

## Case Report

# Renal failure and abdominal pain as the presenting symptoms of a rare tumor of the aorta masquerading as a calcified plaque

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## Abstract

Vascular obstructive causes must be considered in chronic renal failure with no obvious cause. We present the case of a 74-year-old woman with smoldering renal failure who undergoes a renal biopsy that did not uncover a cause. As her symptoms persisted and renal function worsened, her nephrologist proposed magnetic resonance angiography with gadolinium and prophylactic initiation of hemodialysis. Imaging uncovered an occlusive aortic mass, which was removed surgically with improvement in renal function. The case discusses the nature of the mass and the need to weigh the risks and benefits of MR imaging with gadolinium and initiation of hemodialysis against the risk of nephrogenic systemic fibrosis.

**Keywords:** angiosarcoma; coral reef plaque; gadolinium; renal failure

## Introduction

Obstruction of blood flow to the kidneys must be considered in the differential diagnosis of declining kidney function when other causes have been ruled out. Uncovering such causes may involve imaging—computed tomography or magnetic resonance imaging—and contrast. The clinician must weigh the risks to the patient from the use of iodinated contrast or gadolinium against the benefit of uncovering the cause of the renal failure. We present the case of a 74-year-old woman who was transferred to our hospital after an extensive workup, including kidney biopsy, failed to reveal a cause for her renal failure. The patient underwent MRI with gadolinium after initiation of hemodialysis that uncovered an infrarenal aortic mass. The patient remained off hemodialysis after surgical resection of the mass.

## Case report

The patient is a 74-year-old woman with a 20-year history of hypertension whose serum creatinine had risen to 2.4 from 1.1 mg/dL over several months. She complained of nausea, vomiting, post-prandial abdominal pain and an unintentional 20-pound weight loss. She was anemic. Her kidney impairment did not improve with intravenous fluids. The right and left kidneys measured 8.3 cm and 10.0 cm, respectively, on ultrasound, with minimal blood flow on the right. A 24-h urine revealed 700 mg of protein. A renal biopsy (Figure 1) showed sclerosed glomeruli. ANCA and SPEP were negative, and ANA was equivocal.

She presented to an outside hospital prior to transfer with a dry cough, fevers, chills and headaches. Her serum creatinine rose further to 3.5 mg/dL. Her erythrocyte sedimentation rate was elevated at 100; C-reactive protein was high at 89 mg/L. Rheumatoid factor and dsDNA were negative; complement C3 and C4 were normal. Hepatitis and Lyme serologies were negative. A temporal artery biopsy showed no evidence of giant cell arteritis, but she was started on steroids briefly.

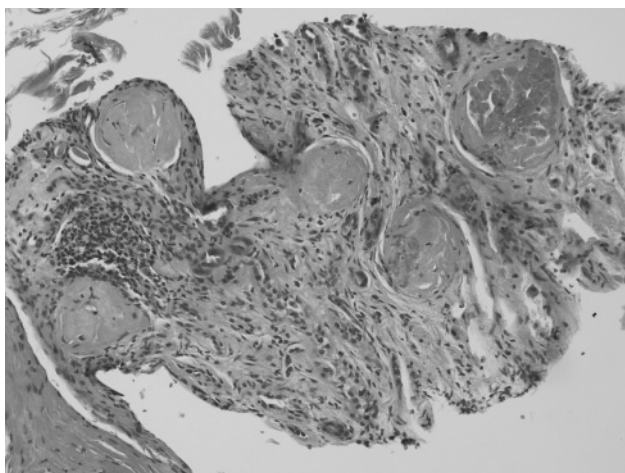
The referring nephrologist suggested a diagnostic MR angiogram to be followed by hemodialysis to remove gadolinium. Dialysis was initiated as her serum creatinine reached 5.7 mg/dL. She underwent MRA and was transferred to the vascular surgery service at this hospital.

On physical examination, temperature was 99.1 F, heart rate 78 beats/min, blood pressure 157–168/67–80 mmHg, respiratory rate 18 and oxygenation 95% on room air with urine output 500 cc over 8 h. She had palpable distal pulses and no bruits in her vascular beds.

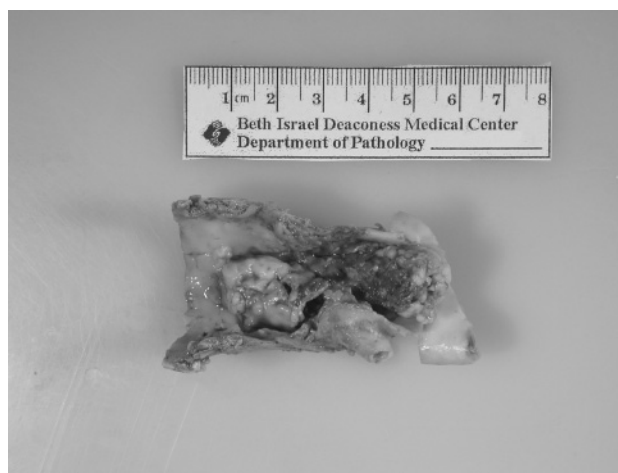
Her urine was straw-colored and clear with specific gravity 1.007, pH 7.0, moderate blood, trace protein and no glucose. Her urine sediment showed no muddy brown or cellular casts. There were no urine eosinophils.

