Case Report

Retroperitoneal capillary arteriovenous malformation mimicking a malignant neoplasm

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Abbreviations & Acronyms
CT = computed tomography
MRI = magnetic resonance imaging
ISSVA = International Society for the Study of Vascular Anomaly

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Introduction: Retroperitoneal tumors account for 0.2% of all neoplasms. Among these tumors, retroperitoneal vascular malformations are particularly rare, with most previously reported cases being venous malformations.

Case presentation: A 72-year-old woman was diagnosed with a retroperitoneal tumor on abdominal computed tomography. The 27-mm diameter tumor was located away from the right kidney and major vessels in the right perirenal adipose tissue. Contrast-enhanced computed tomography revealed a heterogeneously enhanced tumor with well-defined borders. Dynamic contrast-enhanced magnetic resonance imaging revealed rapid enhancement in the arterial phase and a progressive filling-in pattern in the delayed phase. Although vascular malformation was suspected, a definitive diagnosis could not be established. The retroperitoneal tumor was excised laparoscopically for therapeutic and diagnostic purposes, and the histopathological diagnosis confirmed it as a capillary arteriovenous malformation.

Conclusion: Herein, we presented a rare case of retroperitoneal capillary arteriovenous malformation that was difficult to definitively diagnose preoperatively.

Key words: hemangioma, liposarcoma, retroperitoneal tumor, vascular malformation.

Keynote message

We presented a rare case of a retroperitoneal combined capillary arteriovenous malformation. The heterogeneously enhanced mass was located away from nearby organs and major vessels in the retroperitoneum, mimicking a neoplasm. The present case exemplifies a radiologically indeterminate retroperitoneal mass, possibly a malignant neoplasm or a vascular malformation.

Introduction

Retroperitoneal tumors represent 0.2% of all neoplasms.[1] Liposarcoma and leiomyosarcoma are the most common histology, while vascular lesions in the retroperitoneal space are rare, accounting for only 1–5% of all cases.[2,3] Among these vascular lesions, venous malformations, including the so-called cavernous hemangioma, are the most common. Herein, we reported a rare case of a retroperitoneal capillary arteriovenous malformation. Making an accurate preoperative diagnosis on imaging studies was difficult, and the lesion was excised for therapeutic and diagnostic purposes.

Case report

A 72-year-old woman was referred to our department with a retroperitoneal tumor. She had persistent hypertension, and abdominal CT showed a 27-mm diameter tumor in the right perirenal adipose tissue. The patient had no symptoms or medical history and denied a history of blunt abdominal trauma.

The tumor was caudal to the right kidney (Fig. 1). Contrast-enhanced CT revealed a heterogeneously enhanced tumor with well-defined borders. The venous flow drained into the right gonadal vein. The right perirenal adipose tissue density was slightly increased. No increase in the volume of the surrounding adipose tissue was observed. MRI showed low-signal intensity compared to the cortex of the kidneys on T1-weighed imaging and an inhomogeneous high signal on T2-weighed imaging (Fig. 1). Dynamic contrast-enhanced MRI revealed rapid cribriform enhancement in the posteromedial peripheral area of the tumor during the arterial phase (Fig. 2). In the delayed phase, the mass showed a progressive filling-in pattern and was strongly enhanced with spotty low-signal areas (Fig. 2). No fatty components were visible in the tumor. Neither diffusion restriction nor dilated feeding arteries were observed.

