American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2023; 24: e938645
DOI: 10.12659/AJCR.938645

Received: 2022.10.13
Accepted: 2023.01.09
Available online: 2023.01.22
Published: 2023.02.19

Primary Hepatic Angioleiomyoma: A Case Report

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Data Interpretation D
Manuscript Preparation E
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Financial support: None declared
Conflict of interest: None declared

Patient: Male, 60-year-old
Final Diagnosis: Angioleiomyoma
Symptoms: Pain
Clinical Procedure: —
Specialty: Pathology

Objective: Rare disease

Background: Primary hepatic angioleiomyoma is a rare mesenchymal tumor that is characterized by blood vessels and smooth muscle. Herein, we report an extremely rare case of primary hepatic angioleiomyoma and discuss the clinicopathological features.

Case Report: A 60-year-old Mongolian man was diagnosed with a hepatic tumor in the second and third segments of screening in 2012. It had been under control by a physician for 10 years. The patient had discomfort and vague pain in the right side of the abdomen since April 2022. Hepatitis virus markers (hepatitis B and hepatitis C) were negative. Plain computed tomography revealed an 80-mm solitary liver lesion in the left lobe with well-defined margins and heterogeneous enhancement. A left hepatectomy was performed in May 2022. The cut surface of the tumor showed a grayish-white, elastic, hard mass with a diameter of 50×80 mm. Histological findings of the tumor revealed that it was clearly demarcated from the surrounding liver tissues with relatively clear boundaries showing thick, muscle-coated blood vessels with perivascular smooth muscle bundles. Immunohistochemical staining showed that the smooth muscle cells were strongly diffuse and positive for smooth muscle actin.

Conclusions: Clinically, primary hepatic angioleiomyoma should be distinguished from other types of liver tumors, especially liver cancer. In combination with our long-term observation and other case reports, we recommend general follow-up if the preoperative pathological diagnosis can be confirmed and the patient has no other symptoms.

Keywords: Liver Neoplasms • Pathology • Prognosis

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Background

Angioleiomyoma is a rare mesenchymal tumor that typically arises in the dermis or subcutis and is characterized by including blood vessels and smooth muscle [1,2]. Angioleiomyoma occurs from muscular tissue within the blood vessel wall, most often from the walls of veins. Angioleiomyoma is mostly found in patients aged 30 to 60 years, has a higher incidence in women, and is located in the lower extremity [3]. Although angioleiomyomas have been found in different organs [4-6], there are only a few case reports of primary hepatic angioleiomyoma (PHA). Herein, we report an extremely rare case of PHA and discuss clinicopathological features.

Case Report

The patient was a 60-year-old man. A hepatic mass was found during a workplace health check in 2012. Histological diagnosis was not performed because the patient's consent was not obtained. At the health check, a clinician had performed ultrasound and laboratory analysis of transaminases (alanine aminotransferase, aspartate aminotransferase) and alpha-fetoprotein every year for 10 years. The patient had vague pain in the right side of the abdomen since April 2022. The patient's previous medical history included sixth and seventh ribs fracture at 29 years, hydrocele of the testis at 35 years, daily alcohol consumption, and smoking (30 cigarettes a day) from 20 to 60 years old. No abnormalities were found in physical, hematological, and biochemical examination, including tumor markers such as αfetoprotein and carcinoembryonic antigen. Hepatitis virus markers (hepatitis B and hepatitis C) were negative. Plain computed tomography (CT) revealed an 80-mm solitary liver lesion in the left lobe (segments II to III) with well-defined margins and heterogeneous enhancement (**Figure 1**). There were no focal lesions in the spleen, pancreas, and kidneys. Histological diagnosis was not performed because the patient's consent was not obtained. The cut surface of the tumor showed a 50×80 mm, grayish-white, and elastic hard mass.

No capsule was found around the tumor (**Figure 2**). Histological findings of the tumor revealed that it was clearly demarcated from the surrounding liver tissues with relatively clear boundaries, presenting with thick muscle-coated blood vessels with intervascular smooth muscle bundles (**Figure 3A-3C**). Necrosis, hemorrhage, cyst formation, and pleomorphism of the nucleus were absent; however, a few mitotic figures were seen. No sclerosing cholangitis was observed in the hepatic bile ducts of the liver tissues surrounding the liver. Immunohistochemically, the perivascular spindle cells of the tumor showed immunostaining for smooth muscle actin (**Figure 4A**), and the vascular endothelium was positive for CD34 (**Figure 4B**), weakly positive for desmin (**Figure 4C**), and negative for HMB-45 and S-100 (**Figure 4D, 4E**).

