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Case Report

Retroperitoneal leiomyosarcoma mimicking an ovarian tumor diagnosed using a negative ovarian pedicle sign ☆,☆☆,★

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ABSTRACT

Retroperitoneal leiomyosarcoma (RPLMS) is rare and usually presents as a large abdominal mass with poor clinical symptoms. Radiological findings of an RPLMS arising in the pelvis of a woman resemble those of adnexal tumors. Herein, we present a case of RPLMS mimicking an adnexal tumor which was differentiated from having an ovarian origin as the right ovarian vein was passing through the tumor but there was no direct vascular connection with the tumor. Therefore, it is important to identify the ovarian vein to distinguish between these tumors.

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Introduction

Primary retroperitoneal tumors are extremely malignant tumors (70%–80%). Of these, leiomyosarcoma (LMS) is the second most frequent primary retroperitoneal malignancy (28%), followed by liposarcoma [1–3]. A retroperitoneal leiomyosarcoma (RPLMS) usually lacks obvious clinical symptoms and is often incidentally detected as a large abdominal mass. However, the imaging findings of an RPLMS arising from the ovarian fossa are often similar to those of adnexal tumors [4–6]. In this report, we describe a case of RPLMS mimicking an adnexal tumor which was diagnosed because the right ovarian vein was passing through the tumor, differentiating it from the latter.

Case report

A 63-year-old woman presented with abdominal pain in the right flank and lower abdomen for 1 year and right inguinal pain for 2 months. She underwent a transvaginal ultrasound examination as routine screening for uterine cervical cancer at a gynecologic clinic, which revealed a tumor (5 cm in diameter) in the right adnexal region. The patient was referred to our obstetrics and gynecology department for further evaluation and treatment.

Further investigation with contrast-enhanced computed tomography (CECT) revealed a solid tumor (5.7 × 3.5 × 3.5 cm) without calcification adjacent to the uterine fundus on the right side. The boundary was clear, and no bridging vessels were found between the uterus and the tumor (Fig. 1); no enlarged lymph nodes or distant metastases were observed. Magnetic resonance imaging (MRI) also showed a tumor with homogeneous low signal intensity on the T1 weighted image (WI) and slightly heterogeneous low signal intensity on the T2WI; no flow voids were observed between the uterus and the tumor. Furthermore, the axial diffusion weighted images showed slightly high intensity. After administration of contrast material, the tumor was heterogeneously enhanced with patchily unenhanced areas. Accordingly, the origin of the tumor was assumed to be from the right ovary based on its location and clear boundary between the tumor and the uterus, without bridging vessels (Fig. 2). All blood counts, liver and renal function tests, and tumor markers, including CA 19-9 and CA 125, and carcinoembryonic antigen, were within the normal range.

A laparotomy was performed to remove the tumor, which revealed a pelvic mass arising from the retroperitoneal cavity near the normal right ovary; intraoperative frozen section analysis suggested a leiomyosarcoma (LMS). Total hysterectomy and bilateral adnexectomy were performed, and the pelvic tumor was removed along with the right adnexa. There was no direct invasion of the tumor into the right ovary, ileum, or right external iliac vein.

Postoperative histological examination of the specimen confirmed the diagnosis of LMS, which was composed of spindle-shaped atypical cells proliferating in bundles at a mitotic rate of 20/10 high-powered field. On immunohistochemical staining, the sample showed positive results for smooth muscle actin (SMA) and desmin, and the Ki-67 labeling index was high (approximately 40% in the hot spots) (Fig. 3). The final pathological diagnosis was leiomyosarcoma arising from the retroperitoneum. Notably, there was no extension to the right ovary or fallopian tube, no pathological invasion of the right external iliac vein, and the surgical margins were negative.

