

Multidetector CT and MR Imaging Findings of Renal Vein Leiomyosarcoma with Both Extraluminal and Intraluminal Components: A Case Report¹

혈관 내외 구성요소를 모두 갖는 신정맥 기원 평활근육종의 다중 검출기 전산화단층촬영 및 자기공명영상 소견: 증례 보고¹

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Leiomyosarcomas of the large veins are uncommon tumors commonly originating from the inferior vena cava. A primary leiomyosarcoma arising from the renal vein is very uncommon. Herein, we describe the detailed multidetector computed tomography and magnetic resonance imaging findings of a renal vein leiomyosarcoma with both extraluminal and intraluminal components in a 55-year-old woman. The intraluminal component in the renal vein, a heterogeneous enhancement caused by combined necrosis and portions of low or intermediate signal on T2-weighted images are imaging findings that suggest the diagnosis of primary leiomyosarcoma of the renal vein.

Index terms

Leiomyosarcoma
Retroperitoneum
Renal Vein
Multidetector CT
Magnetic Resonance

INTRODUCTION

Leiomyosarcoma is the second most common primary retroperitoneal malignant tumor in adults, which commonly originates in the inferior vena cava (1). A leiomyosarcoma arising from the renal vein is particularly uncommon. After the first report of Varela and Garro in 1967, only about 30 cases have been reported in the published medical literature (2, 3). The renal vein leiomyosarcoma is grossly classified into three subtypes: completely extravascular; completely intravascular and mixed (1, 4). To the best of our knowledge, there have been only few reports detailing the imaging findings of renal leiomyosarcoma with both extra- and intravascular components (1, 4). We report an uncommon case of left renal vein leiomyosarcoma with combined components, focusing on the multidetector computed to-

mography (MDCT) and MR imaging findings with review of the literature. This case report was approved by the ethics committee at our institution and an informed consent was waived.

CASE REPORT

A 55-year-old female underwent an abdominopelvic MDCT to evaluate her epigastric pain for two weeks. The CT examination was performed using a 128-detector-row CT scanner (definition AS+, Siemens Medical Solutions, Forchheim, Germany). The enhanced CT scan of the abdomen showed a bi-lobed, heterogeneous, soft-tissue mass in the great vessel space and the left renal vein, measuring 9.5 cm in longest diameter (Fig. 1A, B). The extraluminal component abutted the uncinate process of the pancreas anteriorly, the aorta posteriorly and the portal vein

Received February 19, 2014; Accepted May 8, 2014

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superiorly (Fig. 1A, C). An expansile and branch-shaped intraluminal component occupied the lumen of the left renal vein, seen with a thin splaying of contrast medium (Fig. 1B). An abdominal MR imaging was performed for further evaluation on a 3.0T system (Magnetom Verio; Siemens Medical Solutions, Erlangen, Germany), using a body phased-array coil. The bi-lobed mass appeared isointense relative to the adjacent muscle on T1-weighted images (Fig. 1D). The extraluminal component

showed a relatively low signal intensity and the intraluminal component showed an intermediate signal intensity with high-signal-intensity foci on T2-weighted images (Fig. 1E, F). Contrast-enhanced, fat-suppressed T1-weighted images showed a heterogeneous enhancement of the mass with a poorly enhancing portion, suggesting necrosis (Fig. 1G, H). The preoperative presumed diagnosis was either retroperitoneal sarcoma, including leiomyosarcoma or non-functioning neuroendocrine tumor

