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Primary retroperitoneal cavernous hemangioma: A case report and review of the literature

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ABSTRACT

Retroperitoneal cavernous hemangioma, a rare vascular tumor, has only 30 PubMed cases. Preoperative diagnostic criteria are unclear and often present asymptomatically until complications such as rupture or compression arise.

We present a 73-year-old with chronic abdominal pain and a giant retroperitoneal tumor. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed an irregular space-occupying mass in the retroperitoneum, suggesting a retroperitoneal chronic expanding hematoma. Total surgical resection confirmed the diagnosis as retroperitoneal cavernous hemangioma.

1. Introduction

Cavernous hemangioma represents a benign vascular neoplasm characterized by abnormal angiogenesis, predominantly affecting the orbital region, liver, brain, and skin, albeit rarely involving the retroperitoneum.[1] Since 1990, only 30 cases of adult retroperitoneal hemangioma have been reported.[2] Typically asymptomatic in the early stages, patients may require hospitalization and surgery as the tumor progresses, presenting with symptoms such as pain, nausea, and vomiting. Diagnostic modalities including ultrasound, CT, and MRI play crucial roles in identifying and locating lesions. However, definitively diagnosing retroperitoneal cavernous hemangioma without surgery remains challenging. In our case, the patient was initially diagnosed with retroperitoneal chronic expanding hematoma but histological analysis after surgical resection confirmed retroperitoneal cavernous hemangioma. This study includes a comprehensive literature review focusing on recent advancements in diagnosis and treatment, accompanied by our case report.

2. Case presentation

A 73-year-old man, with a medical history including left hemiplegia secondary to cerebral hemorrhage, diabetes, hypertension, and dyslipidemia, presented with chronic abdominal discomfort and distension. There were no signs of other systemic symptoms such as fever, jaundice, vomiting, or hematuria. Laboratory tests revealed elevated levels of D-dimer (21.17 μg/ml) and FDP (51.34 μg/ml). Tumor markers were within normal limits. CT revealed a 35 cm tumor adjacent to the left kidney, with no thrombotic findings (Fig. 1). Both arterial and portal phases showed no enhancement of the tumor wall and a heterogeneous concentration of contents within the tumor. No evidence of vascular supply or lymphadenopathy from surrounding organs was observed, and no obstruction of the left urinary tract was noted. MRI indicated a heterogeneous signal intensity of the tumor component on T2-weighted images, with a high signal intensity cystic lumen (Fig. 2). A diagnosis of retroperitoneal chronic expanding hematoma was considered. Despite the absence of invasion into surrounding organs, total tumor resection was performed due to the possibility of malignancy. Postoperatively, the patient recovered well and was discharged after 14 days. Pathological examination revealed benign vascular proliferation consisting of microvascular and capillary structures (Fig. 3A). Immunohistochemical analysis demonstrated diffuse positivity for CD34 on the endothelium of these vascular channels (Fig. 3B), affirming their endothelial origin and confirming the diagnosis of cavernous hemangioma. There was no evidence of malignancy. The patient showed no recurrence of pain or symptoms at the 6-month follow-up.

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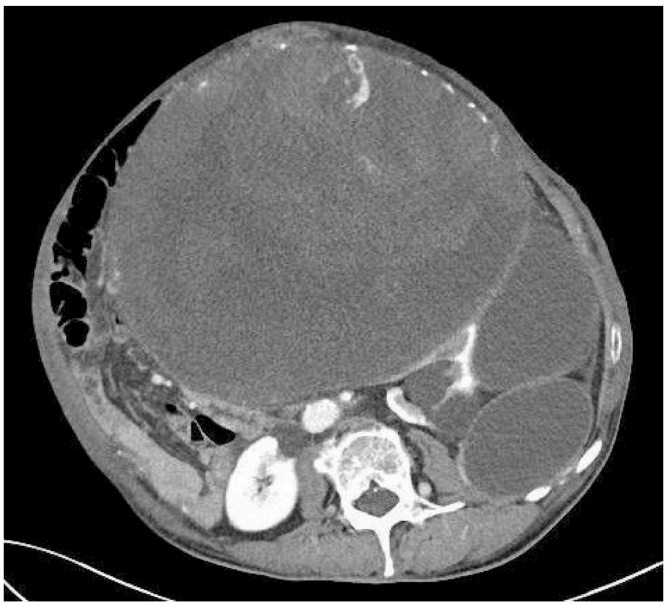

Fig. 1. CT scan showing a 35 cm tumor adjacent to the left kidney.

