Case Report

Journal Homepage: http://crcp.tums.ac.ir

A Rare Case of Primary Hepatic Leiomyoma

Elham Zare, Ali Beheshti Namdar*

Department of Internal Medicine, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.

Use your device to scan and read the article online

Citation Zare E, Beheshti Namdar A. A Rare Case of Primary Hepatic Leiomyoma. Case Reports in Clinical Practice. 2024; 9(5): 203-207. DOI:10.18502/crcp.v9i5.18451

Running Title Primary Hepatic Leiomyoma

Article info:
Received: August 23, 2024
Revised: September 17, 2024
Accepted: October 26, 2024

Keywords:
Leiomyoma; Liver; Gastrointestinal systems

ABSTRACT

Leiomyomas are most commonly encountered in the genitourinary and gastrointestinal systems, with primary hepatic leiomyomas (PHL) being a rare occurrence. We present the case of a 62-year-old man with an incidentally discovered voluminous lesion in the left lobe of the liver, initially suspected to be a hemangioma. The patient was referred for further evaluation, during which advanced imaging modalities were utilized for accurate diagnosis, and tissue biopsies were obtained. A definitive diagnosis of PHL was established through immunohistochemistry based on pathological findings.

Introduction

Leiomyomas are mesenchymal tumors most commonly found in the genitourinary and gastrointestinal systems. Primary hepatic leiomyomas (PHL) are extremely rare, accounting for less than 1% of all benign liver tumors [1, 2]. These tumors are believed to originate from the smooth muscle layer of the biliary tree or the intrahepatic blood vessel wall [1, 3]. Due to their rarity and the absence of specific clinical or radiological features, PHL often presents as a diagnostic challenge.

Here, we present the case of a 62-year-old male with an incidentally discovered voluminous lesion in the left lobe of the liver, initially suspected to be a hemangioma. The lesion was ultimately diagnosed as PHL through immunohistochemistry and pathological findings. This case is particularly noteworthy given the patient's history of renal transplantation and subsequent immunosuppressive therapy, a factor frequently associated with the development of PHL [4]. In addition, we provide a concise review of the clinical presentation, diagnostic approach, treatment options, and potential future research directions for this rare entity.

Case Presentation

A 62-year-old male patient was hospitalized for the assessment of acute kidney injury. He was referred to our facility for further evaluation following the incidental discovery of a 40 mm circular isoechoic lesion in the eighth segment of the right hepatic lobe, detected via an abdominopelvic ultrasound. The patient reported no symptoms such as abdominal discomfort, weight loss, nausea, vomiting, or anorexia.

* **Corresponding Author:**
Ali Beheshti Namdar
Address: *Department of Internal Medicine, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran.*
E-mail: *beheshtia@mums.ac.ir*

The patient's medical history included renal transplantation in 2016, and he had been receiving immunosuppressive therapy since then (including immunoral, prednisolone, cellcept, and insulin). Initial laboratory tests, including liver function assessments and complete blood counts, showed results within normal limits. Tumor markers and viral marker tests, including alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA199), and cancer antigen 125 (CA125), along with Epstein-Barr virus (EBV), hepatitis C virus (HCV), hepatitis B virus (HBV), and human immunodeficiency virus (HIV), were all negative.

A triphasic computed tomography scan was subsequently performed, demonstrating characteristics indicative of a hemorrhagic adenoma, and a stromal tumor was suggested. Considering the patient's immunosuppressed state, a US-guided percutaneous liver biopsy was performed for further evaluation.

Histological examination of the specimen revealed a benign mesenchymal tumor within the hepatic parenchyma. Immunohistochemical testing confirmed positive expression for SMA and desmin in cell proliferation. These pathological findings established the diagnosis of PHL (Figure 1). Additionally, an abdominopelvic CT scan ruled out involvement of other areas or metastatic disease.

Discussion

Primary hepatic leiomyomas (PHL) are exceptionally rare mesenchymal tumors of the liver, believed to originate from the smooth muscle layer of the biliary tree or the intrahepatic blood vessel wall [1, 2]. They account for less than 1% of all benign liver tumors [3]. Despite their rarity and associated diagnostic challenges, PHL represents a compelling topic for review. This paper aims to provide a concise overview of their clinical presentation, diagnostic approach, treatment options, and potential areas for future research.