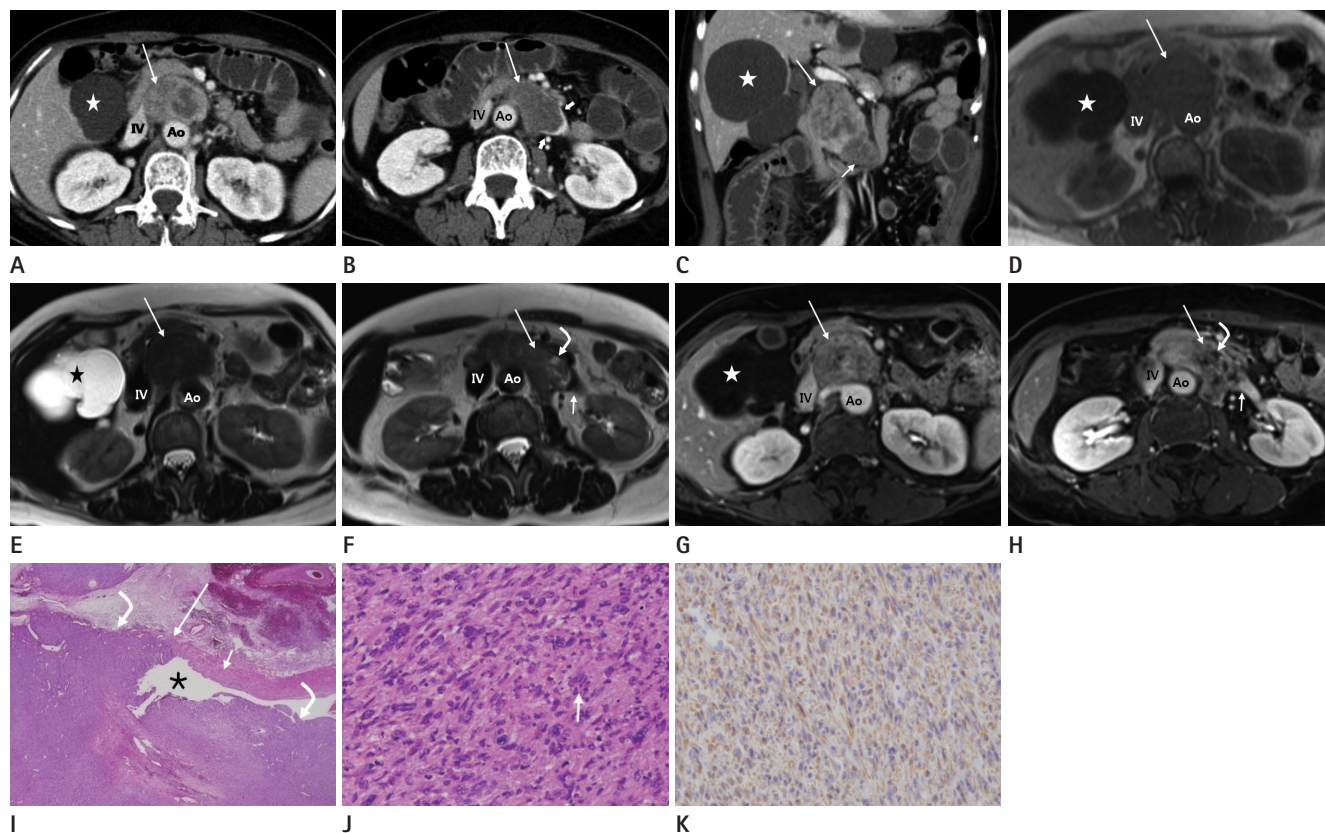


Fig. 1. 55-year-old woman with left, renal-vein leiomyosarcoma.

A, B. Enhanced axial CT scans obtained at the level of the pancreas uncinate process (**A**) and the renal hila (**B**) show a heterogeneous, soft-tissue mass, retroperitoneal mass (long arrows in **A, B**) in the lumen of the left renal vein with thin splaying of contrast medium (short arrows in **B**); the mass abuts the uncinate process of the pancreas anteriorly.

C. Coronal reformatted image demonstrates the extraluminal component of the tumor (long arrow) abutting the portal vein superiorly and the intraluminal component (short arrow) in the left renal vein.

D–H. On axial T1-weighted MR images (**D, E**), the mass (arrow) appears isointense relative to the muscle (**D**). On T2-weighted (**E, F**) MR images, the extraluminal component (arrow in **E**) of the mass shows low signal intensity and the intraluminal component (arrow in **F**) appears heterogeneous, although relatively iso- to hyperintense. The intraluminal component has a high-signal-intensity portion (curved arrow in **F**), a finding that indicates necrosis. Contrast-enhanced, fat-suppressed, T1-weighted MR images (**G, H**) reveal heterogeneous enhancement of the mass (arrow in **G, H**), and a poorly enhancing portion (curved arrow in **H**) suggests necrosis. Splaying of the contrast medium is also noted in the left renal vein (short arrow in **H**).

I. Microscopic examination (original magnification, × 12; H&E stain) illustrates a transition (long arrow) between the leiomyosarcoma (curved arrows) and the intima of the renal vein wall (short arrow). The findings confirm that the tumor arose from the wall of the renal vein. The lumen (asterisk) of the left renal vein is noted.

