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Case Report

CT Findings of Primary Renal Angiosarcoma

Hisashi Komoto[a] Kazuhiro Kitajima[a] Yusuke Kawanaka[a] Nahomi Yoshimura[a] Ryo Kunimoto[a] Hiroyuki Yokoyama[a] Yukako Shinkai[b] Yohei Kaizuka[b] Shingo Yamamoto[b] Takako Kihara[c] Neinei Kimura[c] Seiichi Hirota[c] Koichiro Yamakado[a]

[a]Department of Radiology, Hyogo College of Medicine, Nishinomiya, Japan; [b]Department of Urology, Hyogo College of Medicine, Nishinomiya, Japan; [c]Department of Surgical Pathology, Hyogo College of Medicine, Nishinomiya, Japan

Keywords
Angiosarcoma · Kidney · Computed tomography

Abstract
Primary angiosarcomas of the kidney are very rare but highly aggressive tumors showing poor prognosis. We present a case of primary renal angiosarcoma occurring in a 60-year-old man with left flank pain. CT images depicted a huge exophytic mass (14 cm in diameter) in the left kidney, exhibiting central extensive hemorrhage or necrosis without contrast enhancement. The mass showed centripetal peripheral nodular enhancement on dynamic contrast-enhanced CT images. We suggest its inclusion in the differential diagnosis of cases of hemorrhagic renal tumors with prominent vasculature.

Introduction

Angiosarcoma is a rare malignant tumor accounting for less than 2% of all soft tissue sarcomas [1]. Approximately one-third of primary angiosarcomas occur in skin, one-third in soft tissue, and the remaining one-third in other sites like bone, breast, and liver. Primary renal angiosarcomas are exceedingly rare, but highly aggressive tumors showing poor prognosis [2]. These tumors are predominantly found in older men (60–70 years of age), and patients frequently present with flank pain and a palpable mass [3, 4]. Due to the low incidence of this

Kazuhiro Kitajima
Department of Radiology, Hyogo College of Medicine
1-1 Mukogawa-cho, Nishinomiya
Hyogo 663-8501 (Japan)
kazu10041976@yahoo.co.jp

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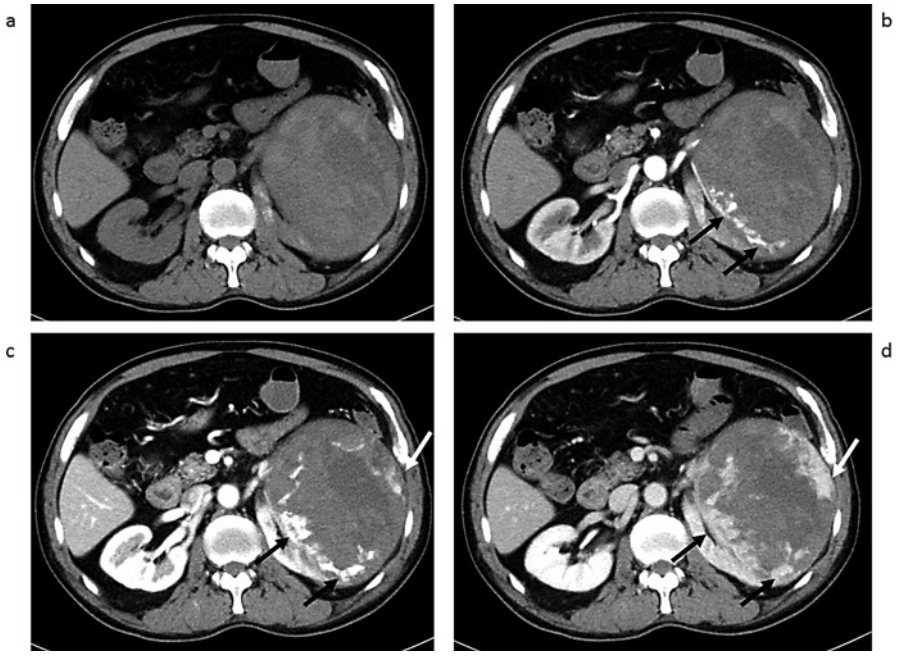

Fig. 1. Axial CT images of pre-contrast (**a**), contrast-enhanced arterial (**b**), corticomedullary (**c**), and nephrographic phases (**d**) depict a 14 × 13 × 11 cm mass with extensive hemorrhage or necrosis in the left kidney, which shows progressive peripheral nodular enhancement with a delayed fill-in (arrows).

tumor, angiosarcoma is not usually considered in the diagnosis of renal tumors associated with retroperitoneal hemorrhage [5]. In this report, we describe a case of primary renal angiosarcoma showing extensive hemorrhage and peripheral nodular early enhancement with an emphasis on dynamic contrast-enhanced computed tomography (CT) imaging features.

