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Case Report

Granulomatosis with polyangiitis presenting as a solitary renal mass: A case report with imaging and literature review

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ABSTRACT

Granulomatosis with polyangiitis (GPA) manifests as necrotizing granulomatous inflammatory masses in the nasal cavity, paranasal sinuses, and lungs. However, a mass in the kidney is extremely rare. We herein report a case of GPA that presented as a solitary mass in the left kidney. The patient was a man in his 60s. A 2.5-cm solitary mass was incidentally discovered in the left kidney at another hospital and was followed-up. Eight months later, the mass had enlarged, and the patient visited our hospital for further examination and treatment. The mass was hypovascular, with unclear margins on contrast-enhanced computed tomography (CT). The signal of the mass was nonuniform and iso- to slightly hypo-intense on T2-weighted and diffusion-weighted magnetic resonance imaging (MRI). Enlarged para-aortic lymph nodes were also detected on the CT and MRI. Based on imaging, malignant tumors were suspected, and nephrectomy was performed. The pathological diagnosis was GPA. We performed a literature review of this rare renal manifestation and present a summary of reported imaging findings. If a hypovascular renal mass with an unclear margin can be found in those with GPA, unnecessary operations may be avoided by actively promoting renal biopsy.

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Introduction

Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis, is an uncommon immunologically mediated systemic disease of unknown etiology that is pathologically characterized by an inflammatory reactive pattern (necrosis, granulomatous inflammation, and vasculitis). GPA often occurs in the upper and lower respiratory tracts and in the kidneys [1]. GPA is an autoimmune multisystemic small vasculitis that is included in the group of anti-neutrophil cytoplasmic antibody (ANCA)-associated small-vessel vasculitis [2]. GPAs of the nasal cavity, the paranasal sinuses, and the lungs are well known, but renal masses are extremely rare.

Case Report

The patient was a man in his 60s. A 2.5-cm solitary mass was incidentally discovered in the left kidney at another hospital and was followed-up. Eight months later, the mass had enlarged, and the patient visited our hospital for further examination and treatment. The patient's existing conditions included Sjögren's syndrome and pneumoconiosis. There were no additional physical findings, such as back pain. Blood tests showed that there was no decrease in renal function. However, increased levels of C-reactive protein (2.44 mg/dL; normal range, 0–0.3 mg/dL), immunoglobulin G4 (2,092 mg/dL; normal range, 870–1700 mg/dL), and soluble interleukin-2 receptor (672 U/mL; normal range, 145–519 U/mL) were noted. There were no abnormal findings in the urine test.

Dynamic contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) were performed. Non-contrast-enhanced CT images revealed a 7.0 cm × 5.5 cm × 6.0 cm mass in the center of the left kidney with a slightly higher absorption than the surrounding renal parenchyma. The mean CT values of the mass were 44 HU, 62 HU, and 72 HU during the noncontrast enhanced, corticomedullary, and nephrogenic phases, respectively (Fig. 1). The boundary between the mass and the renal parenchyma was unclear. The pseudocapsule on the limb of the mass was also unclear. Compared to the CT images taken at another hospital 8 months ago, the

mass had rapidly grown from 2.5 cm to 7 cm in the major axis (Fig. 2a and b). The poor-contrast enhanced area developed in the center of the mass (Fig. 2b), and an enlarged para-aortic lymph node ipsilateral to the mass appeared (Fig. 2c).

MRI (3.0 T) showed an iso-intense mass on T1-weighted image and iso- to slightly hypo-intense mass relative to the kidney on T2-weighted image. There was no pseudocapsule around the mass. On T2WI, a strong low-signal area was found in a part of the renal subcapsule (Fig. 3b). Diffusion-weighted imaging ($b = 1000 \text{ s/mm}^2$) showed nonuniform and iso- to slightly high signal intensity. The apparent diffusion coefficient map was nonuniform and iso-to slightly hypo-intense. The apparent diffusion coefficient values ranged from 0.84×10^{-3} to $1.45 \times 10^{-3} \text{ mm}^2/\text{s}$.