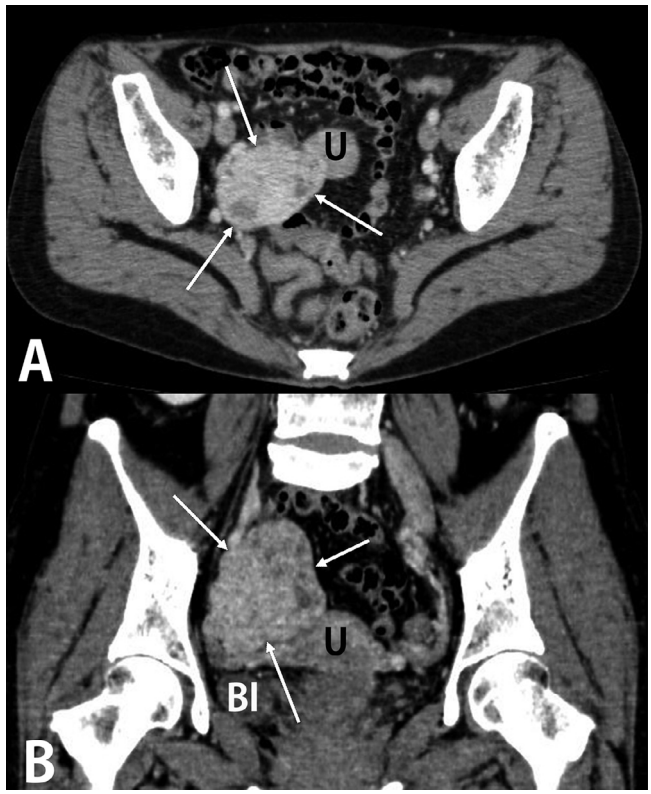

Fig. 1 – Contrast-enhanced (A) axial and (B) reformatted coronal abdominal computed tomography showed a solid tumor (arrow) with non-homogeneous enhancement adjacent to the uterine fundus (U). No bridging vessels were observed between the tumor and the uterus.

Therefore, the CECT was retrospectively reviewed with reference to the pathological findings, in which the right ovarian vein was identified as passing through the tumor, suggesting that the origin of the tumor was not the ovary (Fig. 4).

The patient recovered well postoperatively and was discharged seven days later. Positron emission tomography-computed tomography (PET-CT) performed one month after the operation showed no metastasis. However, the 3-month follow-up PET-CT showed hyperaccumulation of F-fluorodeoxyglucose in the liver and the right fourth and sixth ribs, suggesting metastasis. Additionally, multiple nodules in the right lung suggestive of metastasis were seen on chest CT. However, they were too small to demonstrate hyperaccumulation on PET/CT. Accordingly, chemotherapy was initiated with eight courses of doxorubicin as a single agent, followed by tribulin monotherapy to not exceed the recommended dose of doxorubicin, which was suspended when liver metastases were detected 2.5 years after the operation. Radiation therapy was started to relieve rib pain 3.5 years after surgery.