Globally, only a few cases of PHL have been reported. These tumors typically occur in middle-aged adults, with no clear gender predilection. Due to their rarity, there is limited data from long-term follow-up studies to evaluate outcomes and recurrence risks.

PHL are more frequently reported in immunosuppressed patients, such as the case of our patient

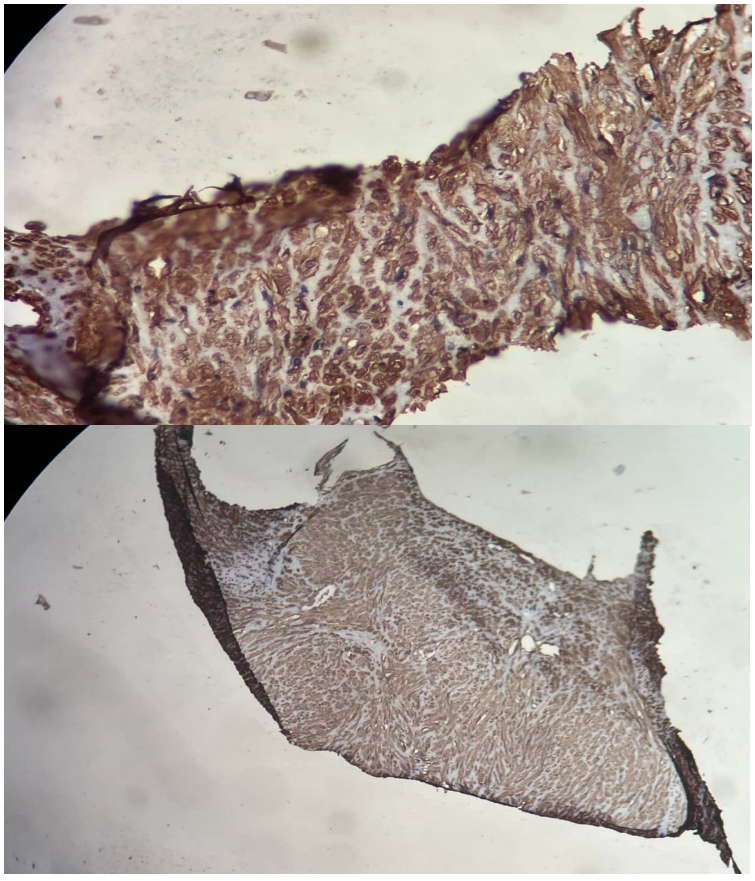

Fig. 1. At the immunohistochemical examination, the lesion showed strong reactivity for smooth muscle actin (SMA) (+), desmin (+) but not for Dog-1 (−), CD117 (−), and Beta-catenin (−) (Fig1. A, B). Based on the diagnostic criteria for PHL, the patient was diagnosed with PHL.

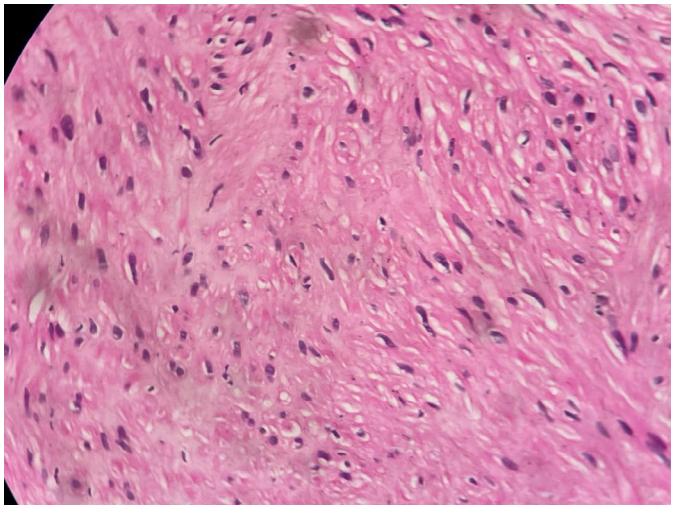

Fig. 2. At histological examination, it was composed of bland spindle cells without atypia in a fascicular pattern. There were no foci of necrosis or mitosis

who has been receiving immunosuppressive therapy following transplantation [4]. Despite the established association between immunosuppression and PHL, approximately 55 cases have been documented in the literature since 1926; of these, half were immunocompetent [2, 5].