The enhancement patterns on dynamic contrast-enhanced MRI reflected its pathological complexity: arterial-rich component, slowly enhanced venous or stromal component, and non-enhanced lesions such as necrotic, thrombotic, or highly

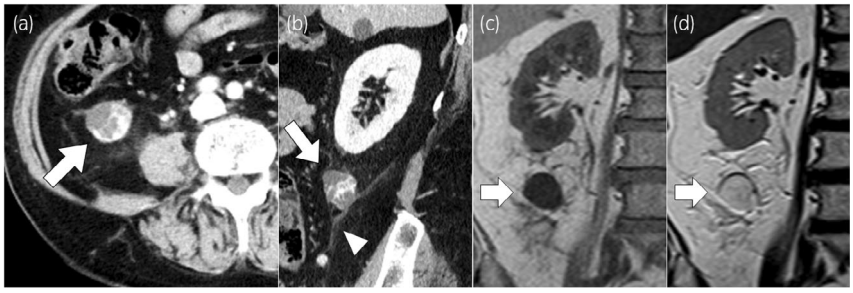

Fig. 1 Contrast-enhanced abdominal CT images of the tumor in the (a) axial and (b) sagittal planes, and (c, d) MRI in the coronal plane. (a, b) The lobulated mass (27 mm in diameter, arrow) is in the right perirenal adipose tissue apart from the right kidney and major vessels. It shows heterogeneous and septal enhancement. The increase of the right perirenal adipose tissue density and thickening of the retroperitoneum (triangle) are shown. (c) The mass (arrow) shows a low signal compared with the cortex of the kidneys on a T1-weighed image and (d) an inhomogeneous high signal on a T2-weighed image.

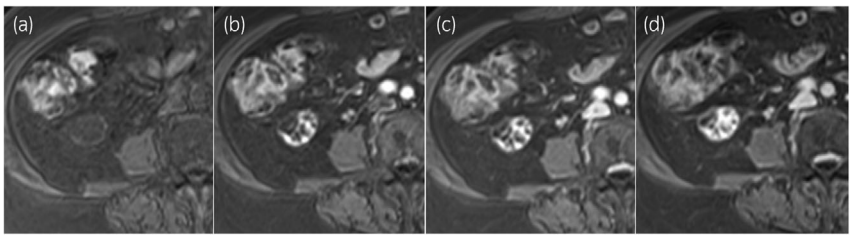

Fig. 2 Dynamic contrast-enhanced MRI (a: pre-, b: 30 s, c: 70 s, and d: 300 s after gadolinium injection) revealed rapid cribriform enhancement at the posteromedial peripheral area of the tumor in the arterial phase (b). A progressive filling-in pattern is shown in the delayed phases (c, d).

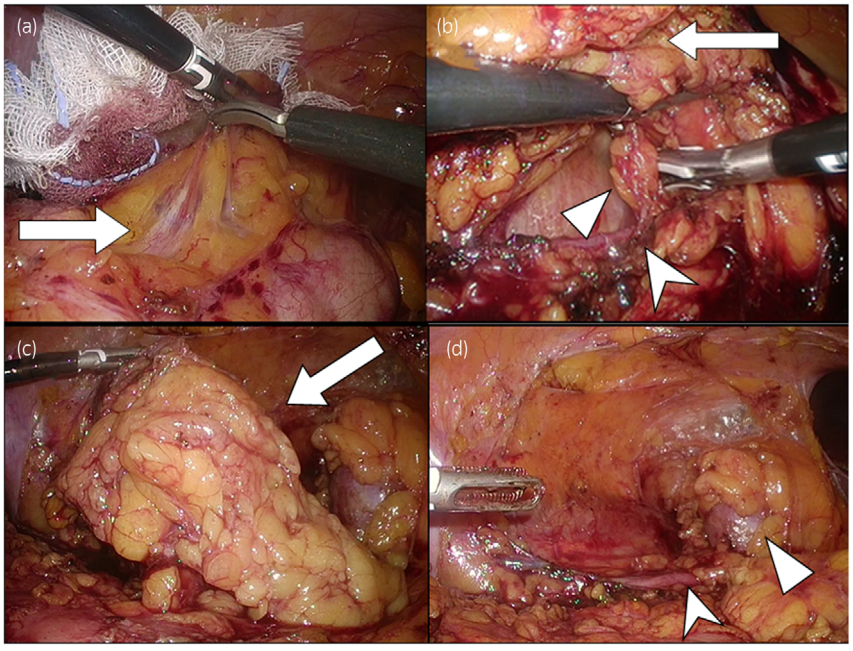

Fig. 3 Surgical images. (a) A whitish tumor (arrow) is visible beneath Gerota's fascia. (b) Blood vessels (triangle) between the tumor (arrow) and the right gonadal vessels (arrowhead) were treated with LigaSure™. (c) The tumor was excised together with the surrounding adipose tissue (arrow). (d) Retroperitoneal space after tumor resection. The right kidney (triangle) and gonadal vessels (arrowhead) were preserved.

