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Renal arteriovenous malformation presenting with massive hematuria

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Renal arteriovenous malformations are abnormal communications between the intrarenal arterial and venous systems. They are a rare cause of hematuria. Color Doppler ultrasonography, multislice CT angiography, and DSA are important tools for making the diagnosis. We describe the case of a 62-year-old man with renal arteriovenous malformation who presented with gross hematuria.

Introduction

Renal arteriovenous malformations (AVMs) are abnormal communications between the intrarenal arterial and venous systems. Gross hematuria is a frequent complaint in urology clinics worldwide. Common causes of gross hematuria include urinary tract infection, urolithiasis, and neoplasms of the genitourinary tract. Renal AVMs are among the rare causes of gross hematuria. We describe the case of a 62-year-old man with renal AVM who presented with gross hematuria.

Case report

A 62-year-old male presented to our institute with a history of gross hematuria with passage of blood clots in urine since early morning. His physical examination reports were normal except for pallor. Blood pressure at the time of presentation was 100/70, and heart rate was 106/min. His abdomen was soft. ECG and chest radiography were normal. The patient had a history of radical left nephrectomy for renal-cell carcinoma two years ago. There was no his-

tory of surgery, trauma, or any instrumentation in the right kidney. He had no personal or family history of bleeding disorders. He was not taking any medication. There was no history of drug abuse or smoking. Blood analysis showed hemoglobin 10gm/dl (normal range, 12.4 to 14.9 gm/dl). His biochemical and coagulation parameters were within normal limits. Urine analysis showed plenty of RBCs. There was no evidence of pus cells or crystals in the urine. Ultrasound of the abdomen was advised.

Ultrasound, performed on an Acuson 500 machine from Siemens using a 3.5-MHz transducer, showed no evidence of kidney or bladder calculus. Large clots were seen in the urinary bladder. There was no evidence of hydronephrosis. There was a cystlike anechoic area seen at the midpole of the right kidney; this was confirmed to be a focal aneurysmatic dilatation with heterogeneous fill-in on color Doppler. Turbulent blood flow with maximum velocity of 455 cm/sec was obtained, indicating a fistula on spectral analysis. There was pulsatile flow detected in the renal vein. CT angiography was advised.

CT angiography, done on a Somatom Sensation 64-slice CT scanner from Siemens, demonstrated aneurysmatic dilatation of the segmental branch and early opacification of the right renal vein on the arterial phase, suggestive of renal AVM (Figs. 1 and 2). All other abdominal arteries and organs were unremarkable. There was no evidence of arteriosclerosis or arteritis. The patient was catheterized, draining gross hematuria with a substantial amount of clots. A bladder clot evacuation was performed, after which he was started on continuous bladder irrigation and intravenous antibiotic coverage. He underwent arterial embolization on the next day.

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Fig. 1. 62-year-old male with renal arteriovenous malformation. Findings: Maximum intensity projection, in axial plane of CT renal angiography in arterial phase demonstrating aneurysmal dilatation of segmental branch (arrowhead) of main renal artery (black arrow) with early opacification of renal vein (white arrow), suggestive of arteriovenous malformation. Technique: Scanner: Siemens Somatom sensation 64, 200Eff mAs, 120 kV, slice thickness 10mm, intravenous contrast: 100 ml Omnipaque 350 mg/dl.

Discussion

Renal arteriovenous malformations (AVMs) or arteriovenous fistulas (AVFs) are abnormal communications between the intrarenal arterial and venous systems. Renal AVM is relatively rare, with an estimated incidence of less than 0.04% (1, 2). Renal AVM can be idiopathic, congenital, or acquired.

Congenital arteriovenous malformations are considered to represent focal spontaneous failures of vascular development occurring during 1st trimester of gestation (3). They make up about 25% of all cases, and usually present in the third to fourth decade of life. Females are three times more likely to be diagnosed with renal AVM than males, with the right kidney slightly more often affected than the left (3). Renal AVM is sometimes associated with genetic disorders such as hereditary hemorrhagic telangiectasia (4).

There are two types of congenital AVM. The most common is the cirroid type. It has multiple communicating vascular channels that form a masslike cluster that lies deep to the uroepithelium in the lamina propria. The less common type is the cavernous (aneurysmal) AVM, which consists of a single feeding artery and a single draining vein communicating via a cavernous chamber. Acquired renal AVMs are frequently referred to as arteriovenous fistulas. This is the most common type (70 to 75% of all cases), frequently occurring due to iatrogenic trauma such as renal

biopsy or surgery. It may also occur due to blunt or penetrating trauma, pyelonephritis, and renal-cell carcinoma. Idiopathic renal AVM composes only up to 5% of all cases; it is thought to be caused by spontaneous erosion or rupture of a renal artery into a neighboring renal vein.



