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Primary Retroperitoneal Cavernous Hemangioma With Extrahepatic Tissue: A Case Report and Literature Review

Christos Vrysis[1, 2], Marios Ponirakos[1], Konstantinos Koufatzidis[1], Athanasios Gkirgkinoudis[2], Aristotelis-Marios Koulakmanidis[2], Dimitrios Giovanitis[3], Konstantinos Papadimitropoulos[1]

[1]Second Department of Surgery, 251 Hellenic Air Force General Hospital, Athens, Greece, [2]First Department of Obstetrics and Gynecology, Alexandra Hospital, National and Kapodistrian University of Athens, Greece, [3]Department of Pathological Anatomy, 251 Hellenic Air Force General Hospital, Athens, Greece

Correspondence: *vryschri@hotmail.com***;** Tel.: + 30 697 8458371

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Abstract

Objective. We present a rare case of primary retroperitoneal cavernous hemangioma, highlighting its clinical, imaging, and histological parameters. **Case Report.** A 54-year-old patient presented with chronic abdominal pain that had been experienced for the past six months. No notable findings were identified in the patient's medical history, clinical examination, or laboratory tests. Full imaging was performed using magnetic resonance imaging and abdominal computed tomography (CT). A mass was found in the retroperitoneal area, located posterior to the stomach and close to the splenic portal, the left lobe of the liver, and the left hemidiaphragm. CT-guided fine-needle aspiration confirmed the presence of a benign tumor, which was surgically excised. Histological and immunohistochemical investigations confirmed the presence of a retroperitoneal cavernous hemangioma with extrahepatic tissue. **Conclusion.** Primary retroperitoneal cavernous hemangiomas are rare retroperitoneal tumors with nonspecific clinical and radiological characteristics, making diagnosis difficult. This case demonstrates the occurrence of extrahepatic tissue involvement, a feature that has been reported only exceptionally in the literature. Surgical resection is the primary treatment for symptomatic patients with a favorable prognosis, and histological examination of the surgical specimen confirms the diagnosis.

Key Words: Cavernous Hemangioma ▪ Retroperitoneum ▪ Primary Retroperitoneal Tumors ▪ Extrahepatic ▪ Case Report.

Introduction

Cavernous hemangiomas are tumors characterized by rapidly multiplying vascular endothelial cells and the ability to induce blood vessel formation (1). This benign tumor is commonly found in the orbital, mucosal, and hepatic regions (2). Rarely, cavernous hemangiomas are located in the retroperitoneal cavity (3).

The most prevalent sites of retroperitoneal hemangiomas include the pancreas, adrenal glands, and kidneys (4). An uncommon manifestation of this condition is a primary retroperitoneal cavernous hemangioma (PRCH), which is distinguished by its clear separation from the surrounding organs. Only five case reports have been published in the literature (3-7). The present study is the sixth report on the occurrence of this condition. Given the extremely limited number of reported cases, this study provides additional clinical and histological evidence, further clarifying the presentation and management of PCRH.

The diagnostic techniques of ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) are crucial for the detection and accurate localization of retroperitoneal cavernous hemangiomas (6, 7). Undoubtedly, the preoperative identification of this clinical entity remains challenging. In our case, the primary diagnosis was angiomyolipoma, and the presence of PRCH was revealed by histological examination after surgical excision.

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This study aimed to present an unusual case of primary retroperitoneal cavernous hemangioma, emphasizing its clinical, imaging, and histological features. This case also highlights extrahepatic tissue involvement, which is rarely reported in the literature.

Case Presentation

A 54-year-old patient visited the outpatient clinic of the 251 Hellenic Air Force General Hospital (Athens, Greece) due to chronic abdominal pain experienced over the past six months. The patient's medical history, clinical examination, and laboratory evaluations were unremarkable. There were no signs of other disorders, such as fever, chills, jaundice, nausea, vomiting, melena, or hematuria. Analysis of laboratory parameters, such as serum amylase, creatinine, alanine and aspartate aminotransferases, bilirubin, and urea nitrogen levels, yielded normal findings. The tumor markers were within the normal range. Imaging studies were performed. An abdominal CT scan revealed a well-circumscribed mass with no significant contrast enhancement during the arterial and portal venous phases (Figure 1). Based on these imaging results, there was no indication of the mass invading adjacent organs, the presence of feeding arteries, or retroperitoneal lymphadenopathy.