Ultrasound-guided biopsy was performed, but the definitive diagnosis could not be determined. A left nephrectomy was performed because the tumor was growing so rapidly.

Macroscopically, a yellowish white mass with a diameter of 9 cm × 7.5 cm was confirmed in the center of the kidney. No pseudocapsule was found between the mass and renal parenchyma, but the renal capsule in contact with the mass had thickened. Part of the capsule was also markedly thickened (Fig. 4a). This was consistent with the area of strongly low signal on T2-weighted image. Findings of the histopathological examination showed granuloma with extensive fibrosis, necrosis, and aggregation of neutrophils (Fig. 4b). In the renal capsule with markedly thickening fibrosis, a granuloma was noted (Fig. 4c). Granuloma and vasculitis with fibrinoid degeneration were also confirmed in the arteries around the ureter and fat tissues around the renal sinus (Fig. 4d). Thus, GPA was suspected. A resected para-aortic lymph node showed reactive lymphadenopathy.

Subsequently, the patient's blood test results showed that myeloperoxidase (MPO)-ANCA was positive (10.4 U/mL; upper limit, 3.5 U/mL), and a definitive diagnosis of GPA was made. Consequently, prednisolone (30 mg/day) was administered. Following prednisolone treatment, the MPO-ANCA turned negative, and there has been no relapse for the past 2 years.

Chest CT before nephrectomy did not present any findings of GPA in the lungs. After the definitive diagnosis was made, paranasal sinus CT was performed, but there were also no findings suggestive of GPA.

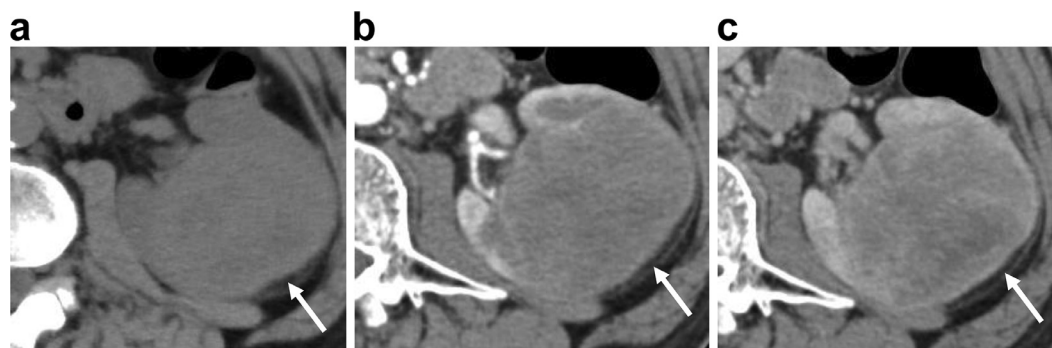


Fig. 1 – Dynamic contrast-enhanced computed tomography. (a) Unenhanced, (b) corticomedullary phase, and (c) nephrogenic phase. There is a 7 cm × 5.5 cm × 6 cm mass in the center of the left kidney (arrow). The contrast effect is weak and incremental: (a) 44 HU, (b) 62 HU, and (c) 72 HU. The border between the mass and renal parenchyma is unclear.

