Case Reports

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Congenital renal arteriovenous malformation with cirsoid and cavernosal-type characteristics: a case report

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Abstract

Renal arteriovenous malformations (AVMs) are infrequent vascular morphological anomalies. About 20% of AVMs are congenital renal AVMs (CRAVMs). A 53-year-old female patient presented with a 5-day history of gross hematuria and right flank pain. The patient underwent the selective renal arteriography and embolization under local anesthesia. Renal computed tomography angiography (CTA) and digital subtraction angiography (DSA) results showed bleeding of the right renal arteriovenous malformation, both nidus and aneurysm, which indicated that the patient had both cirsoid and cavernosal types of CRAVM. Endovascular management was chosen to treat the patient. The patient was cured and discharged, then followed-up for 3 months. These results show that early identification using radiologic tests is important for diagnosis and treatment of CRAVM.

Keywords

Renal arteriovenous malformations, congenital type, malformed vascular bed, coil and liquid embolization, endovascular management, computed tomography angiography, digital subtraction angiography

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Introduction

Renal arteriovenous malformation (AVM) is a rare disease that is characterized by abnormal communications between renal arterial and venous systems. It has a prevalence of less than 0.04%.[1,2] The main clinical feature of AVM is sudden and uncontrollable hematuria, with or without a hack pair move. AVMs may be acquired, and the most common type is congenital renal AVM (CRAVM), which is present in approximately 20% of all renal AVM cases and in less than 1% of the general population.[3,4] CRAVM may cause hematuria leading to death. On the basis of vessel morphology, CRAVM has been classified into the following three categories:[2] the angiomatous type, which is characterized by a single large artery feeding multiple interconnecting distal branches and draining veins; the cirsoid type with multiple interconnecting varix-like malformed vascular bed-communications that lead to nonfunction of the focal glomeruli;[5] and the cavernosal type, which is caused by a preexisting arterial aneurysm that invades and erodes an adjacent vein.[6] In CRAVM, the renal arteriovenous communication does not have a normal arteriovenous capillary network, but instead, it relies on an abnormal vessel (nidus) network.

Case report

A 53-year-old female patient presented to an urgent care clinic with a 5-day history of gross hematuria, right flank pain, and dark blood clots in the urine. The pain was described as pressure-like pain without peritoneal irritation. The patient had no previous history of renal trauma, renal biopsy, renal surgery, or inflammation. The patient complained of dizziness. Physical examination revealed a body temperature of 36.8°C, blood pressure of 118/86 mmHg, pulse of 83 beats per minute, and respiratory rate of 19 breaths per minute. There were no other similar cases in the patient's family history. Cystoscopy was performed after admission, and the results showed a massive blood clot in the bladder (Figure 1).

After removing the clots, cytology indicated that the active bleeding originated from the right ureteral orifice. Renal computed tomography angiography (CTA) revealed bleeding from the right renal arteriovenous malformation, and digital subtraction angiography (DSA) was performed. The imaging results revealed both a nidus and an aneurysm, indicating that the patient had both cirsoid and cavernosal types of CRAVM (Figure 2, Figure 3). After evaluating the risks and benefits of

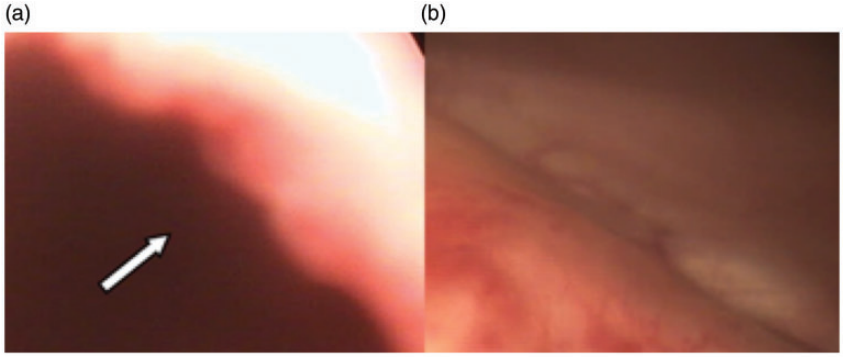

Figure 1. Cystoscope. a. A massive blood clot blocked the camera lens (white arrow); b. No cystorrhagia after suction.