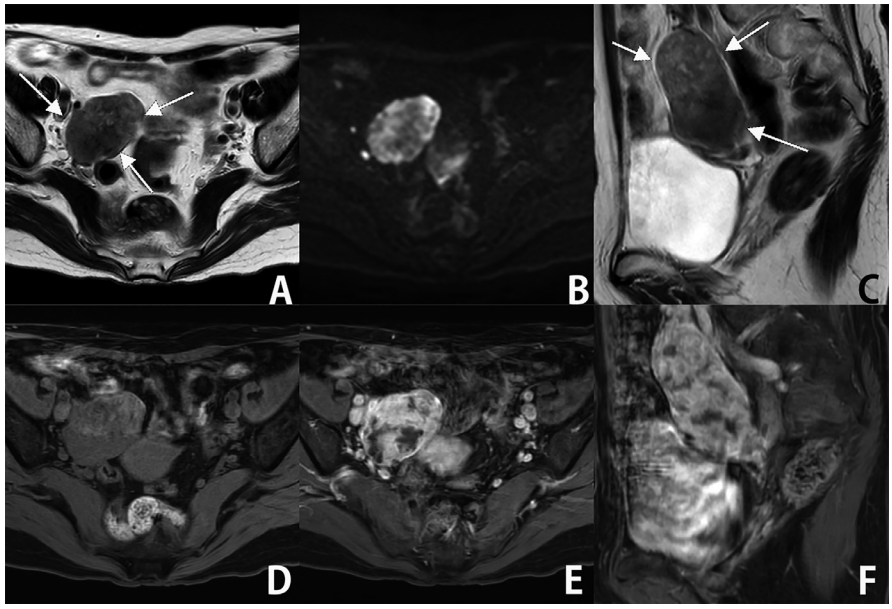

Fig. 2 – (A) Axial T2-weighted magnetic resonance imaging (MRI) showed a slightly hypointense mass located on the right side of the pelvis with a high signal on diffusion-weighted imaging (DWI) (B). (C) Sagittal T2-weighted MRI revealed an oval tumor with a clear boundary with the surrounding tissues. (D) Axial T1-weighted MRI with fat suppression demonstrating the tumor in hypointensity with artifacts on the ventral side originating from the peristalsis of the bowels. (E and F) MRI before and after administration of contrast material. On contrast-enhanced axial and sagittal MRI, the tumor was heterogeneously enhanced with patchy hypo-signal areas.

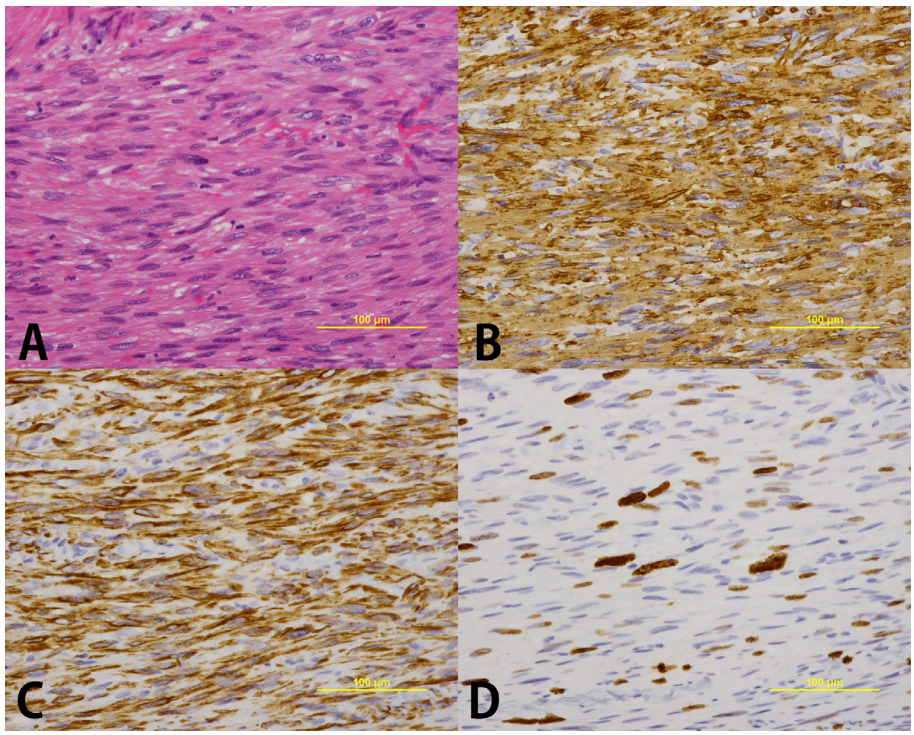

Fig. 3 – (A) Histological examination revealed proliferation of spindle cells with mitosis and nuclear atypia (hematoxylin and eosin stain, x200). Immunohistochemistry demonstrated strong and diffuse staining of the smooth muscle action stain (B) and desmin (C) (x200). (D) The Ki-67 labeling index was high at approximately 40% in the hot spots (x200).