Case Report

A 60-year-old man was admitted to our hospital with a complaint of discomfort in the left flank area starting 2 weeks earlier, but no hematuria. The initial routine laboratory tests showed the hemoglobin level 10.4 g/dL (normal range: 12–18 g/dL) and hematocrit 30.6% (normal range: 40–52%). The remaining blood tests were normal, and creatinine and urinalysis were within normal limits.

CT images depicted a huge exophytic mass, measuring 14 × 13 × 11 cm in diameter, in the superior pole of the left kidney (Fig. 1, Fig. 2). The mass also exhibited central extensive slight mixed high- and low-dense area on non-enhanced phase, showing no contrast enhancement, suggesting hemorrhage or necrosis. The mass showed peripheral nodular enhancement, as shown on arterial-phase CT, accompanied by delayed centripetal filling and corticomedullary and nephrographic-phase CT images. There were neither lymphadenopathy nor additional mass lesions observed in other solid organs in the abdomen. Based on these imaging findings, the differential diagnoses included hemangioma, angiosarcoma, angiomyolipoma, and hemangioendotheliomas.

The macroscopic appearance of the solid tumor measured 14.0 × 12.5 × 11.5 cm. It was located in the upper middle of the left kidney and was dark red in color with marked hemorrhage and necrosis (Fig. 3a). The surgical margins were free of tumor and the tumor did not

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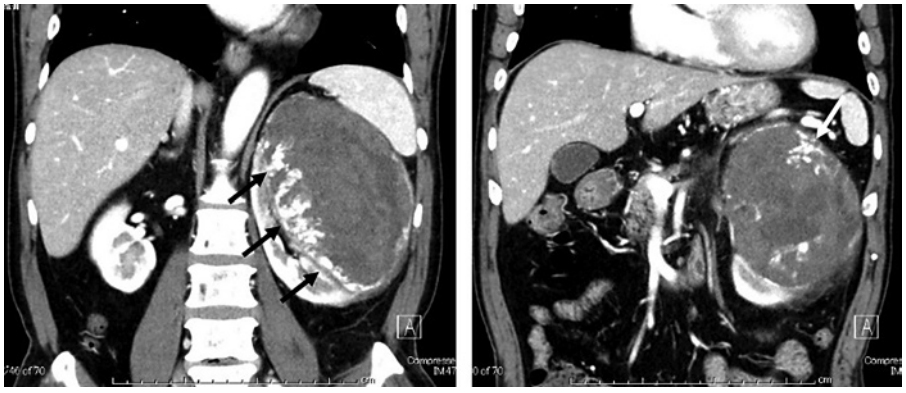

Fig. 2. Reconstructed coronal CT images of corticomedullary phase show the huge exophytic mass in the left kidney, exhibiting central extensive hemorrhage or necrosis without contrast enhancement and centripetal peripheral nodular enhancement (arrows).

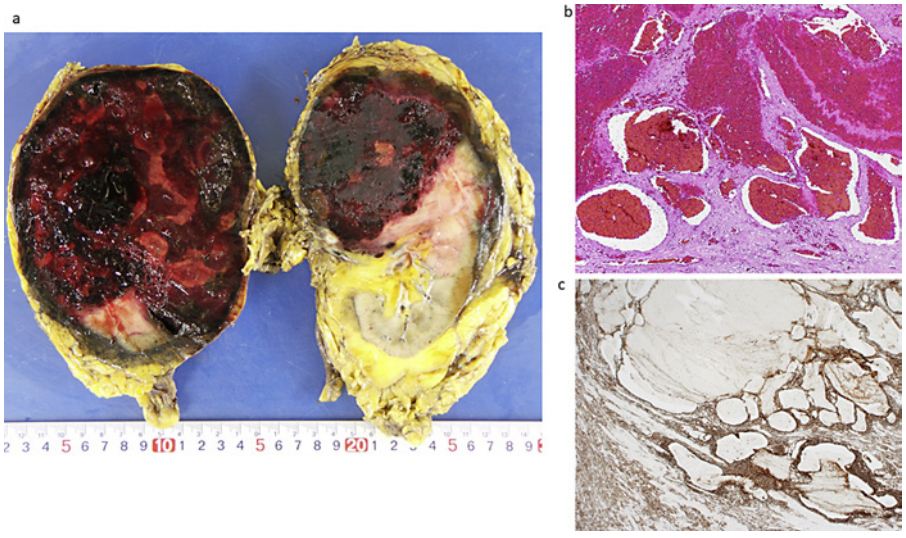

Fig. 3. Resected specimen and pathological finding. **a** Open surgical specimen shows an extensive hemorrhage and necrotic mass in the upper pole of the left kidney. **b** Histological sections reveal proliferation of the tumor vessels and anastomosing channels with obvious vasoformation and dissecting growth pattern (H&E stain). **c** Immunostain for CD31 shows intense staining of most cells.