J. Photomicrograph (original magnification, × 200; H&E stain) shows typical histologic features of leiomyosarcoma: interlacing fascicular pattern with nuclear atypia and mitosis, proliferating spindle-shaped cells (arrow).

K. Immunohistochemical staining (original magnification, × 200) is positive for smooth muscle actin, which appears brown.

Note.—Ao = abdominal aorta, IV = inferior vena cava, star = incidental hepatic cyst

with renal-vein thrombosis.

A complete excision of the tumor was performed. After the tumor and the affected segment of the left renal vein were removed, an 8 mm Dacron graft was used for end-to-end anastomosis between the left renal vein and the inferior vena cava. On gross examination, the extraluminal component measured 6.5-cm and the intraluminal component measured 3.5-cm in the longest diameter. The mass was gray and soft and showed hemorrhagic foci. The microscopic examination showed typical histologic features of leiomyosarcoma, including an interlacing fascicular pattern of proliferating spindle-shaped cells with nuclear atypia as well as a transition between the leiomyosarcoma and the intima of the renal-vein wall (Fig. 1I, J). These findings confirmed that the tumor arose from the wall of the renal vein. Immunohistochemical staining was positive for smooth muscle actin, appearing as brown (Fig. 1K).

DISCUSSION

Approximately 5% of leiomyosarcomas originate from the smooth muscles of the large blood vessels (5). The inferior vena cava is responsible for more than 50% of retroperitoneal leiomyosarcoma cases (2, 5). Primary leiomyosarcoma originating from the renal vein is quite uncommon with 50 to 60 years as prevalence peak age. It more commonly arises from the right renal vein compared to the left renal vein (2, 4). Long-term outcomes are closely related to tumor size and the feasibility of complete tumor resection (5).

On MDCT, a small leiomyosarcoma presents as a solid, soft-tissue density mass that is well-marginated and homogenous. However, a large mass can show necrosis and occasionally hemorrhage (6). Leiomyosarcoma shows low or intermediate signal intensity on T2-weighted MR images and low-intermediate signal intensity on T1-weighted MR images. It may present with several different degrees of enhancement and delayed enhancement compared to skeletal muscle (7). The signal intensity and enhancement seen on MRI depends on the amount of muscular and fibrous components and the extent of necrosis (8). In our case, both extraluminal and intraluminal components showed intermediate signal intensity on T2-weighted images with a portion of high-signal intensity observed inside the mass. The intraluminal component showed a fewer enhancement than the extraluminal component,

probably because the intraluminal component contained more necrotic tissue and hemorrhagic foci. Unlike other retroperitoneal sarcomas that show a growth pattern which encases and narrows the renal vessel, neoplasms originating from the vessel wall show an intraluminal growth pattern or an intravenous propagation of the tumor (9). The presence of a solid and necrotic extravascular component with a contiguous intravascular enhancing portion is a more suggestive imaging finding of a retroperitoneal leiomyosarcoma (9).

A complete resection of the tumor and the involved renal vein using graft replacement is the treatment of choice. However, even following complete tumor resection, the five-year survival rate has been reported to be between 31% and 62% as more than half of these patients develop a tumor recurrence (4, 10).

In conclusion, the imaging findings of leiomyosarcoma are relatively non-specific. However, imaging findings such as an intraluminal component in the renal vein, heterogeneous enhancement due to combined necrosis and portions of low- or intermediate-signal intensity seen on T2-weighted MR images can suggest the diagnosis of a primary leiomyosarcoma of the renal vein.

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혈관 내외 구성요소를 모두 갖는 신정맥 기원 평활근육종의 다중 검출기 전산화단층촬영 및 자기공명영상 소견: 증례 보고¹

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정맥 기원의 평활근육종은 드문 종양으로 주로는 하대정맥에서 기원하며 신정맥 기원의 평활근육종은 매우 드물다고 알려져 있다. 우리는 55세 여자 환자의 신정맥에서 기원한 평활근육종을 경험하여 이에 대해 보고하고자 한다. 후복막강 종괴가 있을 때, 신정맥 내에서 기원하는 것으로 보이는 혈관 내 종괴가 있고 자기공명영상의 T2 강조영상에서 저신호 내지는 중등도의 신호강도를 보인다면 신정맥 기원의 평활근육종을 진단할 수 있다.

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