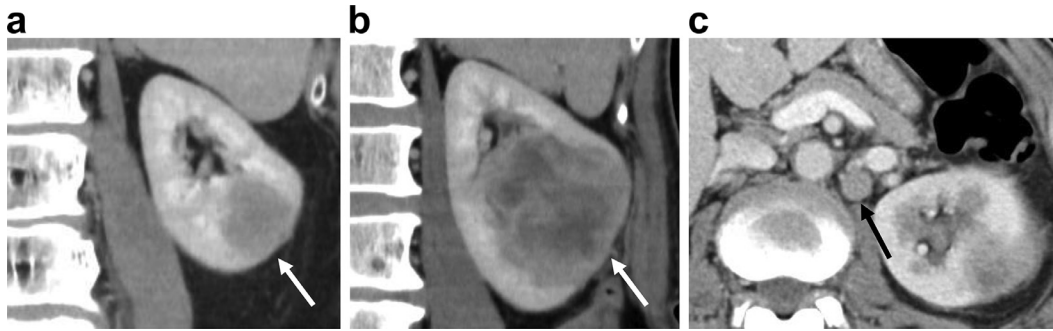


Fig. 2 – (a) Contrast-enhanced computed tomography (CT) image taken at another hospital 8 months ago. (b and c) Contrast-enhanced CT images taken at our hospital. The tumor grew from 2.5 cm to 7 cm in length, in 8 months. Poor-contrast area inside of the mass (b), and the ipsilateral para-aortic lymph node is enlarged (c, arrow).