Furthermore, positive EBV infection test results have been reported in the majority of PHL cases, suggesting a potential role for EBV in the pathogenesis of smooth muscle tumors, particularly under immunosuppressive conditions [4, 6–8]. However, in our patient, EBV test results were negative, as observed in some previously reported cases. This highlights that the etiology of PHL is multifactorial and remains poorly understood [3, 4, 6, 9, 10].

Clinically, one-third to half of patients with primary hepatic leiomyomas remain asymptomatic until a significant increase in tumor size occurs [4, 3]. These tumors are often discovered incidentally during imaging performed for unrelated reasons [1, 2, 11], as observed in our case. When symptoms do occur, they are typically non-specific and resemble those of other liver masses, including vague abdominal pain, a sense of fullness, or a palpable mass—features common in liver malignancies [11, 12].

In rare cases, PHL may present with liver function abnormalities [13]. Screening for tumor markers such as alpha-fetoprotein, carbohydrate antigen 19-9, and carcinoembryonic antigen is generally negative [5, 12, 14]. In our case, all tumor marker tests yielded negative results, and liver function tests remained within normal ranges.

There are no specific radiological findings for primary hepatic leiomyomas . However, imaging modalities play a crucial role in the initial diagnosis [1, 15]. Generally, leiomyomas are described as markedly hypervascular [11]. On ultrasonography, leiomyomas typically appear as well-defined, hypoechoic lesions with varying degrees of heterogeneity [15–17]. On CT scans, lesions are hypodense with enhancement during arterial and portal phases but without washout in late phases [17, 18].

MRI may reveal lesions with hyperintensity on T1-weighted images and hyperintensity on T2-weighted images, along with non-homogeneous contrast uptake [19–21]. However, PHL can mimic other, more common hepatic neoplasms [22], and is often indistinguishable from metastatic leiomyomas or other hepatic spindle cell tumors based solely on imaging [1, 12, 14, 22, 23]. In cases like ours, a biopsy was indispensable, as imaging initially suggested suspicious findings such as liver adenoma and hemorrhagic lesions [23].

The diagnostic criteria for PHL recommended by Hawkins include two key points: first, the tumor must originate from leiomyocytes; second, there should be no evidence of leiomyoma in other body sites, such as the gastrointestinal tract [24].

Histopathological examination of a biopsy specimen is considered the gold standard for definitive diagnosis [12,25]. Leiomyomas are characterized by smooth muscle cells; in the absence of atypical features, tumor cells typically exhibit smooth muscle morphology and stain positive for markers such as desmin, smooth muscle actin, and caldesmon [1]

(Figure 2). Immunostaining for smooth muscle markers can further support the diagnosis [3,26]. Differential diagnosis may include other liver tumors with smooth muscle differentiation, such as inflammatory myofibroblastic tumors or hepatic leiomyosarcomas. Molecular studies may provide additional insights into the genetic profiles of PHL by using techniques like next-generation sequencing (NGS) or fluorescence in situ hybridization (FISH) [12].

Due to PHL's unique nature, gathering sufficient data for comprehensive management guidelines remains challenging. The primary treatment modality for PHL is surgical resection, as complete excision offers the best chance for curative outcomes [12,14]. However, the presence of underlying liver disease, as well as the tumor's size and location [26], may impact the surgical approach. Minimally invasive techniques are utilized in some cases to reduce postoperative morbidity and enhance recovery.

Adjuvant therapies have shown limited efficacy in treating primary liver leiomyoma and are generally reserved for unresectable cases or recurrences. The prognosis of PHL is generally favorable following complete surgical resection, with low rates of recurrence or metastasis reported in the literature. However, long-term monitoring is recommended to detect disease progression or recurrence. Due to the limited number of similar cases reported in the medical literature, it is challenging to draw definitive conclusions about its management approach. We have attempted to provide a comprehensive review.