Considering the limited spread of the tumor and the absence of invasion or metastasis to other organs, the suspected diagnosis was a benign tumor, such as a lymphangioma cyst, gastrointestinal stromal tumor, or primary retroperitoneal benign tumor. CT-guided fine-needle aspiration was performed, and angiomyolipoma was suspected. However, the definite nature of the lesion could not be established preoperatively. Laparotomy was performed because of the size of the lesion and to alleviate the patient's discomfort.

Beyond the surgical issue, no concurrent diseases were discovered during routine preoperative examination. The patient received both general and epidural anesthesia throughout the surgical procedure. A left Kocher incision was performed. A thorough examination of the peritoneum and abdominal organs revealed no metastatic lesions. The gastrocolic ligament was then opened, the short gastric vessels were tied off, and access to the smaller sac was gained. The mass was identified

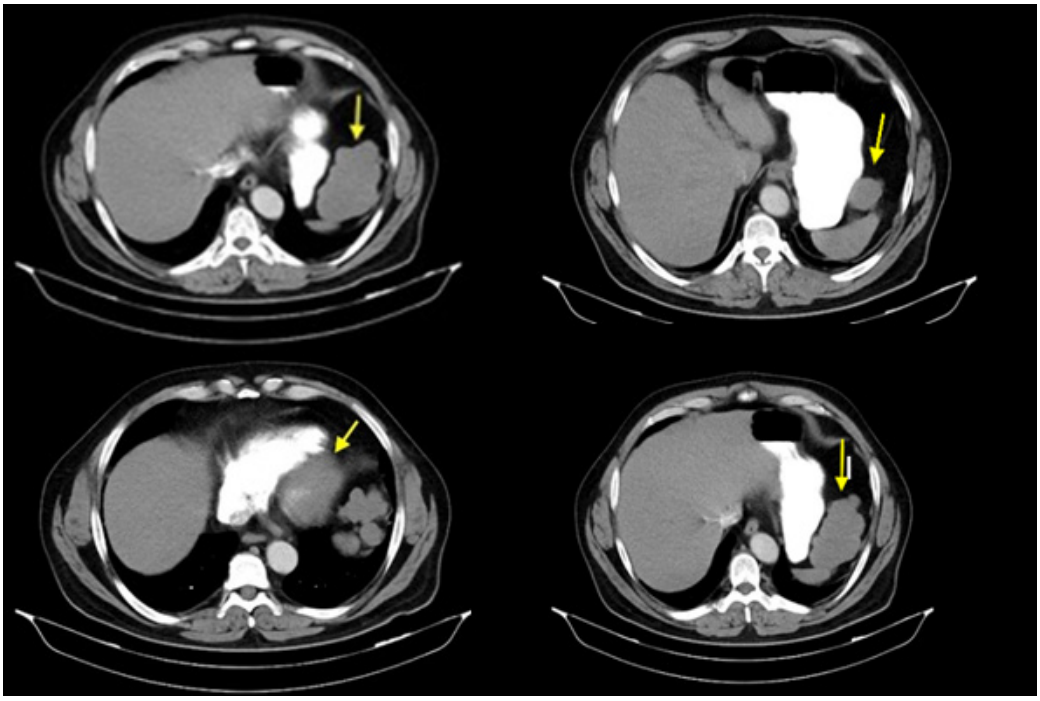

Figure 1. Computed tomography of the abdomen with oral and intravenous contrast.