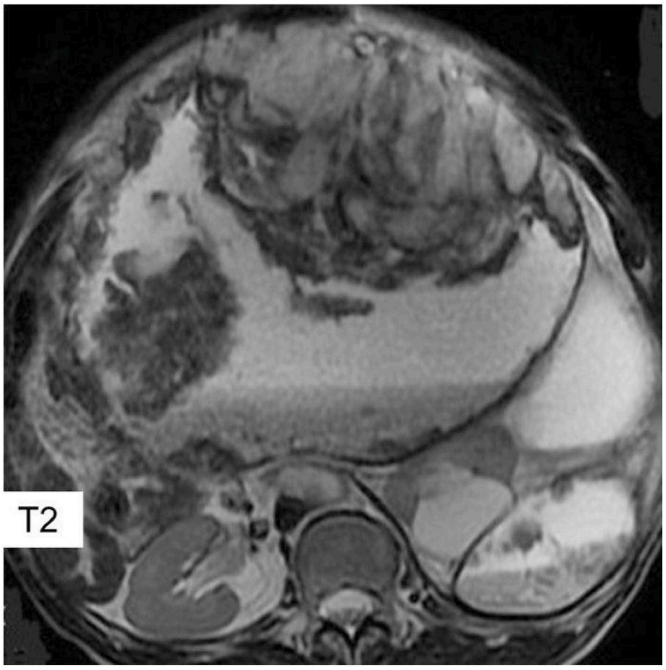

Fig. 2. T2-weighted image of an MRI showing a heterogeneous signal intensity of the tumor component with a high signal intensity cystic lumen.

3. Discussion

We presented a case of retroperitoneal cavernous hemangioma. Consistent with findings from previous studies, achieving a preoperative diagnosis was challenging, with pathology ultimately confirming the definitive diagnosis. Retroperitoneal tumors are exceptionally rare, comprising less than 0.2% of all tumor types.[1] Liposarcomas and leiomyosarcomas dominate as the primary malignant retroperitoneal tumors, while teratomas, cysts, and neurinomas are prevalent among benign masses in the same anatomical region.[1] Cavernous hemangioma is extremely rare. In the literature, 30 cases have been reported since 1990.[2] Based on our literature review, there are 8 cases of retroperitoneal cavernous hemangiomas, including our case, 9 cases originating

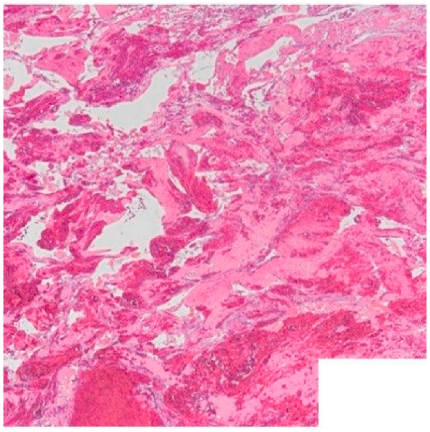

Fig. 3A. (A) Vascular proliferation consisting of venular and capillary structures (X 100).

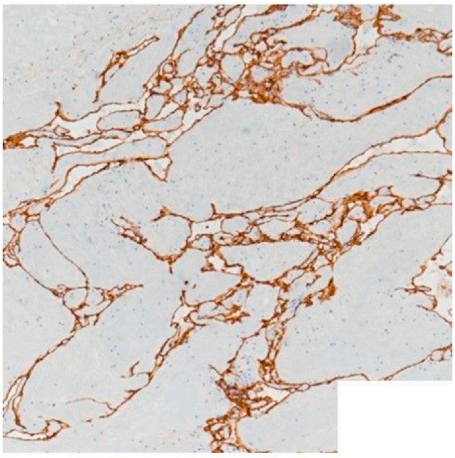

Fig. 3B. (B) CD34 immunostaining positivity observed on the endothelium of the vascular channels (X 100).

from the adrenal gland, 11 cases originating from the pancreas, 2 cases with unspecified origin, and 1 case originating from the psoas major muscle (Table 1).[2] Cavernous hemangiomas usually remain asymptomatic in the early stages due to the limited size of the occupied lesions. However, the compression or invasion of nearby anatomic structures can cause nonspecific symptoms such as vague abdominal discomfort, hematuria, renal vein thrombosis, hydronephrosis, and dilation of the upper ureter.[3] The occurrence of hemorrhage leading to hemorrhagic shock has been documented as a complication of cavernous hemangiomas, underscoring the significant risk associated with tumor rupture or bleeding.[4] The imaging findings of cavernous hemangiomas also exhibit diversity. CT and MRI are commonly employed in the preoperative assessment of cavernous hemangiomas. Hemangiomas typically demonstrate significant enhancement during the arterial phase on CT scans. On T1-weighted MRI imaging, cavernous hemangiomas typically exhibit low signal intensity, whereas on T2-weighted imaging, hemorrhage is often characterized by high intensity.[1] However, variations in the proportions of cystic and solid components within the tumor can result in varying degrees of enhancement during the arterial phase, posing challenges to diagnosis solely through imaging.[5] While CT scans can identify the tumor and discern density alterations, pinpointing the origin and achieving a definitive qualitative diagnosis is often elusive. Diagnosing retroperitoneal cavernous hemangiomas preoperatively presents considerable difficulty. In cases where retroperitoneal hemangioma remains asymptomatic, additional treatment or surgical