Conclusion

PHL remains rare in the realm of hepatic lesions. Further research is needed to elucidate our understanding of the underlying molecular characteristics, pathogenesis, and optimal management strategies for this enigmatic tumor, as well as to explore novel minimally invasive techniques and targeted therapies.

Ethical Considerations

Ethical approval

Case reports are exempt from ethical approval in our institution.

Funding

There are no sources of funding.

Conflict of Interests

The authors have no conflict of interest to declare.

Consent

The patient provided written informed consent to publish this case report and accompanying images. The editor-in-chief of this journal can review a copy of the written consent upon request.

References

[1] Zimmermann, A. (2017). Leiomyomatous Tumors of the Hepatobiliary Tract. In: Tumors and Tumor-Like Lesions of the Hepatobiliary Tract. Springer, Cham. https://doi.org/10.1007/978-3-319-26956-6_63

[2] Jia B, Jin Z, Gao P, Liu Y. Primary hepatic leiomyoma in a Chinese female patient without underlying disease: a case report. BMC Surg. 2019 Oct 7;19(1):140. https://doi.org/10.1186/s12893-019-0598-1

[3] Djokic M, Hadzialjevic B, Luzar B, Trotovsek B. Primary Hepatic Leiomyoma in a Healthy Middle-Aged Woman: Literature Review and Case Report. Front Surg. 2021 Jun 14;8:691674. https://doi.org/10.3389/fsurg.2021.691674

[4] Nwankwo EC Jr, Burton SJ, Brotherton T, Carpenter D, Qureshi K. Asymptomatic Hepatic Leiomyoma in an Immunocompetent Middle-Aged Woman. ACG Case Rep J. 2022 Nov 24;9(11):e00899. https://doi.org/10.14309/crj.0000000000000899

[5] Coletta D, Parrino C, Nicosia S, Manzi E, Pattaro G, Oddi A, et al. Primary leiomyoma of the liver in an immunocompetent patient. Intractable Rare Dis Res. 2020 Nov;9(4):251-255. https://doi.org/10.5582/irdr.2020.03075

[6] Miyahara J, Shimazu K, Saito A, Saito M, Fukuda K, Yoshida T, et al. Clinical Course of a Rare Epstein-Barr Virus-Associated Smooth Muscle Tumor and Its Genomic Analysis. Case Rep Oncol. 2023 Aug 4;16(1):577-584. https://doi.org/10.1159/000530383

[7] Cheuk W, Li PC, Chan JK. Epstein-Barr virus-associated smooth muscle tumour: a distinctive mesenchymal tumour of immunocompromised individuals. Pathology. 2002 Jun;34(3):245-9. https://doi.org/10.1080/00313020220131309

[8] Kanazawa N, Izumi N, Tsuchiya K, Sakurai K, Hamano K, Itakura J, et al. A case of primary leiomyoma of the liver in a patient without evidence of immunosuppression. Hepatol Res. 2002 Sep;24(1):80. https://doi.org/10.1016/s1386-6346(02)00014-1

[9] Perini MV, Fink MA, Yeo DA, Carvalho CA, Morais CF, Jones RM, et al. Primary liver leiomyoma: a review of this unusual tumour. ANZ J Surg. 2013 Apr;83(4):230-3. https://doi.org/10.1111/j.1445-2197.2012.06257.x

[10] Vyas S, Psica A, Watkins J, Yu D, Davidson B. Primary hepatic leiomyoma: unusual cause of an intrahepatic mass. Ann Transl Med. 2015 Apr;3(5):73. https://doi.org/10.3978/j.issn.2305-5839.2015.03.40

[11] Santos I, Valls C, Leiva D, Serrano T, Martinez L, Ruiz S. Primary hepatic leiomyoma: case report. Abdom Imaging. 2011 Jun;36(3):315-7. https://doi.org/10.1007/s00261-010-9648-y

[12] Omiyale AO. Primary leiomyoma of the liver: a review of a rare tumour. HPB Surg. 2014;2014:959202. https://doi.org/10.1155/2014/959202