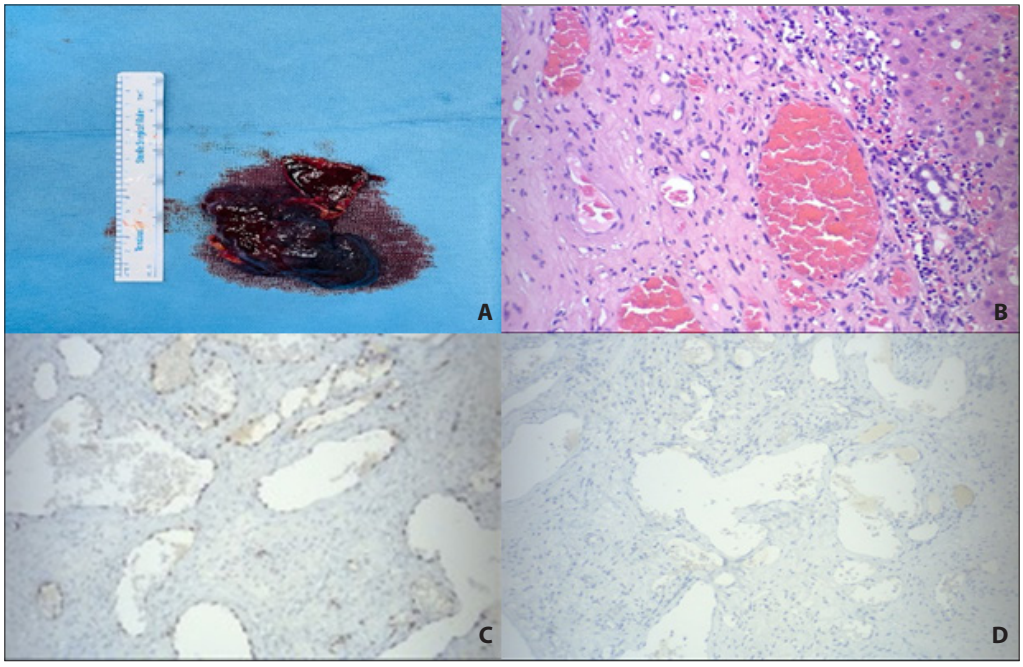

Figure 2. Macroscopic and microscopic examinations. (A) Surgical specimen; (B) Dilated and congested blood vessels with adjacent liver parenchyma (H&E, 10 ×); (C) ERG nuclear positivity (10 ×); (D) Absence of HMB-45 expression (10 ×).

in the posterior region of the stomach, adjacent to the splenic portal, and in direct contact with the left hemidiaphragm and left lobe of the liver. The tumor's blood supply was determined to arise from the retroperitoneal tissue rather than from the abdominal artery or other organs. The operation did not reveal any signs of invasion into the inferior vena cava, stomach, liver, renal capsule, pancreas, spleen, or any other nearby organs. The process involved surgically removing the lesion and establishing thorough control of bleeding. The tumor measured approximately 10 cm × 5 cm × 3 cm and had a propensity to bleed upon contact. At the macroscopic level, the tumor was characterized by large cavities filled with blood clots, surrounded by numerous small fluid-filled spaces (Figure 2A).

Histopathological examination of the resected specimen confirmed the diagnosis of a retroperitoneal cavernous hemangioma with extrahepatic tissue. Histological examination revealed numerous vascular spaces of different sizes, lined by a single layer of flattened cells in close proximity to the liver parenchyma. The tissues also showed blood vessels with varying lumen sizes and uneven wall thicknesses. Furthermore, there was evidence of coagulation in certain blood arteries, which is consistent with the features of a cavernous hemangioma. Immunohistochemical analysis revealed positive expression of CD34, CD31, SMA, and ERG, indicating that the cells were derived from endothelial tissue. As the HMB-45 and MelanA tests yielded negative results, the preoperative diagnosis of angiomyolipoma was excluded. No evidence of malignancy was detected (Figure 2B-D). The patient was discharged on postoperative day 7. The patient did not receive any adjuvant therapy. At the 10-month follow-up, the patient had not developed recurrence, their condition remained satisfactory, and they reported excellent quality of life.