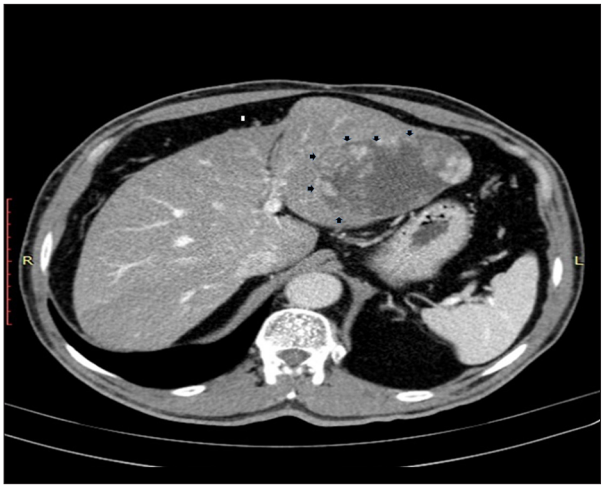

Figure 1. Plain computed tomography revealed a solitary liver lesion in the left lobe with well-defined margins and heterogeneous enhancement (black arrowheads).

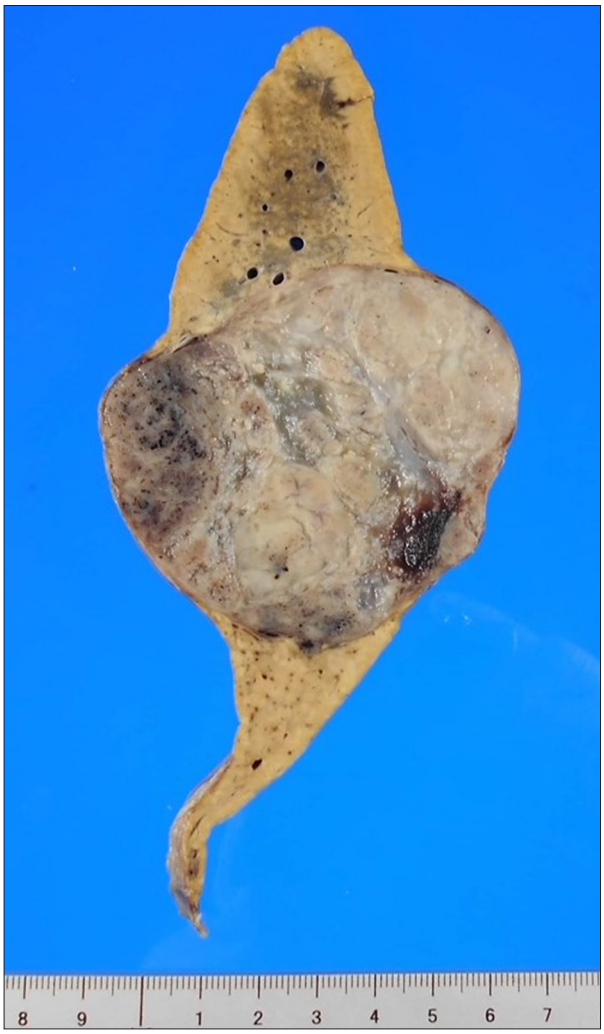

Figure 2. The liver mass is seen after surgery. Tumors are well-circumscribed and have grayish-white cut surfaces.