involve perinephric fatty tissue, ureter, and adrenal gland. The microscopic features revealed proliferation of the tumor vessels and complex anastomosing channels with obvious vasoformation and endothelial papillae (Fig. 3b). An immunohistochemical study showed a vascular tumor strongly and diffusely positive for vimentin, CD31 (Fig. 3c), CD34, and p53, and partially positive for Ki67 (about 40%), while negative for AE1/AE3, EMA, CK7, and CD10. These findings indicated that the tumor was angiosarcoma of the kidney.

No adjuvant chemotherapy or radiation therapy were performed due to the patient's wish. After 7.6 months from the initial surgery, a CT scan showed local recurrence, peritoneal dissemination to the small bowel, and hepatic metastasis. One week later, he underwent the second surgery including tumor resection and partial resection of small bowel. After 9.2 months from the initial surgery, he died due to this disease.

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Discussion

Angiosarcoma is an aggressive malignant neoplasm originating from endothelial cells of the blood and lymphatic vessels [6, 7]. Among the various malignant tumors that can occur in the kidney, primary angiosarcomas are extremely rare, with only about 68 cases reported to date in the literature published in 2019 [8]. Although the presence of several risk factors, such as thorotrast, arsenic, polyvinyl chloride, radiotherapy, and chronic lymphedema, have been reported in angiosarcomas arising at other sites in the body, there is no evidence of a direct relationship between these predisposing factors and primary angiosarcomas of the kidney [3–5]. The tumors are usually large, measuring from 3.7 to 30 cm in diameter, and are detected in advanced stages of the disease [4]. As the tumor has a tendency to bleed, patients may also complain of massive hematuria and a retroperitoneal hematoma following spontaneous rupture of the mass [6]. Histologically the tumor is composed of anastomosing vascular spaces lined by malignant cells containing nuclei with pleomorphism and mitotic activity. Immunohistochemical staining is useful in further characterizing the tumor. It stains positive for CD34, CD31, and factor VIII-related antigen. Most tumor cells stain negative for CD10, HMB45, and EMA. Thus, a panel of markers is needed to conclusively diagnose this rare renal tumor [3].

With respect to imaging findings, the tumors have been described as an extensive hemorrhagic or necrotic mass with variable peripheral enhancement similar to our series, or a large necrotic mass [2–6, 8, 9]. However, there are very few useful imaging features, suggestive of primary renal angiosarcomas. Although magnetic resonance imaging (MRI) examination was not conducted in this case, Heo et al. [9] reported that T2-weighted imaging demonstrated a tangled mesh of tumor vessels with signal voids in the periphery of the mass, corresponding to the areas with strong enhancement on contrast-enhanced MRI.

The differential diagnosis includes hemangioma, angiomyolipoma, hemangioendothelioma, and renal cell carcinoma.

Due to the rarity of this tumor, there are no standard treatment guidelines for primary renal angiosarcomas [2–4, 6]. However, most of the reported cases involved patients who underwent radical nephrectomies [3, 4]. Radiation therapy and chemotherapy may be subsequently used in localized and metastatic disease, respectively [2, 4]. Anti-angiogenesis factors like the vascular endothelial growth factor (VEGF) receptor blockers have been used successfully in the management of angiosarcoma recently and their use can be extended to renal angiosarcoma in the future [10]. The prognosis is very poor, with more than 70% of the reported cases dying within a mean interval of 7.3 months [4]. The prognosis of primary renal angiosarcoma depends on the tumor size and metastasis status at presentation [5]. The primary tumor with size more than 10 cm tends to show poorer prognosis. Metastases are found mainly in the lung, liver, and bones.

Conclusion

We present a case of primary renal angiosarcoma with extensive hemorrhage and suggest its inclusion in the differential diagnosis of cases of hemorrhagic renal tumors with prominent vasculature.

Statement of Ethics

This report complies with the guidelines for human studies and includes evidence that the research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. The authors have no ethical conflicts to disclose. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Concept and design, H.K., K.K.; acquisition of data, H.K., K.K., Yu.K., N.Y., R.K., H.Y., Y.S., Yo.K., S.Y., T.K., N.K., S.H.; drafting of the manuscript, K.K.; critical revision of the manuscript for important intellectual content, S.Y., T.K., K.Y. All authors approved the final version of the manuscript.

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