Table 1

No	Gender	Age	Preoperative diagnosis	Origin	Publication year
1	Male	20		Psoas major muscle	1990
2	Female	71	No	Pancreatic	1996
3	Male	63	No	Pancreatic	1998
4	Male	36	Retroperitoneal sarcoma	Retroperitoneal	1999
5	Female	58	Cystadenoma of the pancreas	Pancreatic	2002
6	Female	16	Hemorrhoids, Colonic vasodilatation	Unspecified	2005
7	Female	51	Neurogenic tumor	Unspecified	2006
8	Male	57	Cortical cancer	Adrenal	2008
9	Male	66	Cavernous hemangioma	Adrenal	2008
10	Male	79	No	Pancreatic	2009
11	Female	71	No	Retroperitoneal	2009
12	Male	55	Cystadenoma	Pancreatic	2012
13	Male	35	Lymphangioma cysts, GISTs, Primitive retroperitoneal benign tumors	Retroperitoneal	2012
14	Female	47	Malignant tumor	Retroperitoneal	2013
15	Male	14	Tumor with hemorrhage	Adrenal	2014
16	Female	57	Gastrointestinal mesenchymal tumors (GISTs), Carcinoid tumors, Neurogenic tumors, Metastatic lymphadenopathy, Other rare tumors	Adrenal	2016
17	Male	67	Adrenal tumor, Pheochromo cytoma	Adrenal	2017
18	Female	70	Benign adrenal tumor	Adrenal	2018
19	Male	49	Pancreatic cancer	Pancreatic	2018
20	Female	60	Cystic pancreas adenoma, Cystic pancreas adenocarcinoma	Pancreatic	2019
21	Female	41	Cystic dominant pancreas cystadenoma, Islet cell pancreas carcinoma	Pancreatic	2019
22	Female	30	Pancreas benign tumor	Pancreatic	2019
23	Female	37	No	Pancreatic	2020
24	Female	63	Pancreatic cancer	Pancreatic	2020
25	Male	38	No	Retroperitoneal	2020
26	Male	35	Adrenal pheochromocytoma	Adrenal	2021
27	Male	57	Left adrenocortical carcinoma, Pheochromocytoma, Retroperitoneal mesenchymal-derived tumor	Adrenal	2021
28	Male	70	No	Adrenal	2021
29	Female	54	Malignant tumor	Retroperitoneal	2022
30	Female	43	Retroperitoneal lymphatic duct cyst	Retroperitoneal	2023
31	Male	73		Retroperitoneal	Our own case

intervention is unnecessary. However, surgical excision of the mass is advised if the tumor exhibits rapid growth, compresses neighboring organs, or manifests nonspecific symptoms.[1] Considering combined resection of the mass and adjacent organs appears justified to mitigate the risk of local recurrence. Pathological examination remains the definitive method for diagnosing retroperitoneal cavernous hemangiomas. From a pathological perspective, cavernous hemangiomas are predominantly characterized by an expanded, anomalous vascular lumen enveloped by a monolayer of vascular endothelial cells exhibiting CD34-positive staining by immunohistochemistry.[1] It is composed of a sponge-like structure. Depending on the size of the vascular lumen, it can be capillary or cavernous. Preoperative diagnosis of retroperitoneal cavernous hemangioma is difficult and requires a thorough understanding of imaging features in combination with relevant clinical and pathologic studies to exclude the possibility of malignancy. This is important to minimize surgical and postoperative complications.

Limitations include the fact that retroperitoneal cavernous hemangiomas are difficult to diagnose preoperatively, progress asymptomatically, and are difficult to suspect and consider because the general urologist rarely has the opportunity to see them. The final diagnosis cannot be made without pathology.

4. Conclusion

Retroperitoneal cavernous hemangioma presents a diagnostic challenge preoperatively due to its rarity and nonspecific clinical or radiological features.

Surgical resection remains the primary and often the only curative treatment option. This approach not only provides histopathological confirmation for diagnosis and rules out malignancy but also effectively prevents complications associated with tumor invasion and compression of adjacent structures. Given the difficulty in clinical or radiological diagnosis, histopathological examination remains crucial for confirmation. Sharing these case reports and conducting a comprehensive review would serve to enlighten the public about the challenging nature of the diagnosis, emphasizing the necessity for thorough examination and pathology confirmation.

Consent

Written informed consent was obtained from the participant for the publication of this case report. A copy of the written consent is available for the editorial review.

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Statement of conformation to the declaration of Helsinki

This study was conducted according to the principles mentioned in the Declaration of Helsinki.

CRediT authorship contribution statement

Yuki Matsui: Conceptualization, Formal analysis, Writing – original draft, Writing – review & editing. **Sho Okada:** Data curation. **Yoshihiro Nakagami:** Supervision. **Takashi Fukagai:** Project administration, Validation. **Kazuhiro Matsuda:** Supervision. **Takeshi Aoki:** Supervision.

Declaration of competing interest

The authors declare no conflicts of interest associated with this manuscript.

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