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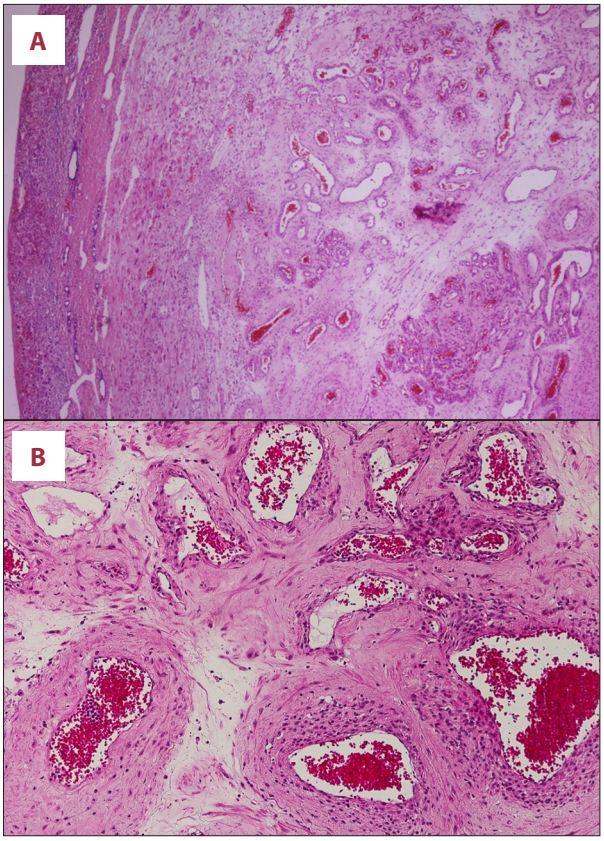

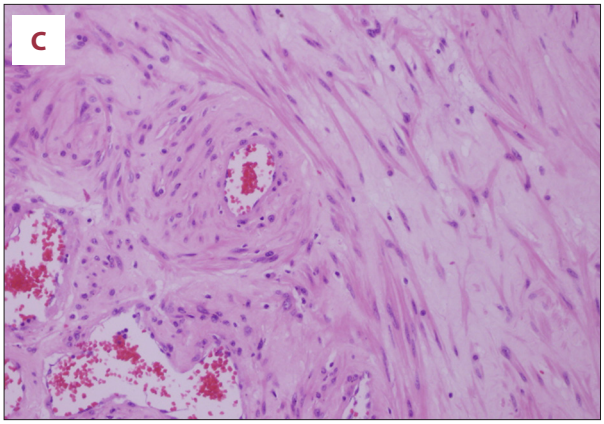

Figure 3. At low magnification, a well-circumscribed tumor in the liver is seen (**A**, hematoxylin and eosin, 40×). At high magnification, presenting with thick muscle-coated blood vessels (**B**, hematoxylin and eosin, 200×) with inter- and perivascular smooth muscle bundles (**C**, hematoxylin and eosin, 200×).

Discussion

Angioleiomyoma occur on the neck, head, and limbs, are mostly small size, and most often present as a subcutaneous tumor that can be painful and/or tender [2,4,7]. The clinical presentation of visceral angioleiomyomas is correlated with tumor location and size [8-11]. Therefore, PHA can also be suggested by abdominal pain or a palpable mass. The present patient had definite clinical symptoms of discomfort and was admitted for abdominal pain.

Recently, imaging findings of angioleiomyoma in various organs have been reported. Hu et al reported angioleiomyoma of the pulmonary artery [11]; contrast-enhanced CT of the chest revealed a well-demarcated mass. Bagheri et al reported angioleiomyoma of the orbita, which showed an oval mass by CT [12]. Furthermore, Sun et al reported uterine angioleiomyoma involvement into the veins [13]. They commented that preoperative CT diagnosis of angioleiomyoma was extremely difficult, as previous case reports did not provide common CT characteristics. The ratio of smooth muscle cells, fibrous tissue, and intratumoral vascular lumen morphology can lead to nonspecific imaging of PHA. Therefore, due to atypical clinical symptoms, no specificity in laboratory examination, and the lack of characteristic imaging findings, angioleiomyoma can be misdiagnosed as a different liver tumor.

Preoperative diagnosis of PHA is very difficult; thus, diagnosis often requires postoperative or biopsy pathological analysis. Histologically, PHAs are composed of numerous smooth muscle fiber cells, mixed with abundant thick-walled vascular walls without common morphologic characteristics of malignant tumor. Histologically, the present patient had a well-demarcated, encapsulated tumor characterized by thick muscle-coated vessels with intervascular smooth muscle bundles [4-6,14,15]. In most of the previous studies, there was no significant mitotic activity in angioleiomyoma in many organs [14,15]. In the present study, we observed mitotic figures; however, there are no definitive criteria of mitosis to differentiate between benign and malignant angioleiomyoma. Furthermore, necrosis, hemorrhage, cyst formation, and nuclear pleomorphism were absent. Therefore, we concluded that our case was benign angioleiomyoma.