myxoid components. A retroperitoneal vascular malformation was suspected; however, making a definitive diagnosis was difficult. The differential diagnoses included solitary fibrous tumor, paraganglioma, schwannoma with degenerative changes, intravascular epithelioid hemangioendothelioma, and sarcomas including dedifferentiated liposarcoma and undifferentiated pleomorphic sarcoma. A paraganglioma or extra-adrenal pheochromocytoma was considered less likely because of low urinary metanephrine and normetanephrine levels and negative uptake on ^{123}I-metaiodobenzylguanidine scintigraphy. A percutaneous biopsy was not performed due to the fear of possible bleeding. Surgical resection of the retroperitoneal tumor was planned for therapeutic and diagnostic purposes.

The patient underwent laparoscopic surgery via the transperitoneal approach using three ports and standard laparoscopic devices under general anesthesia (Fig. 3). The right colon was reflected medially along the white line of Toldt to visualize the anterior surface of Gerota's fascia. The tumor was easily detected beneath the fascia. Blood vessels were identified between the tumor and the gonadal vessels; however, the feeding arteries could not be separately identified. They were coagulated and cut using a LigaSure™. The tumor was excised with the surrounding adipose tissue, and the right kidney and gonadal vessels were spared.

The tumor was a white solid 30 × 10 mm mass within the adipose tissue. Histopathologically, the tumor had an incomplete fibrous capsule with a large hyalinized area at the center (Fig. 4). Blood vessels of various sizes surrounded the hyalinized areas. The most predominant were capillaries, which proliferated densely and anastomosed with each other, adjacent to the inner surface of the fibrous capsule. Large venous vessels with thrombi and arteries were observed, some of which were encased in capillaries. The arteries were primarily located at the tumor's periphery. Cytological atypia or mitoses of vascular endothelial cells were not observed in the arterial, large venous, or capillary vessels. The excised adipose tissue was mature without atypical lipoblasts. Thus, the tumor was diagnosed as a retroperitoneal vascular malformation involving arterial, venous, and capillary vessels.

The patient's postoperative course was uneventful, and she was discharged at 5 days postoperatively. The patient is doing well without recurrence at 10 months postoperatively.

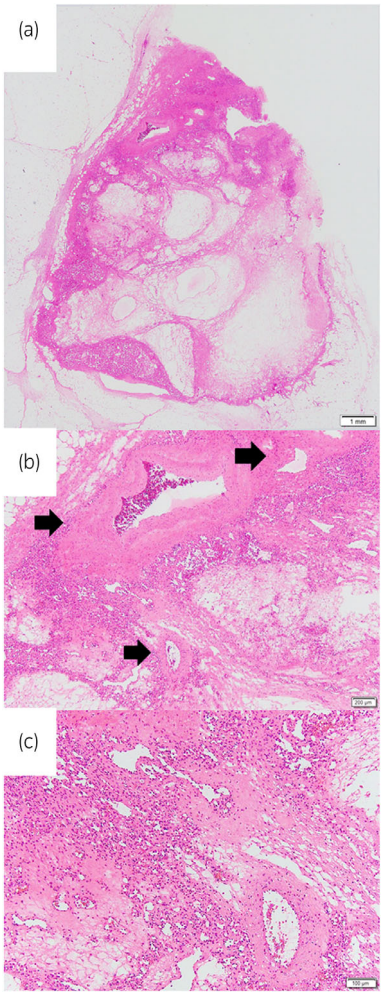

Fig. 4 Histopathology of the specimen (hematoxylin and eosin). (a) The lesion has an incomplete fibrous capsule. Blood vessels of variable sizes surround the large hyalinized area. (b) Arteries of variable sizes (arrows) are visible at the lesion's periphery. (c) Capillaries are densely proliferated and anastomosed with each other adjacent to the inner surface of the fibrous capsule.

