A Rare Cause of Hematuria: Congenital Renal Arteriovenous Malformation

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Abstract
Renal arteriovenous malformation (AVM) is a rare disorder. The diagnosis of the disease is difficult unless it is symptomatic. The most common clinical presentation is hematuria. Hypertension, left ventricular hypertrophy, cardiac failure, and abdominal and/or lumbar pain are other clinical signs of renal AVM. Here we presented an interesting case of a patient with congenital renal AVM who was admitted to our hospital with massive hematuria and was treated with catheter embolization. Renal ultrasonography of the patient demonstrated hematoma in the urinary bladder and solid hyperechoic nodular cortical masses within the right kidney. In contrast-enhanced computed tomography of the abdomen, mild hydronephrosis and approximately 2-cm-diameter saccular aneurysm associated with segmental arteries were revealed in the lower pole of the right kidney. Right renal arteriography showed a large cirsoid-type renal AVM with aneurysm in the lower pole of the kidney. Renal artery embolization was performed in the patient. At follow-up, no recurrence of hematuria was observed. Congenital renal AVM is one of the rare causes of hematuria, and it should be kept in mind that it may lead to macroscopic hematuria. Renal artery embolization can be considered as a safe and effective treatment for renal AVM.

Keywords: Arteriovenous malformation, embolization, hematuria

INTRODUCTION
Renal arteriovenous malformation (AVM) is an uncommon vascular abnormality, including aberrant communications between the intrarenal arterial and venous systems. It can be congenital or acquired. Acquired renal AVM accounts for majority of the cases and is usually a result of previous renal biopsy, trauma, infection, renal surgery, or malignancy. Congenital renal AVM is less common, and 2 subtypes of congenital renal AVM are described. Cirsoid type is described as having a tortuous knotted appearance of numerous feeding vessels and interconnecting fistulas. The other type, called cavernous (angiomatous), has a single vessel feeding multiple small interconnecting vessels (1, 2). The most common clinical presentation of congenital AVM is gross hematuria, whereas acquired renal AVM presents with hemodynamic changes, such as hypertension, cardiomegaly, and heart failure (3-5). We report a rare case of a 27-year-old man who presented to our hospital with gross hematuria because of congenital renal AVM and was treated with renal artery embolization.

CASE PRESENTATION
A 27-year-old man, previously healthy and nonsmoker, was admitted to our hospital with painless gross hematuria. His past surgical history included appendectomy 1 year ago. On physical examination, he appeared pale and tired. His blood pressure was 110/70 mm Hg. Laboratory findings revealed a serum creatinine level of 0.8 mg/dL (76 µmol/L) and hemoglobin level of 9.5 g/dL (hemoglobin level before admission for appendectomy...
my was 15.5 g/dL). The other biochemical and hematological parameters were within normal limits. Iron deficiency was not detected in the patient. Urine microscopic analysis showed 900 erythrocytes/high-power field, and the result of the urine culture was negative. Urine sediment examination was not performed. Renal ultrasonography of the patient revealed hematoma in the urinary bladder and solid hyperechoic nodular cortical masses within the right kidney. A three-way urethral catheter was placed, and continuous bladder irrigation and intravenous hydration were initiated. Ureterorenoscopy showed no ureteral lesion, no active bleeding, no stone, and no lesion in the urinary bladder. Cytological examination was unremarkable. Contrast-enhanced computed tomography (CT) of the patient revealed mild hydronephrosis and approximately 2-cm-diameter saccular aneurysm associated with segmental arteries in the lower pole of the right kidney (Figure 1). Digital subtraction angiography (DSA) was performed to confirm the diagnosis and treatment plan. Right renal arteriography demonstrated a large cirsoid-type renal AVM with aneurysm in the lower pole of the right kidney (Figure 2). At the same time, the patient successfully underwent renal artery embolization (Figure 2b). No complications occurred during or after the procedure, and renal function of the patient remained stable. At follow-up, hematuria had completely regressed, hemoglobin level had returned to normal levels, and the patient was discharged well. At the end of the 1-year follow-up, there was no recurrence of hematuria. Written informed consent was obtained from the patient.

**Main Points**

- Renal arteriovenous malformation may be either congenital or acquired. Acquired malformations are usually caused by trauma or intervention.
- Renal arteriovenous malformation should be considered in patients with gross hematuria for which no alternative explanation has been found in the initial testing.
- Diagnosis is confirmed by digital subtraction angiography, which can be combined with selective embolization therapy in the same setting.

**Figure 1.** Three-dimensional volume rendering image shows right-sided arteriovenous malformation of the kidney (black arrow)

**Figure 2. a, b.** a) Right renal angiography shows a large cirsoid-type renal arteriovenous malformation with aneurysm (black arrows) in the lower pole of the right kidney before embolization. b) Right renal angiography shows a large cirsoid-type renal arteriovenous malformation with aneurysm (black arrows) in the lower pole of the right kidney after embolization
DISCUSSION
Renal AVM is a rare clinical entity. It is characterized by anomalous connections between the arteries and veins through a vascular nidus with a bypass of the capillary bed. Renal AVM can be either congenital or acquired. Approximately 70%–80% of all AVM are acquired and may occur as a result of renal biopsy, blunt or penetrating trauma, inflammation, malignancy, or renal surgery (2). Acquired renal AVM is often termed as renal arteriovenous fistula (AVF). Congenital AVM is different from iatrogenic or traumatic AVF, which is characterized by a single direct communication between an artery and a vein without an intervening vascular nidus (6). Patients with renal AVM are usually asymptomatic. When they are symptomatic, the most common clinical presentation is hematuria. The patients may also present with flank pain, hypertension, and cardiac failure. Renal AVM may be detected in asymptomatic patients by physical examination depending on flank or abdomen bruits. Sometimes, it may be incidentally detected on imaging studies performed for other reasons (7). A limited number of cases have been reported in the literature. In the case reports, clinical presentation can vary between incidental findings to flank pain (8), secondary hypertension (9), cardiomegaly, and intermittent or gross hematuria (2, 3). In clinical practice, it can be difficult to establish a diagnosis of renal AVM only by clinical presentations. DSA is the gold standard for diagnosis and treatment of renal AVM. In our patient, the presenting complaint was gross hematuria. Urinary ultrasonography suggested the presence of renal masses in the right kidney and hematoma in the urinary bladder. Therefore, renal angiomyolipoma, renal infarction, renal vasculitis, and renal cell carcinoma or ureteral/bladder carcinoma were considered in the differential diagnosis. Nevertheless, subsequent contrast-enhanced abdominal CT showed saccular aneurysm. Thereafter, renal angiography confirmed the diagnosis of congenital renal AVM, and the patient was successfully treated with renal artery embolization (10).

CONCLUSION
Renal AVM is an unusual cause of hematuria, which may lead to gross hematuria. Diagnosis of renal AVM should be considered in patients with unexplained massive hematuria. Renal artery embolization can be considered as a safe and effective treatment for renal AVM.

Informed Consent: Written informed consent was obtained from the patient.

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