Angioleiomyomas are divided into 3 histologic variants (solid, venous, and cavernous), based on the predominant histologic findings [3]. Solid-type angioleiomyomas are characterized by closely compacted smooth muscle bundles and intervening small, thin-walled, slit-like vascular structures. Venous-type tumors are often found in areas with thick muscle walls, in which there is smooth muscle of the vascular wall with intervascular smooth muscle bundles. Cavernous-type tumors are characterized by dilated vascular structures with small amounts of smooth muscle, and the muscular walls of these vessels are difficult to distinguish from intervascular smooth muscle bundles. Our case consisted of thick muscle-coated blood vessels with intervascular smooth muscle bundles; therefore, it was considered to be of the venous type based on histological findings, which may have contributed to the CT findings.

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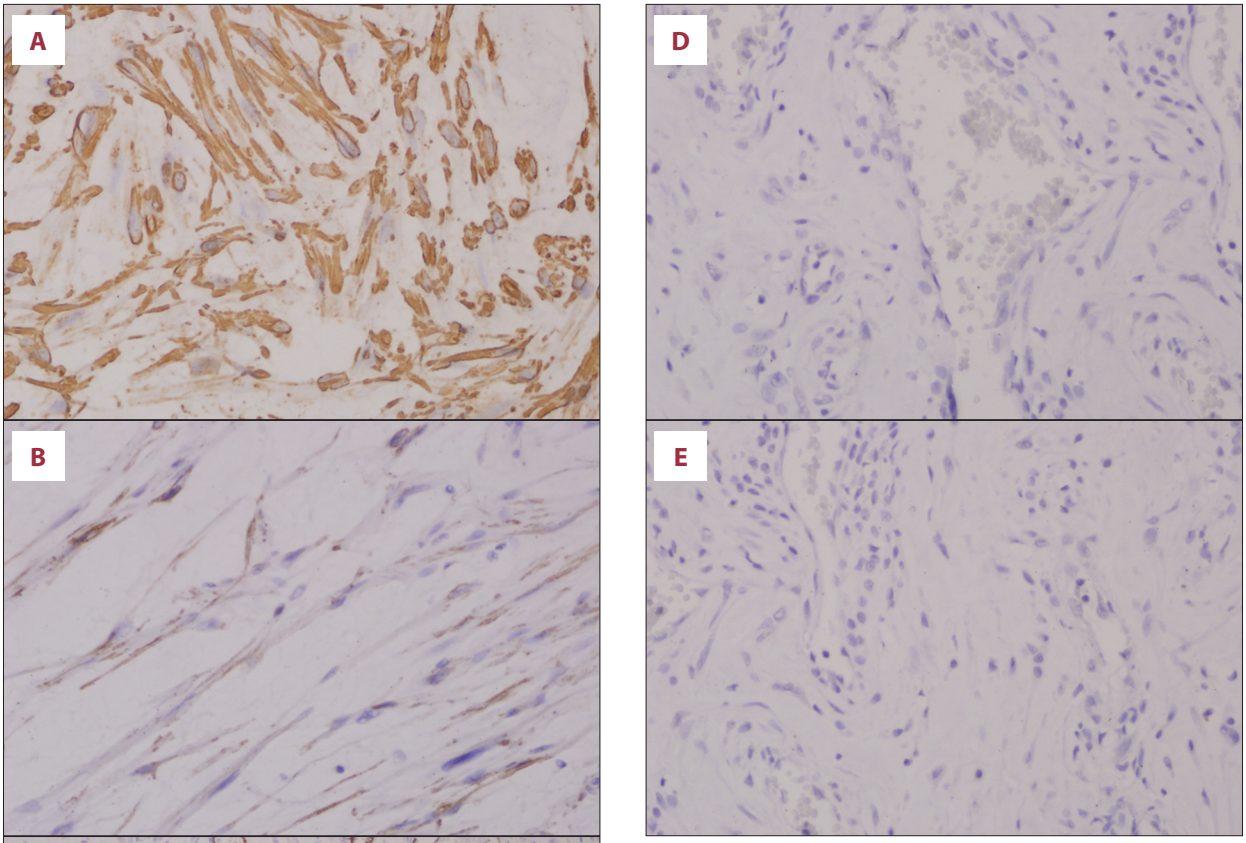

Figure 4. Immunohistochemically, the tumors are positive for small muscle actin (**A**, 400×) and weakly positive for desmin (**B**, 400×). Perivascular spindle cells of the tumor were negative for CD34 (**C**, 200×), HMB-45 and S-100 (**D, E**, 400×).