Fig. 2. 62-year-old male with renal arteriovenous malformation. Findings: Volume-rendered oblique coronal image of right kidney in arterial phase of CT angiography showing aneurysmal dilatation of segmental branch at lower pole with early opacification of renal vein, suggestive of arteriovenous malformation. Technique: Scanner: Siemens Somatom sensation 64, 200Eff mAs, 120 kV, slice thickness 10mm, intravenous contrast: 100 ml Omnipaque 350 mg/dl.

The patient may remain asymptomatic; the AVM may be detected in asymptomatic individuals during physical examination based on a frank bruit or may be incidentally detected on imaging studies performed for other reasons. The cirroid type of renal AVM frequently causes hematuria, as it is commonly found beneath the mucosa of the renal collecting system; gross hematuria as the primary complaint was reported in 72% of cases (5). Hematuria results from minute rupture of the thin-walled veins into the collecting system from increase in pressure. This is why even small, peripherally located AVMs can cause massive hematuria (3). There are few reported cases of AVMs in asymptomatic patients with microscopic hematuria (6). Patients may present with flank pain from obstruction of the renal collecting system by blood clots. The idiopathic or acquired types cause abdominal bruit, hypertension, headache, palpitation, cardiomegaly, or congestive cardiac failure resulting from the large amount of blood flowing through the AVF (7).

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Several imaging modalities have been used to evaluate renal AVMs and AVFs. These include CT scan, Doppler ultrasound, and MRI. However, arteriography is still used as the gold standard to confirm this diagnosis. On grayscale ultrasound, small AVFs usually remain invisible. The large AVMs and AVFs appear as uni- or multilocular cystic lesions, or they may simulate hydronephrosis. Color and spectral Doppler studies show aliasing and high-velocity, turbulent flow with low resistive index in AVMs/AVFs. The arterial feeder shows high peak systolic velocity and low resistive index. The draining vein shows an arterial flow pattern and pulsation. On angiography, a cirroid AVM appears as tortuous veins with multiple communications between arteries and veins. The cavernous type of AVM appears as a single artery and vein connected via a dilated chamber. An acquired AV fistula is characterized as a solitary communication between an artery and a vein. Idiopathic AVF has a similar angiographic appearance to that of the acquired type.

Our patient had a left renal mass two years ago for which he underwent contrast-enhanced CT in a private hospital, but angiography was not performed at that time, so the renal AVM remained undetected. This explains the importance of proper CT protocol and the angiographic phase in the evaluation of any patient with hematuria. In cases of suspected AVMs, selective renal arteriography can be both diagnostic and therapeutic in the absence of an apparent cause. In our patient, there was no clinical evidence indicating development of acquired fistula (iatrogenic or traumatic); taking into consideration the angiographic appearance and clinical presentation, the cavernous type of AV malformation was suspected.

The goal of treatment is maximal preservation of functioning renal parenchyma and the eradication of symptoms and their hemodynamic effects (8). Small peripheral AVMs without hemodynamic effects and minimal or no symptoms can be managed conservatively. Transcatheter arterial embolization has become the management of choice, replacing open surgery (3). However, surgical treatment remains a reasonable choice in unstable patients or in patients whose renovascular anatomy is not favorable for endovascular treatment, when there is large cirroid malformation, and in cases related to malignancy. There have been documented cases of renal AVM requiring urgent nephrectomy (9). Since our patient had massive hematuria, selective renal arteriography was carried out, and transcatheter supraseductive embolization of the lesion was performed. The risks of embolotherapy include reflux of obliterating agents into proximal vessels, resulting in loss of normal renal parenchyma and pulmonary embolism from migration of the agent. A postembolization syndrome may occur, characterized by pain in the embolized area, nausea, vomiting, and fever lasting up to 5 days. None of these complications were observed in our patient.

There have been some reports of spontaneous regression (10, 11). However, the possible mechanism remains unknown. There are several hypotheses. Hemorrhage and hematoma may promote thrombosis and associated vaso-

spasm, and edema can cause reduction of blood flow to the AVF. Increased bloodflow disturbance may cause subacute or chronic thrombosis, leading to regression of the AVF. Selective catheterization and angiography can damage arteriovenous malformation and result in the formation of thrombosis.

In our patient, hematuria disappeared after embolization. A Doppler study 6 months after embolization was normal. However, long-term followup is still needed, as the malformation may recur due to the rapid development of collateral vessels or recanalization of the abnormal vessel.

Teaching point

Arteriovenous malformation is uncommon and should be kept in mind in when evaluating a patient with hematuria. Color Doppler ultrasonography, multislice CT angiography, and digital subtraction angiography (DSA) are the most important tools for making the diagnosis in an urgent setting. In CT, the angiographic phase should be included in patients with history of hematuria. The therapeutic decision must be made by considering the general condition of the patient and symptoms. Embolization by selective catheterization can be considered safe and effective.

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