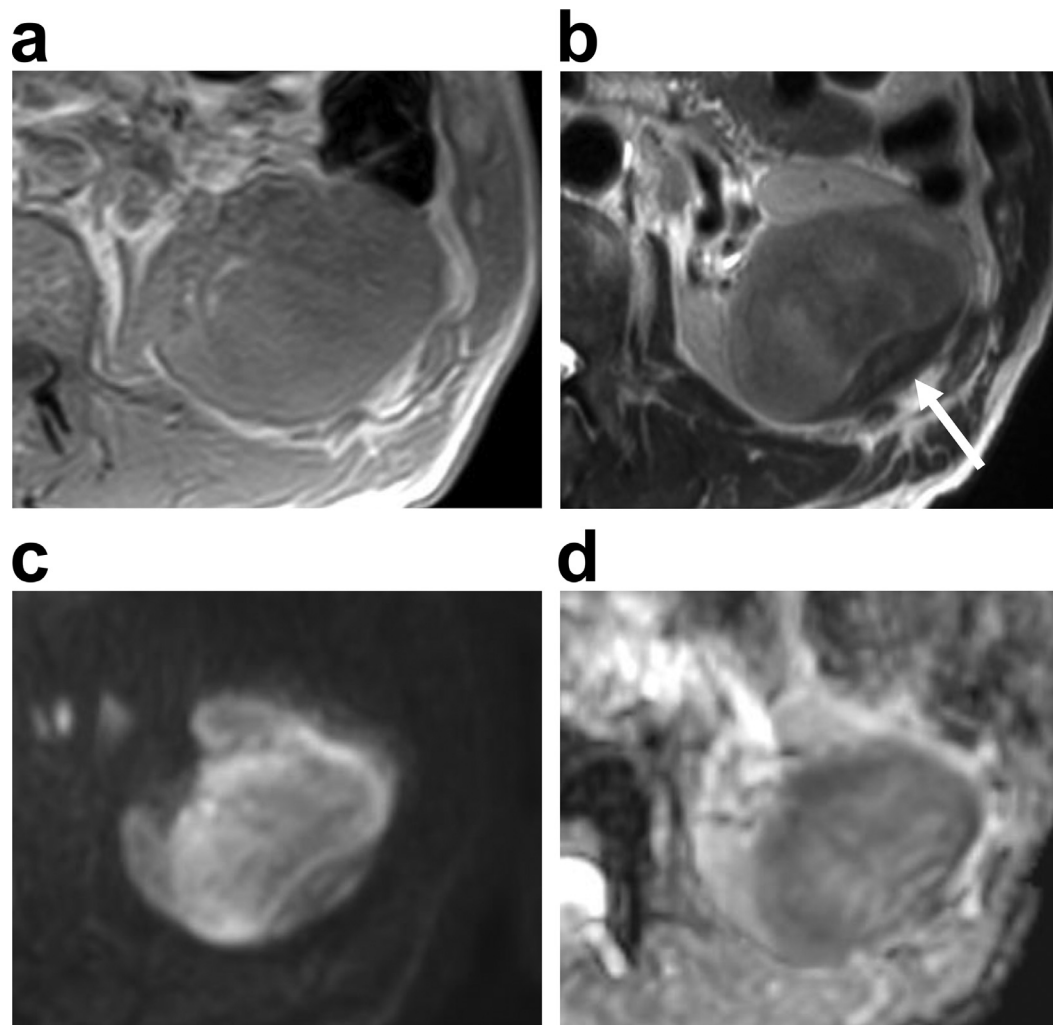


Fig. 3 – Magnetic resonance imaging (3.0 T). (a) T1-weighted image shows iso-intense mass relative to the kidney. (b) T2-weighted image shows nonuniform iso- to hypo-intense mass relative to the kidney. There is no pseudo-capsule around the mass. A convex lens-shaped strong low-signal area is found in renal subcapsule (arrow). (c) Diffusion-weighted image ($b = 1,000 \text{ s/mm}^2$) presents nonuniform and iso- to slightly high-intensity. (d) Apparent diffusion coefficient (ADC) map presents nonuniform and iso- to slightly hypo-intensity, with ADC values ranging from 0.84×10^{-3} to $1.45 \times 10^{-3} \text{ mm}^2/\text{sec}$.

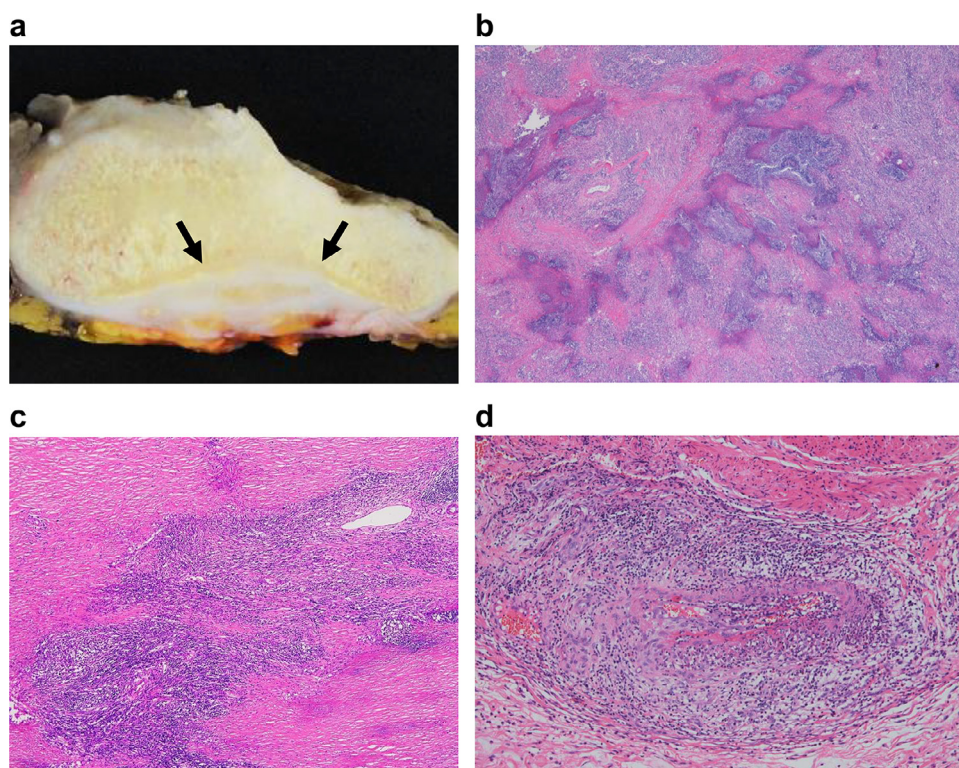


Fig. 4 – Resected specimen (a) and pathological specimens (b-d). (a) Macroscopically, a 9 cm x 7.5 cm yellowish white mass is found in the center of the kidney. Resected specimen shows thickening of the renal capsule in contact with the GPAs and marked thickening of part of the capsule (arrow). Hematoxylin and eosin staining. (b) 20 x magnification, (c) 40 x magnification, and (d) 100 x magnification. Histologically, the lesion shows granuloma with extensive fibrosis, necrosis, and aggregation of neutrophils (b). Granuloma was noted in the renal capsule with markedly thickening fibrosis (a, arrow), (c). Vasculitis with fibrinoid degeneration was confirmed in the small arteries around the ureter (d).