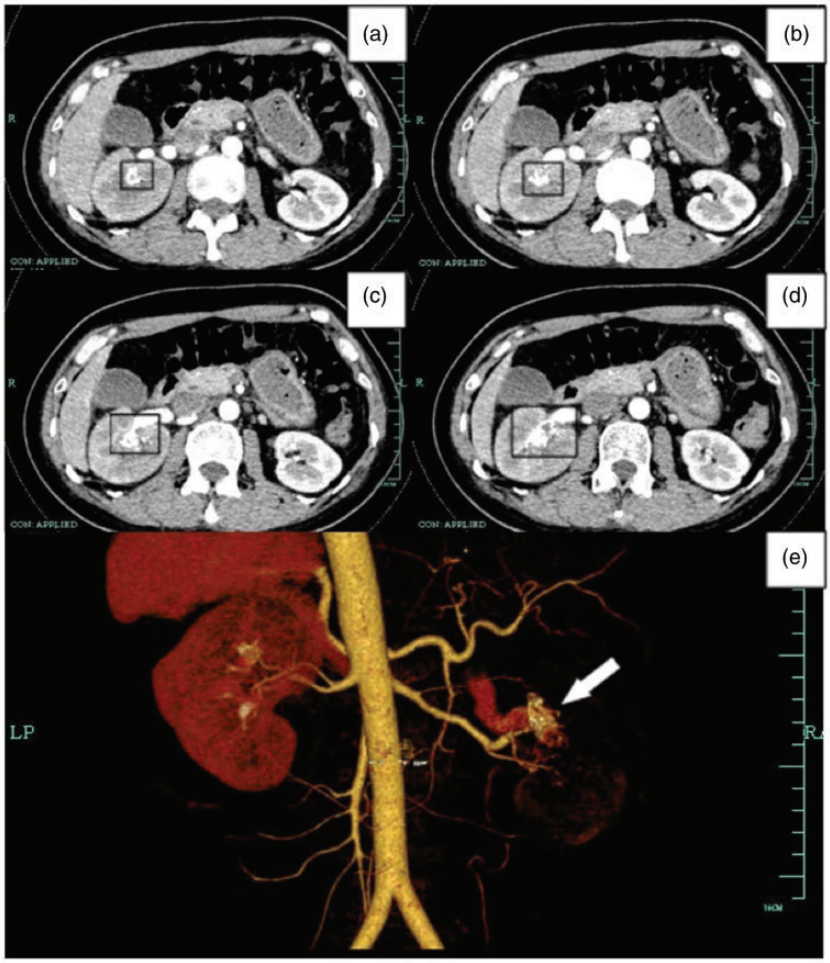

Figure 2. Computed tomography angiography. a: Cross-sectional CT shows high-density shadows (black quadrangles) with a diameter of 0.8 cm. b: A 1.1-cm diameter high-density shadow (black quadrangles) can be seen on the cross-sectional CT. c: A 1.4-cm diameter high-density shadow (black quadrangles) can be seen on the cross-sectional CT. d: The cross-sectional CT shows a 1.6-cm diameter high-density shadow (black quadrangles). e: A 3D image shows a lesion that is located in the right kidney, which is about $1.6 \times 1.5 \times 3.5$ cm (white arrow). Enlargement of the right renal feeding artery (black quadrangles), and the right upper pole arteriovenous malformation. 3D imaging showed an aneurysmal lesion in the right kidney (white arrow).
CT, computed tomography; 3D, three-dimensional.

more invasive surgery, endovascular management was chosen. Selective renal arteriography and embolization were performed under local anesthesia. Using the Seldinger technique, we punctured the right femoral artery and inserted a 5F vascular sheath. The renal angiography was performed using a 4F C2 catheter. The catheter was placed into the right renal artery. Angiography showed the right renal arteriovenous malformation with complex feeding arteries, and the contrast agent returned

