We report a patient with renal arteriovenous malformation (AVM), which was difficult to differentiate from renal pelvic tumor by retrograde pyelography (RP). A 44-year-old female visited our hospital due to sudden onset of gross hematuria. RP revealed irregular filling defect in the pelvocalyceal system, which could be an urothelial tumor and/or blood clot. Magnetic resonance imaging (MRI) showed several tiny flow voids adjacent to the renal pelvis with tangle of enhancing vessels in the mid pole of the right kidney and multiple fistulous connections with early filling of renal vein and IVC. Therefore, the diagnosis was renal AVM. The patient was treated with superselective transarterial embolization (TAE) with n-butyl 2-cyanoacrylate (NBCA). The symptom resolved after TAE. Imaging modalities and endovascular techniques for diagnosis and treatment of renal AVM were discussed.

Congenital renal AVMs are rare. Although arteriography still remain the gold standard, MRI may be useful as a non-invasive tool for the diagnosis of renal AVM.

CASE REPORT

A 44-year-old woman visited our hospital due to sudden onset of asymptomatic macroscopic hematuria. Physical examination was normal. Blood pressure was 116/75 mmHg. Pulse rate 66/min, Hematocrit was 24.6% and hemoglobin of 9.1g/dl. Urinalysis showed RBC numerous/HPF, WBC 8-10/HPF. There was no history of trauma, renal surgery or infection, whereas a similar attack with gross hematuria was once noted 10 months ago, which at that time resolved under medication and the cause of the hematuria was not disclosed.

Under the impression of renal neoplasm, she was admitted for further evaluation and management. Cystoscopy revealed a blood clot lodged in the right ureteral orifice and irregular filling defect within the pelvocalyceal system on RP (Fig. 1). This suggested either an urothelial tumor and/or a blood clot. MRI was performed for diagnosis and staging of urothelial carcinoma (1.5T, Magnetom Symphony, Siemens Medical Solutions). MR images showed flow voids in the upper right kidney adjacent to the renal pelvis on both T1- and T2-weighted fast spin-echo images (Fig. 2). Dynamic contrast-enhanced MR images and magnetic resonance angiography (MRA) with an IV injection of gadopentetate dime-
glumine (Magnevist, Schering) at a dose of 0.2 ml/kg of body weight were performed, which showed a nidus with tangle of enhancing vessels and multiple fistulous connections in the upper right kidney and early filling of renal vein as well as IVC (Fig. 3). The lesion located near the collecting system and characterized as cirsoid with multiple AV communication. Therefore, the lesion was diagnosed as congenital renal AVM [1].

Angiography was performed using an angiographic unit (Allura Medical system, Philips Corporation) confirmed the diagnosis of renal AVM. DSA demonstrated tortuous vascular channels with multiple fistulous connection between segmental arteries and veins on upper renal portion (Fig. 4). An immediate superselective endovascular embolization with n-butyl 2-cyanoacrylate (NBCA) (Histoacryl, Braun, Melsungen, Germany) mixed with lipiodol was performed to complete occlude the AVM.

During the ensuing 3 days after embolization the patient experienced mild right flank pain which resolved gradually. Hematuria was no longer present in more than 18 months of follow up.

**DISCUSSION**

Renal arteriovenous pathology can be labeled congenital, acquired or idiopathic. Congenital AVMs constitute 20% of the total AV communications, acquired lesions account for almost three-quarters, and idiopathic lesions account for 3-5% of all renal AV communications [2].

The lesion in our patient was categorized as congenital renal AVM, which consist of multiple tortuous communications between arteries and veins without interlaying capillaries. These connections through which the blood shunts, comprise the

**Figure 1.** Right retrograde pyelogram showed hematoma within the pelvocalyceal system as an irregular filling defect at the ureteropelvic junction region, mimicking urothelial carcinoma.

**Figure 2.** MR images of renal AVM. **a.** Axial fast spin-echo T1 weighted images (TR/TE: 553/7.5 ms) and **b.** Axial fast spin-echo T2 weighted images (TR/TE: 3678/102 ms) showed high signal intensity hematoma within the renal pelvis and tangled flow voids lesions adjacent to the renal pelvis, suggestive high flow vascular malformation.
so-called nidus of the AVM. These tortuous, varix-like vessels are immediately beneath the urothelium of the pelvic and calyceal wall, leading to hematuria as the presenting finding in as many as 72% of cases. Whereas hematuria is seen in less than 20% of those patients with idiopathic and acquired AVMs [2]. The difference in frequency of hematuria may be secondary to the submucosal location of the congenital type AVMs.