Discussion

GPA is included in ANCA-associated vasculitis (AAV) and classified as a group of small-vessel vasculitis [3]. Most patients with AAV in Europe and the US have GPA, and 80–90% of GPA patients have proteinase 3 (PR3)-ANCA [4]. However, in Japan, more than 80% of AAV patients were MPO-ANCA-positive [5], and 54.6% of GPA patients were positive for MPO-ANCA [6]. The International Chapel Hill Consensus Conference (CHCC2012) defines the pathological features of GPA as necrotizing granulomatous inflammation usually involving the upper and lower respiratory tracts, necrotizing vasculitis affecting predominantly small to medium vessels, and necrotizing glomerulonephritis [3]. A necrotizing granulomatous inflammatory mass in the kidney due to GPA is extremely rare. To examine the characteristic imaging findings of this rare condition, we performed a literature review using PubMed and searched papers using English keywords “granulomatosis with polyangiitis,” “Wegener’s granulomatosis,” and “renal mass or pseudotumor.” In addition, we checked the references from the identified papers. A total of 24 cases were selected [7–27]. In 21 of 24 cases, the sites of the mass could be confirmed, and there were three patterns of appearance. The solitary-type (62%, 13/21) was the most common, followed by multiple

bilateral masses (28.5%, 6/21), and then multiple unilateral masses (9.5%, 2/21).

In 15 cases, contrast-enhanced CT findings were confirmed [11–24,27]. Contrast-enhanced CT findings from previous reports were limited to 1 phase. To our knowledge, our report is the first to present the dynamic contrast-enhanced CT findings. As a result of the review, a number of distinctive findings were also observed. Findings such as hypovascular mass (100%, 15/15) and unclear margins (93%, 14/15) were present in almost all cases [11–24,27]. Although the opacity of peri-renal fat was not confirmed in our case, this finding was noted in 20% (3/15) of the reviewed cases [13,16,18]. Enlargement of the para-aortic lymph nodes, which was also found in our case, was confirmed in 7% (1/15) of reported cases [20].

There were only two case reports with MRI findings [24,25]. One patient underwent contrast-enhanced MRI [24], in which the mass was iso-intense relative to the renal parenchyma on T1-weighted image and nonuniform and slightly hypo-intense on T2-weighted image, and there was no pseudocapsule. These findings were similar to those of our case. The contrast effect was poor. In the other case, only T2-weighted image was provided, and the mass had a high signal [25]. In our case, a convex lens-shaped strong low-signal area was found in the renal subcapsule on T2-weighted image. Pathology showed that the area with a renal

capsule had marked thickening fibrosis, and there was no hematoma.

Although [18F]-fluorodeoxyglucose positron emission tomography/computed tomography (¹⁸F-FDG PET/CT) was not performed in our case, according to reports, there was accumulation of ¹⁸F-FDG in the masses [26,27].

Although not found in our case, half of the reviewed cases (12/24) also had GPA lesions in other sites, including the nasal cavity, paranasal sinus, middle and inner ear, lungs, and/or pituitary gland.

On contrast-enhanced CT, distinguishing features of GPA-induced renal mass, such as hypovascular mass and unclear margin, were found in the reviewed cases and our case. However, these findings are not specific and have been found in many other malignancies, such as renal pelvis carcinoma, collecting duct carcinoma, and malignant lymphoma [28,29].

In our case, the renal capsule in contact with the GPA was thickened with fibrosis. It was thought to be a GPA-induced inflammatory thickening of the renal capsule because no pseudocapsule was found between the GPA and the renal parenchyma. A part of the renal capsule was markedly thickened, and we could identify the findings on T2-weighted image. However, this has not been reported in the past, and it was not possible to know if this was a characteristic finding among GPAs. In our case, the biopsy did not yield a diagnosis, but in many reviewed cases, the biopsy yielded a definitive diagnosis of GPA and patients improved with steroid pulse therapy or immunosuppressive therapy.

If a hypovascular renal mass with unclear margins is found in a patient with GPA or those suspected of GPA, it is important to perform a biopsy, in order to prevent unnecessary surgery.

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