Discussion

Retroperitoneal tumors are highly uncommon, comprising less than 0.2% of all tumor types (8). Liposarcoma and leiomyosarcoma are the most common malignant tumors in the

retroperitoneum, whereas teratomas, cysts, and neuromas are the most common benign tumors (9, 10). A cavernous hemangioma is a benign proliferation composed of blood vessels, commonly located on the skin or mucosal surfaces. The liver, spleen, kidneys, adrenal glands, and pancreas give rise to more visceral cavernous hemangiomas (CH). Occasionally, hemangiomas have been associated with the Kasabach-Merritt phenomenon, a congenital genetic abnormality that occurs during childhood (11).

While case reports of adult CHs in retroperitoneal organs are scarce, adult PRCH is even rarer, with only five recorded occurrences in the published literature (3-7). In line with these observations, our case adds further evidence by documenting an additional occurrence of PRCH and describing its distinct clinical and histological characteristics. The clinical data from these studies are presented in Table 1.

Early-stage cavernous hemangiomas often lack distinct clinical manifestations owing to their limited spatial distribution. Nonspecific symptoms and indications, such as stomach pain and anemia, manifest only when there is compression or invasion of the adjacent tissues involved (12). In terms of complications, there is evidence of hemorrhagic shock occurring from CH, highlighting the substantial risk associated with this tumor in the case of rupture or hemorrhage (13).

Table 1. Literature Review of Primary Retroperitoneal Cavernous Hemangiomas

*Publication	Age†/Sex	Symptoms	Preoperative imaging	Preoperative diagnosis	Surgery method	Outcome
Matsui et al., 2024 (6)	73 / M‡	Chronic abdominal pain and distension	CT¶ scan: a 35 cm mass adjacent to the left kidney, no contrast enhancement, no indication of lymphadenopathy or vascular supply from nearby organs MRI**: a heterogeneous signal intensity mass	Retroperitoneal chronic expanding hematoma	Open surgery	No recurrence of symptoms during the 6-month follow-up
AlBishi et al., 2023 (4)	43 / M‡	No	CT¶ scan: a 7.9 × 7.3 × 7.2 cm mass abutting the kidney, heterogeneous contrast enhancement implying cystic and solid components	Sarcoma	Laparoscopic surgery	NA††
Debaibi et al., 2022 (7)	35 / M‡	Chronic abdominal pain	CT¶ scan: a 4.5 × 2.7 × 2.2 cm cyst-like mass near the inferior vena cava and the third part of the duodenum, no contrast enhancement, no evidence of vascular supply from surrounding organs or lymphadenopathy	Benign tumor	Open surgery	No recurrence of symptoms during the 12-month follow-up
Fathi et al., 2018 (5)	58 / F§	Right hypochondriac discomfort for one month	US‖: a 12.5 × 14.5 × 16.5 cm mass with central cystic areas. CT¶ scan: a 16 × 14.4 × 18.2 cm well-defined heterogeneous mass at the right suprarenal area with irregular nodular enhancement	Hemangioma of the right lobe of the liver	Open surgery	NA††
He et al., 2012 (3)	38 / M‡	Epigastralgia	US‖: a cystic mass in the right upper quadrant, accompanied by right hydronephrosis and right upper ureter ectasia CT scan: an 8.7 cm cyst-like mass, hypodense, mild enhancement of the thick wall, no evidence of vascular supply from the nearby organs or of lymphadenopathy	Benign isolated retroperitoneal lesion	Open surgery	No recurrence of symptoms during the 3-month follow-up

*Authors and year of publication; †Year; ‡Male; §Female; ‖US=Ultrasound; ¶CT=Computed tomography; **MRI=Magnetic resonance imaging; ††NA=Non-available.