Renal AVM is relatively rare, with an estimated incidence of less than 0.04%. The true prevalence might be higher because many renal AVMs remain clinically asymptomatic [2]. Even after renal AVMs become symptomatic, they may last years before diagnosis is established.

Since renal AVMs are so uncommon and usually present with hematuria, the first imaging investigations are directed at exclusion of renal parenchymal or urinary tract tumor. Intravenous pyelography and RP can visualize urinary obstruction but has a low sensitivity and specificity in uncovering tumorous masses. As in our case, the images of RP mimics an urothelial carcinoma. MRI is useful for differentiation and characterizing renal lesions,
which was initially performed in our patient for diagnosis of urothelial carcinoma, while turned out to be a congenital renal AVM.

A diagnosis of congenital renal AVM should fulfill following criteria: no history of renal injury or disease, typical angiographic findings, and tortuous vascular channels between segmental or interlobar arteries and veins [3]. Arteriography is the gold standard for evaluating renal AVM [3], while it is an invasive procedure. We should search for a safe and noninvasive method. Ultrasound is the modalities of choice in excluding malignancy, but their role in detecting small renal AVMs remains unknown. Recently color Doppler sonography was proved useful in detecting abnormal high velocity flow in AVM [4]. Spiral computed tomography (CT) is also promising but is limited by the risks associated with iodinated contrast material and ionizing radiation [5]. Non-contrast enhanced MRI can demonstrate AVM with flow void vascular fistula on conventional images, thus MRI can be performed in patient with poor renal function. Donnelly et al. [6] reported that MRI is the primary imaging technique for the evaluation of suspected vascular malformations. Conventional MRI demonstrated high-flow vessels of high-flow vascular malformations as signal voids on spin-echo imaging and bright signal on gradient echo sequences [7]. In particular, the high accuracy of heavily T2-weighted images in defining the extent of vascular malformations has been described before and is already widely used in clinical practice, while literature on this subject remains scarce.

In addition to defining the extent of vascular malformations, dynamic contrast-enhanced MR images can provide information about the hemodynamics of vascular lesions [8]. Arterial phase contrast enhanced imaging will established the diagnosis of AVM by demonstrating abnormal vessels with an early draining vein, which is typical for arterial venous shunting. Angiography is the definitive procedure for determining the precise location and extent of renal AVM, as well as the number of renal arteries, feeding and draining vessels. Early opacification of the renal vein and inferior vena cava can also be demonstrated on angiography.

Renal AVMs provoking hematuria are usually located in the peripelvic region. Surgery of renal AVM need extensive branch artery dissection with high risk of renal parenchymal damage even resulted in nephrectomy [9]. Transcatheter embolization can replace open surgery, since it allows preservation of maximal unaffected normal renal parenchyma with relative low risk of complications and eliminating the risk of recurrent hemorrhage [10, 11].

CONCLUSION

Although there are few reports and experiences of MRI used for diagnosis of renal AVM, MRI is considered to be a non-invasive method for evaluating renal AVM before angiography.

Angiography remains the gold standard in establishing the diagnosis of renal AVMs. It demonstrates the detailed anatomy of the feeding and draining vessels, which facilitate accurate planning prior to intervention. Transcatheter superselective embolization is a well accepted method for treatment of renal AVM.

REFERENCES

腎動靜脈畸形之磁振造影影像：病例報告

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一位四十四歲女性患者，因突發性大量血尿到院求診，逆行性腎盂攝影檢查顯示不規則填充缺損在腎盂腎盂系統位置，疑似腎上皮腫瘤。進一步做磁振造影檢查顯示無信號病灶在右腎中間腎盂旁位置，且在注射完對比劑後呈現糾纏血管叢顯影增加現象，合併多處腎管及腎靜脈與下腔靜脈的早期顯影。由磁振造影檢查顯示，可以正確診斷為腎動靜脈畸形，病患隨即接受動脈栓塞手術治療。症狀也因此消失，影像檢查特性和栓塞治療技術也將被加以討論。先天性腎動靜脈畸形很罕見，目前仍以血管攝影檢查做為診斷標準。而磁振造影為一種非侵入性且準確性高的檢查。以此病例做磁振造影術檢查得以正確、快速診斷出腎動靜脈畸形，由此得以證明磁振造影在診斷病症的重要性。