypoechoic or heterogeneous echogenic mass. CT findings generally describe a hypodense and sometimes heterogeneous mass with inhomogeneous and often peripheral enhancement after intravenous contrast administration, which may show regions of cystic degeneration, demonstrated in our case[17]. Ferrozzi *et al*[18] reported that hemorrhagic necrosis, along with cystic degeneration and necrosis, are likely to be secondary to tumor growth. Magnetic resonance imaging characteristically displays homogeneous or heterogeneous hypointense T1-weighted images and hyperintense T2-weighted images[19]. PET-CT scan SUV max can be correlated with the tumor size, tumor-node-metastasis staging, and histology subtype[20]. In our case, the SUV max of the primary tumor was 6.4, with a baseline hepatic parenchymal SUV of 2.96.

Therapeutic options vary depending upon the tumor size and/or stage on initial presentation. Hepatic resection (wedge resection, segmentectomy, lobectomy, or extended hepatectomy with the intention of R0 resection) remains the only potentially curative surgical option for non-metastatic including our case. However, 4 patients with tumors confined within the liver had liver transplantation [14,21].

Some authors have also reported that adjuvant chemotherapy consisted of various drugs, including doxorubicin and ifosfamide, which help to attain prolonged survival after complete resection. In addition, three cases have been treated with radiotherapy as part of a combined adjuvant treatment along with chemotherapy[5,22-24].

Transarterial chemoembolization and transarterial infusion of epirubicin and carboplatin were also reported in individual cases as treatment modalities for PHL[25].

The median survival is 37.5 mo with a 5-year survival rate of about 40%; two large case series reported 67% disease-specific survival at 5 years after R0 resection. Matthaei *et al*[14] in a case series, reported more than 10 years of survival after hepatectomy. Chi *et al* reported a median overall survival of 19 mo with 1-, 2-, and 5-year survival rates of 61.2%, 41.1%, and 14.5% respectively[26].

Age is another crucial predictive factor for prognosis, with patients below 50 years achieving better survival. Currently, no effective therapeutic options have been reported for unresectable PHL. Fujita *et al*[9] reported a patient with metastatic leiomyosarcoma surviving 3 mo after diagnosis, who received only palliative and conservative therapy.

## CONCLUSION

In conclusion, PHL is a rare malignant disease with often delayed presentation, relatively poor prognosis, and aggressive metastatic potential. The most preferred management options include surgical excision combined with chemotherapy and radiotherapy. However, very little is known about the efficacy of available therapeutic options because of the rarity of the disease. Therefore, in-depth studies are required to assess its causative, prognostic, and predictive parameters.

## FOOTNOTES

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