The MRA (Figure 2) showed a mass involving the right anterolateral abdominal aorta. She went to the operating room on hospital day 7 after transfer. The preoperative diagnosis was suprarenal atherosclerotic occlusion. The vascular surgeons removed a large calcified plaque with a ‘cauliflower-like’ soft tissue mass nearly occluding the

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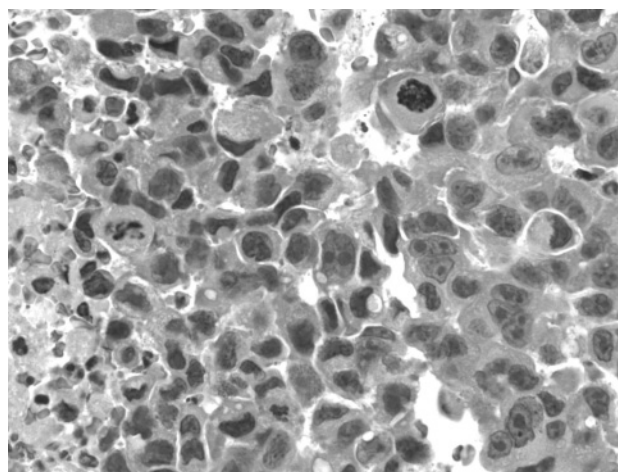
**Fig. 1.** Sclerotic glomeruli at 20× magnification.



**Fig. 3.** Gross surgical specimen of the aorta demonstrates a predominantly necrotic, 5 cm mass, which nearly occludes the suprarenal lumen.



**Fig. 2.** MRA with gadolinium showing a mass primarily involving the right anterolateral aspect of the aorta.



**Fig. 4.** Viable tumor surface is seen composed of poorly differentiated epithelioid cells with large nuclei. An atypical mitotic figure is seen in this field at 60×.

lumen of the aorta (Figure 3). The patient remained off hemodialysis after transfer and surgery. Her serum creatinine declined to 1.6 mg/dL at the time of discharge, post-operative day 24.

The pathology specimen (Figure 4) was subsequently diagnosed as poorly differentiated neoplasm consistent with high-grade epithelioid angiosarcoma, positive at the distal specimen margin but with no invasion of the aortic wall. The oncology consultants suggested chemotherapy and radiation after the patient regained further renal function and strength. She was discharged to a rehabilitation facility.

## Discussion

The case represents an unusual vascular cause of renal failure—an angiosarcoma of the abdominal aorta.

Based on diagnostic imaging, our patient's aortic mass was initially thought to be a 'coral reef' plaque—rock-hard whitish atheromatous material with calcification oc-

cluding the aorta [1]. In one series, presenting symptoms of such plaques included hypertension, claudication, abdominal angina, reduced kidney function and acute renal failure [2]. The mean time between onset of symptoms and operation was 41.2 months. The main reason for delay was a failure by physicians to recognize the disease, which presents as hemodynamic compromise of the kidneys.

In our patient, the surgeons found a large neoplastic mass dominating the aortic lumen. Primary malignant tumors of the aorta are extremely rare, first described in 1873 with a total of 25 reported in the vascular surgery literature to date [3]. The tumor presents clinically as embolic occlusion of a peripheral or mesenteric artery, and diagnosis is made after surgery. Symptoms commonly include claudication, abdominal pain, back pain and fatigue. Aortic angiosarcomas can also masquerade as vasculitis [4,5].

A proposed diagnostic algorithm [6] suggests that magnetic resonance angiography is the most sensitive imaging

for detecting an aortic tumor. Prognosis is poor, with mean survival 12.8 months in one series. Surgical resection is the preferred therapy. European guidelines suggest doxorubicin and ifosfamide as adjuvant chemotherapy.

In this case, initiation of hemodialysis before administration of gadolinium-containing contrast to prevent nephrogenic systemic fibrosis (NSF) presented another challenge to the renal consultant. The strong association between NSF and gadolinium prompted the U.S. Food and Drug Agency to ask all manufactures of gadolinium-containing contrast to update warning labels on their products to include the risk of NSF in patients with an estimated glomerular filtration rate of 30 mL/min/1.73 m<sup>2</sup> or less [7].

Experts recommend weighing the risks of MRI with gadolinium against the risk of not doing the imaging or considering other imaging. If the clinician decides that MRI with gadolinium is essential for patient care, then the lowest dose of gadolinium possible should be used and gadodiamide, which has been implicated in most cases of NSF, avoided. Experts recommend hemodialysis within 2 h of exposure to gadolinium-based contrast and again within 24 h for patients already on hemodialysis and switching to hemodialysis for peritoneal dialysis patients [8]. European guidelines do not recommend initiating hemodialysis for the sole reason of removing gadolinium-based contrast in non-dialysis-dependent patients because of the risks associated with dialysis itself [9]. U.S. guidelines argue that insufficient data exist to recommend initiating dialysis in patients with stage III CKD, and that the risk of initiating hemodialysis should be weighed against the risk of developing NSF in patients with stage IV and V CKD [10].

In our patient, imaging was essential for making the diagnosis of an aortic mass, and surgery led to an unexpected etiology—a rare malignant aortic tumor mas-

querading as a ‘coral reef’ plaque—and resolution of the renal failure.

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*Conflict of interest statement.* None declared.

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