[13] M Yanase 1, H Ikeda, I Ogata, A Ohno, A Moriya, N Miura, et al. Primary smooth muscle tumour of the liver encasing hepatobiliary cystadenoma without mesenchymal stroma. Am J Surg Pathol. 1999;23(7):854–9. https://doi.org/10.1097/00000478-199907000-00016

[14] Belli G, Ciciliano F, Lannelli A, Marano I. Hepatic resection for primary giant leiomyoma of the liver. HPB (Oxford). 2001;3(1):11-2. https://doi.org/10.1080/136518201753173692

[15] Tan W., Wu G., Zheng C. Imaging findings of primary hepatic leiomyoma. *Chin-Ger J Clin Oncol*. 2009;8(3):134–136. https://doi.org/10.1007/s10330-008-0163-3

[16] Urizono Y, Ko S, Kanehiro H, Hisanaga M, Aomatsu Y, Nagao M, et al. Primary leiomyoma of the liver: report of a case. Surg Today. 2006;36(7):629-32. https://doi.org/10.1007/s00595-006-3204-y

[17] Qian XH, Yan YC, Gao BQ, Wang WL. Prevalence, diagnosis, and treatment of primary hepatic gastrointestinal stromal tumors. World J Gastroenterol. 2020 Oct 28;26(40):6195-6206. https://doi.org/10.3748/wjg.v26.i40.6195

[18] Gandhi SN, Brown MA, Wong JG, Aguirre DA, Sirlin CB. MR contrast agents for liver imaging: what, when, how. Radiographics. 2006 Nov-Dec;26(6):1621-36. https://doi.org/10.1148/rg.266065014

[19] Raber EL, Cheng AL, Dong WF, Sutherland F. Primary hepatic leiomyoma in a transplant patient: characterization with magnetic resonance imaging. Transplantation. 2012 Jan 27;93(2):e4-5. https://doi.org/10.1097/tp.0b013e31823ec0cf

[20] Sadler M, Mays WL, Albert P, Javors B. Hepatic leiomyomas in two adult patients with AIDS: intravenous contrast-enhanced CT and MR imaging. Emerg Radiol. 2002 Sep;9(3):175-7. https://doi.org/10.1007/s10140-002-0214-y

[21] Marin D, Catalano C, Rossi M, Guerrisi A, Di Martino M, Berloco P, Passariello R. Gadobenate dimeglumine-enhanced magnetic resonance imaging of primary leiomyoma of the liver. J Magn Reson Imaging. 2008 Sep;28(3):755-8. https://doi.org/10.1002/jmri.21519

[22] Keramida M, Machairas N, Dorovinis P, Sotiropoulos G, Machairas A. A primary leiomyoma of the liver mimicking a giant hepatic hemangioma: report of a rare case. HPB (Oxford). 2023;25:S400. http://dx.doi.org/10.1016/j.hpb.2023.07.419

[23] Sousa HT, Portela F, Semedo L, Furtado E, Marinho C, Cipriano MA, Leitão MC. Primary leiomyoma of the liver: accurate preoperative diagnosis on liver biopsy. BMJ Case Rep. 2009;2009:bcr09.2008.0898. https://doi.org/10.1136/bcr.09.2008.0898

[24] Hawkins EP, Jordan GL, McGavran MH. Primary leiomyoma of the liver. Successful treatment by lobectomy and presentation of criteria for diagnosis. Am J Surg Pathol. 1980 Jun;4(3):301-4.

[25] Guy CD, Yuan S, Ballo MS. Spindle-cell lesions of the liver: diagnosis by fine-needle aspiration biopsy. Diagn Cytopathol. 2001 Aug;25(2):94-100. https://doi.org/10.1002/dc.2011

[26] Colagrande A, Cazzato G, Fedele S, Andriola V, Ingravallo G, Resta L, Vincenti L. A Unique Case of the Transformation of a Hepatic Leiomyoma into Leiomyosarcoma with Pancreatic Metastases: Review of the Literature with Case Presentation. Reports. 2022; 5(1):2. https://doi.org/10.3390/reports5010002