We performed a differential diagnosis with angiomyolipoma, myopericytoma, and solitary fibrous tumor. Angiomyolipoma is a rare tumor that is composed of blood vessels, smooth muscle, and adipose tissue of different proportions. Angiomyolipoma is divided into 4 groups depending on the dominant tissue [16]. The hybrid type is the most common and consists of similar proportions of smooth muscle and adipose tissue within the tumor. Smooth muscle is the dominant component within the myoma-type tumors, adipose tissues are the dominant component of the lipoma-type, and vascular tissue is the dominant component of the hemangioma-type; however, all types contain adipose components in the tumor. In our case, we did not observe adipose components in hematoxylin and eosin staining. Immunohistochemical staining can be performed to increase accuracy of histological diagnosis for angiomyolipoma. HMB-45, a specific immunohistochemical staining for human melanocytic tumor, only reacts with angiomyolipoma and hepatoblastoma in liver tissue [16]. Together, with there being no expression of S-100, we excluded the diagnosis of angiomyolipoma in the present case.

Myopericytoma is a well-circumscribed tumor showing perivascular and concentric arrangement of uniform plump oval-to-spindle-shaped myoid cells. These myoid cells are positive for small muscle actin but negative or focally positive for desmin. Matsuyama et al reported the clinicopathological features of 122 angioleiomyomas [5]. Of these cases, desmin was diffusely stained in 77 cases (63.1%) and partly positive in 24 (19.7%). In contrast, desmin was negative in 9 of 12 (75%) myopericytoma cases. In the present case, we excluded the diagnosis of myopericytoma due to the morphology and expression of desmin.

Solitary fibrous tumor shows uniform fibroblastic spindle cells, varying cellularity, and branching staghorn vessels that

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are patternless, all of which differ from the present case [17]. In addition, solitary fibrous tumors are typically positive for CD34; however, in the present case, CD34 was positive for only vascular endothelium; thus, we excluded the diagnosis of solitary fibrous tumor.

Beissert et al [2] reported the first adult PHA case in 2002; there have been several reports of prognosis in PHA. In the present patient, a pathological diagnosis of the liver mass was not available because he refused a pathological biopsy. As no pathological examination was performed, we selected to follow up on the liver mass. During the long 10-year follow-up period, PHA did not metastasize, and no new masses were seen in other organs or tissues. These findings suggest that PHA was a benign tumor in the present patient. In cases in which a pathological diagnosis cannot be confirmed and the possibility of malignancy is suggested, or the patient has obvious discomfort preoperatively, it may be reasonable for the surgeon to choose surgical treatment. In contrast, in cases in which the liver tumor is asymptomatic PHA, surgical treatment strategies can compromise patient benefit. Thus, in situations in which a pathological diagnosis confirms PHA, follow-up can be an option. Our case may be a supportive reference for clinical strategies for patient treatment.

Conclusions

We present a case of incidentally untreated PHA with long follow-up. As no metastasis was observed and the patient was alive, biological behavior of this tumor seemed to be benign. Clinically, for cases with suspected PHA, biopsy should be performed. It is necessary to distinguish PHA from other types of liver tumors, especially liver cancer. From combining our long-term observation and other case reports, we suggest that general follow-up is recommended if the preoperative pathological diagnosis can be confirmed and the patient has no other symptoms. In contrast, if the pathological diagnosis cannot be confirmed before surgical treatment and the possibility of malignant disease cannot be completely ruled out, or if there are obvious symptoms of discomfort, surgical treatment can be considered.

Acknowledgments

The authors thank Uranchimeg Bayarmagnai and Ariun-Uils Gankhuyag for their excellent technical assistance.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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