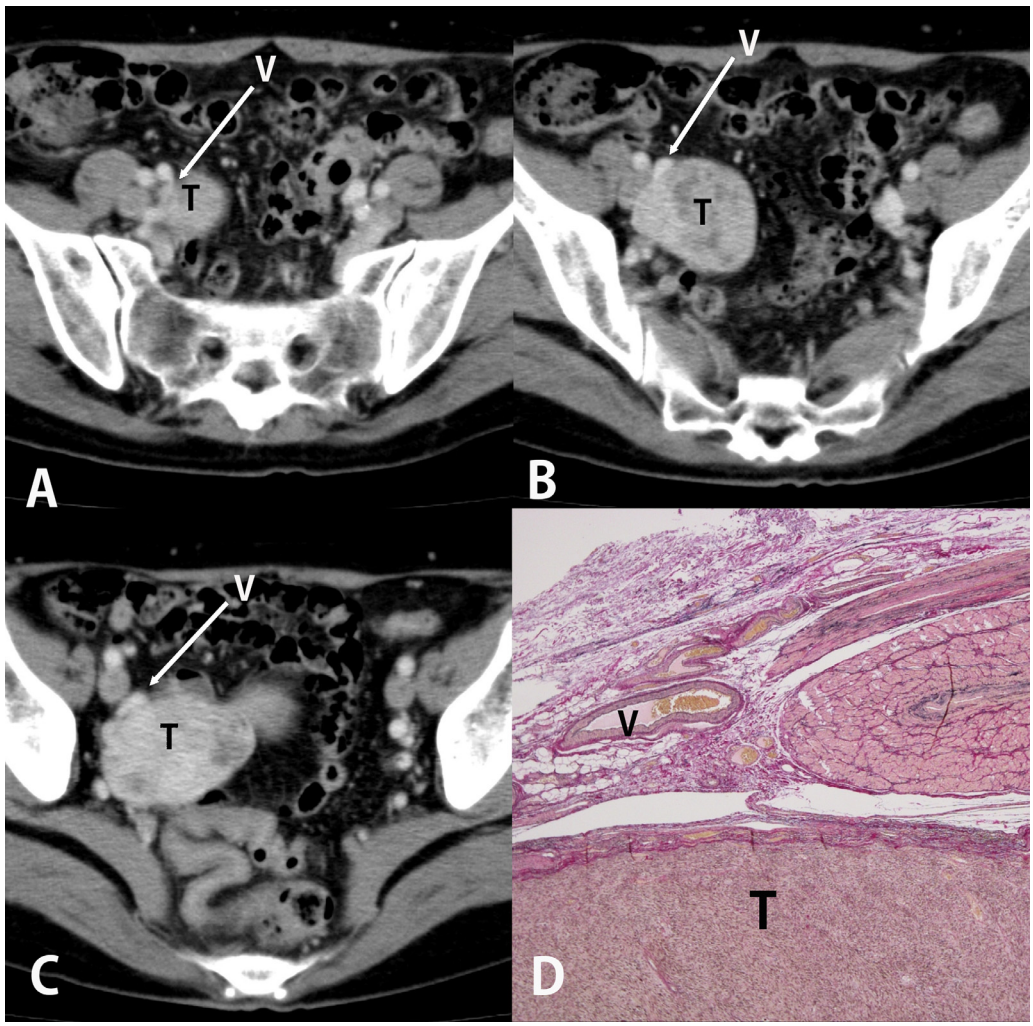

Fig. 4 – (A–C) Intermittent contrast-enhanced axial computed tomography images demonstrated the right ovarian vein (V, arrows) adjacent to the tumor (T), which does not have a connection to the tumor as a drainage vein (V). (D) Microscopic examination revealed no connection between the right ovarian vein (V) and the tumor (T).