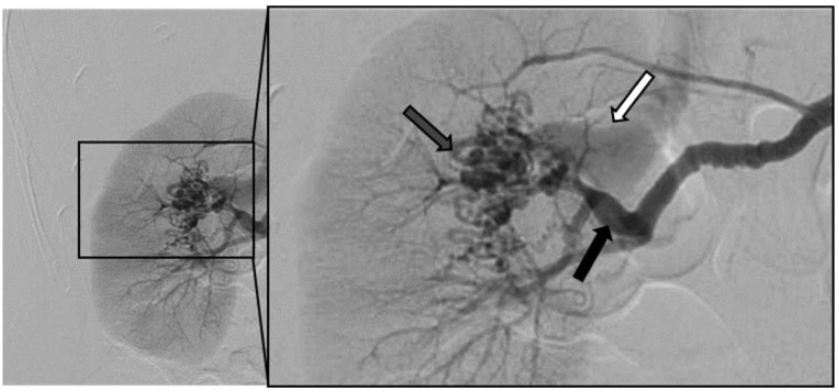

Figure 3. Renal angiography. Communication of the right upper pole feeding artery is a complicated, arterial feeder (black arrow) that communicates with a tortuous vascular group, which is highly suspected to be the varicosity type (gray arrow), and a venous aneurysm, which is highly suspected to be an angiomatous type (white arrow).

rapidly into the renal vein. A 1.98F ASAHI micro-catheter and a 2.4F Minreton micro-catheter were passed through and positioned into the targeted AVM feeding artery. Embolization was performed using Onyx-34, and its slow injection successfully embolized the nidus. Renal arteriography showed that the malformation disappeared after the management procedure, and the right renal superior pole artery was preserved. No symptoms were noted 7 days after the management procedure. The patient was cured and discharged and followed-up for 3 months.

Discussion

We reported the case of a 53-year-old female patient with symptoms of a renal arteriovenous malformation (AVM), such as the gross hematuria, right flank pain, and dark blood clots in the urine. Renal CTA and DSA showed bleeding from the right renal arteriovenous malformation, both nidus and aneurysm, which included both cirsoid and cavernosal types of CRAVM. The cirsoid type of CRAVM is characterized by morphology of many tiny twisted spiral vessels forming a network that abnormally connects arteries to veins in the lesion nidus (the core of the renal AVM). Most of the lesions were located below the mucosal lamina propria of the transitional epithelium in the renal collecting system. Venous valves become unstable because of a lack of elastic fibers in the malformed arteriovenous membrane, inaccurate communications between vessels, and focal hemodynamics. Furthermore, abnormal vessels rupture frequently and induce severe grass hematuria without pain.[7] Imaging of the cirsoid AVM showed no obvious tortuous feeding artery or drainage vein, but there was circular nidus staining in the nephrographic phase.[4] The cavernosal type of CRAVM mainly occurs in older people, and there is no nidus,[6] but there is a single artery and vein that are connected via a dilated chamber.[8]

Diagnosis of this disease mainly depends on imaging data. Therefore, DSA has become the gold standard, which accurately shows and determines the type of lesion. Multi-slice spiral enhanced CT combined with image post-processing has also been recently used to rapidly diagnose malformed vessels, and this depends on the correct scanning protocol and timing using a

collimation of at least 1 mm and a reconstruction interval of 0.5 mm to 1 mm (Figure 4). Cystoscopy is also helpful to identify the source of hematuria, and it is practical to use for a follow-up inspection. CRAVMs occur because of focal vascular development failures between embryonic weeks 4 and 10. At this stage, smooth muscle alpha-actin (αSMA) levels in vascular smooth muscle cells (VSMC) are low and incomplete, and the elastin coverage rate of the elastic protein layer (IEL) in AVM was also decreased, which are associated with the Notch2 mutation.[9] Most CRAVM patients remain asymptomatic until they are approximately 35 years old,[10] and most cases occur in older people, which might be relevant to the vascular walls that become fragile.