Discussion

Vascular tumors and malformations in the retroperitoneal space are rare, with fewer than 30 cases reported in the English literature.[4–15] Among them, the most prevalent is venous malformation, including cavernous hemangioma. To our knowledge, this is the first report of an isolated retroperitoneal capillary arteriovenous malformation.

In previous reports,[4–15] retroperitoneal vascular malformations affected patients of all ages; approximately half were 20–40 years old. No obvious bias was observed in sex or laterality. The manifestations were nonspecific and included abdominal pain, back pain, dyspepsia, epigastralgia, and abdominal distension, although some were found incidentally. In most cases, a definitive diagnosis was not made on imaging studies and the lesions were surgically resected.

Based on advances in understanding the underlying pathogenetic mechanisms, the histopathological classification of vascular anomalies has changed recently.[16–18] In the updated classification system proposed by the ISSVA, vascular anomalies are classified as vascular tumors (neoplastic lesions) or vascular malformations (non-neoplastic lesions) based on the presence or absence of proliferation of vascular endothelial cells.[16–18] Vascular malformations lack proliferative changes in endothelial cells and primarily consist of structural vascular abnormalities. They are composed of capillaries, lymphatics, veins, arteries (simple vascular malformations), or a combination thereof (combined vascular malformations). The term "hemangioma" was previously used to describe benign masses of clusters of blood-filled cavities lined with endothelial cells. However, lesions which were previously described as cavernous hemangioma and venous hemangioma are

simple venous malformations according to the latest ISSVA classification, and the term hemangioma is now reserved only for definite vascular tumors such as infantile and congenital hemangioma.

Dynamic contrast-enhanced MRI aids the differential diagnosis of vascular anomalies.[19,20] Vascular malformations include slow- and fast-flow types. The slow-flow type includes capillary, venous, and lymphatic malformations, while the fast-flow type includes arteriovenous malformations and fistulae. In the present case, strong enhancement was observed in the arterial phase, reflecting the existence of an arterial component.

Making an accurate preoperative diagnosis of vascular malformation in imaging studies was difficult. The most common previously reported cases of retroperitoneal vascular malformations are venous malformations, which include cavernous and venous hemangiomas. On imaging studies, cavernous hemangiomas are often characterized by a lobulated septate mass without a clear capsule (cotton wool appearance).[4,10,11] They are typically attached or adjacent to surrounding organs or major vessels.[5,6,8–10,12–15] In contrast, in the present case, the lesion formed a capsule that was separated from other organs and major vessels, similar to a neoplasm. Surgical resection is possible for diagnostic and therapeutic purposes, although intervention for vascular malformations is indicated only for large lesions with a threat of incidental rupture and/or bleeding. Our case exemplifies a radiologically indeterminate case of a retroperitoneal mass that was a malignant neoplasm or vascular malformation.

Author contributions

Kazuki Yanagida: Conceptualization; data curation; investigation; writing – original draft. Tomoyuki Kaneko: Conceptualization; data curation; investigation; writing – review and editing. Koji Saito: Data curation; investigation; visualization; writing – review and editing. Masayoshi Yamamoto: Data curation; investigation; writing – review and editing. Asako Yamamoto: Data curation; investigation; visualization; writing – review and editing. Yukio Yamada: Investigation; writing – review and editing. Tohru Nakagawa: Data curation; investigation; project administration; validation; writing – review and editing.

Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an Institutional Reviewer Board

Not applicable.

Informed consent

Written informed consent was obtained from the patient for the publication of the details of this medical case and any accompanying images.

Registry and the Registration No. of the study/trial

Not applicable.

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