Discussion

This case report describes a rare case of RPLMS arising from the ovarian fossa, which was originally mistaken as having an ovarian origin. LMS accounts for 5%-10% of all sarcomas, and 50% of these tumors arise in the abdomen, including the retroperitoneum [1,2,7]. Usually found as a large abdominal mass as the tumor enlarges without clinical symptoms, LMS may occasionally be accompanied by symptoms such as abdominal pain and distention [4]. Preoperative imaging plays an important role in estimating the organ of origin, the tumor component, tumor extent, absence or presence of enlarged lymph nodes, and distant metastases [8]. The first-line treatment for RPLMS is surgical resection with negative margins. Radiation therapy and chemotherapy may be administered preoperatively or postoperatively; however, their efficacy remains unclear.

Typically, LMS is seen as a large lobulated mass, often with internal necrosis, hemorrhage, and heterogeneous strong enhancement; calcification within LMS is rare. It commonly demonstrates hypo- to isointense signal intensity on T1WI and high signal intensity in solid components on T2WI [9,10]; however, if the tumor has hemorrhagic areas, it shows high signal intensity on T1WI. After administration of the contrast material, LMS may be heterogeneously enhanced based on the presence of necrotic areas [3]. In the present case, CT showed a well-defined, well-circumscribed, cylindrical mass measuring 5.7 cm along the longest axis, with heterogeneous enhancement, while MRI showed low signal intensity on T1WI and heterogeneous low-to-high signal on T2WI. The entire mass showed strong heterogeneous enhancement and relatively strong diffusion limitation, which was consistent with the imaging findings of an LMS.

Initially, the tumor was presumed to have originated from the ovary due to several reasons. First, it was relatively small to be a retroperitoneal sarcoma, which is reportedly measured as >10 cm in 70% of cases [11]. Second, the tumor was located adjacent to the uterine fundus, and the pedunculated uterine leiomyoma was ruled out because of the absence of bridging

vessels between the tumor and the uterine body [12]. Third, normal ovaries were not evident because the patient was postmenopausal although identification of normal ovaries is a key finding for denying ovarian origin [6]. Similar cases of RPLMS mimicking ovarian tumors have been demonstrated in several previous reports [4–6].

When retrospectively reviewed, the right ovarian vein in our patient did not connect to the tumor, which may point toward the diagnosis of a non-ovarian tumor. Previous studies have proposed an "ovarian vascular pedicle sign" which suggests that the origin of the tumor is ovarian if a direct connection can be established between the asymmetrically enlarged gonadal veins and the pelvic mass (with 92% sensitivity, 87% specificity, and 91% accuracy) [13,14]. In addition, ovarian veins have been reported as reliable land markers with a high delineation rate of 92% on CECT [15]. However, the negative predictive value of the ovarian vascular sign is reported to be 69%, which may be a limitation to its clinical application [13]. Although the previous study examined the imaging findings of uterine and ovarian tumors, this sign may be extrapolated to the differential diagnosis of ovarian tumors from non-ovarian masses arising in the ovarian fossa [16]. However, the diagnostic performance of this sign is not clear for differentiating between these tumors because of a lack of previous studies considering this discussion. The RPLMS is reported to be present in extraluminal space in cases of 62% [3], in which the ovarian veins pass by the tumor. In the current case, the absence of a direct connection between the ovarian vein and the tumor (negative ovarian vascular pedicle sign) might have been the reason for the correct diagnosis.

Conclusion

Although RPLMS is a rare tumor, it is still the second most frequent primary retroperitoneal malignancy. In this case, the adnexal tumor-mimicking RPLMS was distinguished from the adnexal tumor based on the absence of a direct vascular connection (right ovarian vein). Therefore, adequate contrast-enhanced imaging and visualization are essential to delineate the organ of origin of RPLMS.

Patient consent

Consent for publication has been obtained from the patient.

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