We chose endovascular management to treat this patient who had both cirsoid and cavernosal types of CRAVM, and the results were good. The patient recovered and was discharged. Surgical treatment for CRAVM including total or partial nephrectomy is only recommended for patients with hemodynamic instability or anatomic abnormality because the surgeries are invasive with a tendency to form a renal arteriovenous fistula.[11] Highly selective renal arterial embolization, especially multiple embolization technology, is the most efficient therapy for AVM, which embolizes the feeding arteries using spring (stainless steel or platinum) coils or an Amplatzer plug combined with gelfoam particles. Selection of the appropriate coil size is the key to a successful operation.

Recently, liquid ablation embolization has provided a new method for AVM treatment.[12] The liquid embolization agents include gelatin foam, ethyl alcohol, and tissue glue, and they are applied to the labyrinth to dehydrate and erode proteins inside vessels, damage the endothelial cell layer, and cause lesion necrosis, and to rapidly form a thrombus. Liquid embolization is very destructive, and the dose should be strictly controlled. In high-flow AVMs, polyvinyl alcohol foam particles (PVA) are used in advance to slow the flow velocity before liquid ablation. Viscous sclerosing liquid embolic agents (NBCA, Onyx, and Squid) perform better than alcohol. The angiomatous type AVMs are mainly treated with metal coils and movable balloons to enlarge the embolus and decrease the risk of coil migration. For pulmonary

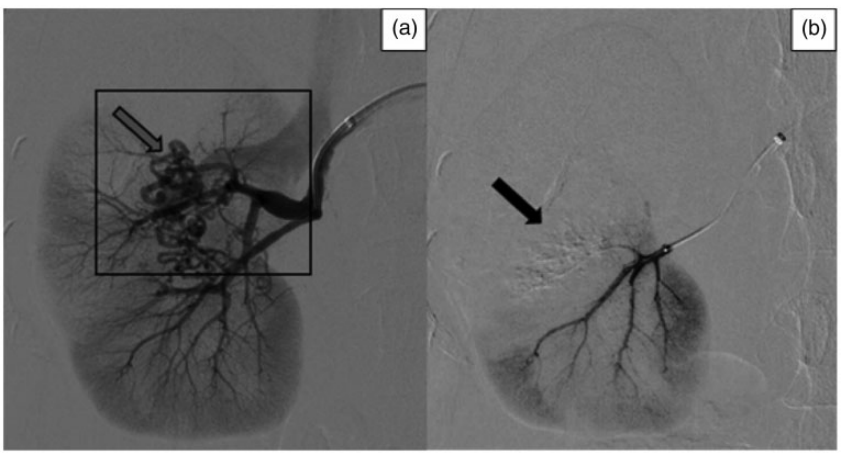

Figure 4. Renal angiography before and after management. a. the malformed lesion (black quadrangles) has disappeared; b. visible postembolization features (black arrow).

embolism, there can be hundreds of coils in some extremely high-flow cases. In one case, the renal vein was successfully blocked using balloon anti-reflux technology before liquid ablation.[13] The surgical achievement ratio would further be improved by combining the Interlock detachable coil (IDC) and the Guglielmi detachable coil (GDC) using a steel ring for embolization.

Conclusion

We report the case of a 53-year-old female patient with both cirsoid and cavernosal types of CRAVM, which was diagnosed using cystoscopy, renal computed tomography, CTA, and DSA. The patient's symptoms included the gross hematuria, right flank pain, and dark blood clots in the urine. Endovascular management was chosen to treat CRAVM in this patient, and she had a good outcome. Therefore, early identification by radiologic tests is important for diagnosis and treatment of CRAVM.

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Author contributions

Resources: Yuyong Wang.
Writing of the original manuscript: Jieru Cai.
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Review and editing: Li Ding, Yiwen Xie.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics approval and consent to participate

This study was approved by the Ethics Committee at the Affiliated Hangzhou First People's Hospital. Written consent was obtained from the study participant. This study is reported in accordance with the CARE guidelines.[14]

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