In addition to clinical presentation, radiological findings were also critical for diagnosis. The imaging modalities for CH are diverse, partly due to the limited presence of distinctive radiographic features, which makes it particularly susceptible to misdiagnosis. Potential causes of atypical findings include the presence of new blood vessels (neovascularity), blood clot formation (thrombosis), and bleeding (hemorrhage) (14). A CT scan precisely determined the placement of the tumor and distinguished variations in density. Typically, cavernous hemangiomas appear as low-density lesions with no significant contrast enhancement throughout the arterial or portal phase (6, 7, 15, 16). Magnetic resonance imaging may have been valuable in elucidating the internal architecture of the tumor (17, 18). On T1-weighted imaging, hemorrhage often appears as a dark signal, whereas on T2-weighted imaging, it may appear as a bright signal (13, 15).

The definitive diagnosis of cavernous hemangiomas primarily relied on histological examination. Microscopic examination revealed that the CH was composed of enlarged, aberrant blood vessels lined with a single layer of cells. These blood vessels are surrounded by fibrous tissues that are unevenly distributed, resulting in a sponge-like structure (19). Additionally, immunohistochemical staining commonly reveals a substantial positive response for CD31, CD34, and EGR, while the Ki-67 index displays a low value (20-22). A significant finding of our study is that examination of the surgical sample confirmed the presence of hepatic parenchyma despite not being attached to the liver during its surgical removal. To the best of our knowledge, this is the first documented case of its kind.

No significant complications were observed following surgery in our case or in previously reported cases of large retroperitoneal CHs (3, 6, 7, 18). Based on the consensus in the published literature, surgical intervention appears to be the optimal choice for two distinct reasons. The primary objective is to obtain histological confirmation of the mass, thereby ruling out any potential malignancy. The second purpose is to attain therapeutic goals while reducing the potential risk of complications, such as bleeding, infiltration, and compression (13). The predominant method of operation in the listed cases was open surgery (3, 5-7). Laparoscopic surgery was performed in only one case of PRCH, indicating that minimally invasive techniques may be considered depending on the characteristics and location of the tumor (4).

Conclusion

Retroperitoneal cavernous hemangioma is a rare type of tumor. Its vague clinical and imaging characteristics make diagnosis challenging. In our case, it was identified as primary, as it was not connected to any specific feeding artery and was detached from the surrounding organs. Both a multidisciplinary approach and an early acquisition of tissue biopsy are required. The primary approach for treating symptomatic patients is surgical resection, which often results in a favorable prognosis. Surgical resection remains the primary treatment for symptomatic patients and often results in a favorable prognosis. This case highlights the clinical relevance and uniqueness of identifying extrahepatic liver tissue within the tumor, a finding that has not been previously reported.

What Is Already Known on This Topic:

Retroperitoneal tumors are quite rare. The most commonly observed malignant tumors in the retroperitoneum are liposarcoma and leiomyosarcoma. On the other hand, cavernous hemangiomas are benign tumors composed of a proliferation of blood vessels. Adult primary retroperitoneal cavernous hemangiomas are even rarer, with only five reported cases in the literature. Early-stage cavernous hemangiomas often do not exhibit distinct clinical symptoms. Various imaging modalities can be used to identify them. On a CT scan, a cavernous hemangioma typically appears as a low-density lesion with no significant contrast enhancement during either the arterial or portal phase. The definitive diagnosis of cavernous hemangiomas primarily depends on histological examination. Surgical intervention is generally considered the optimal treatment option for these tumors, with open surgery being the predominant method used in reported cases.

What This Study Adds:

This study provides detailed insights into the imaging, histological diagnosis, and management of these rare tumors. Preoperative imaging, which included both CT and MRI scans, offered a precise representation of the anatomical structures surrounding the tumor. Histological and immunohistochemical markers were used to identify and analyze this condition, providing valuable information for future research in this area. A significant finding of our study is that examination of the surgical sample revealed the presence of hepatic parenchyma, despite

it not being attached to the liver during surgical removal. To the best of our knowledge, this is the first documented case of such a finding. Our primary objective was to expand the limited body of research on retroperitoneal cavernous hemangiomas by highlighting the importance of being aware of and conducting thorough assessments of similar cases.

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Conflict of Interest: The authors declare that they have no conflict of interest.

Ethical Approval and Consent: Informed consent was obtained from the patient for the publication of this case report and accompanying images.

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