

Bilateral congenital renal arteriovenous malformation: A rare entity with uncommon presentation.

Radhiana H^a, Mohd Shafie A^b and Mohd Ariff MA^c

^a Radiology Department, International Islamic University Malaysia ^b Radiology Department, Hospital Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia ^c Department of Diagnostic Imaging, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia.

ABSTRACT

Renal arteriovenous malformation (AVM) is a rare congenital anomaly of the urinary system. We present a patient with bilateral renal AVMs who presented with back pain and microscopic hematuria. This case highlights the importance of careful diagnostic work-up in the evaluation of upper tract hematuria.

KEY WORDS: Renal AVM, Transarterial Embolization, Hematuria.

INTRODUCTION

The term renal arteriovenous malformation (AVM) is usually reserved for congenital abnormal communications between the intrarenal arterial and venous systems. It usually presents with gross hematuria and hypertension. Back ache and microscopic hematuria as the presenting problems as illustrated in this case are less common. Embolization is currently the method of choice in treating this condition and proved to be effective in this patient.

CASE REPORT

A 45-year-old woman presented with mild back pain present for one year. Even though the pain was persistent, it did not affect her daily activities. Clinical examination was unremarkable. All blood investigations were normal. Urinalysis showed microscopic hematuria which was persistent on every follow-up. She was treated for pulmonary tuberculosis in the past, and there was no sign or symptom of reactivation. She was also a Hepatitis B carrier. There was no other significant medical history.

Ultrasound of the kidneys showed prominent vessels in the right kidney. No hydronephrosis or calculus was seen. Subsequent CT scan showed multiple rounded lesions in both kidneys with enhancement pattern suggesting abnormal vessels. The inferior vena cava showed early abnormal enhancement. Renal angiogram showed bilateral renal arteriovenous malformations in the lower pole of both kidneys. Coil emboliza-

tion was done at the same setting resulted in almost total occlusion of the AVMs in the right kidney (Figure 1a-c). On the left side, there was about 70% occlusion of the AVMs after the first embolization and total occlusion of the inferior interlobar arteries and the arteriovenous malformation after the second embolization. There was about 60% residual arterial supply to the upper and midpole of the left kidney (Figure 2a-c).

Subsequent follow up after two years revealed that patient was asymptomatic and urinalysis was normal.

DISCUSSION

Renal arteriovenous malformations (AVMs) or arteriovenous fistulas (AVFs) are abnormal communications between the intrarenal arterial and venous systems. The term renal arteriovenous malformation is usually reserved for congenital arteriovenous anomalies, characterized by multiple communications between the main or segmental renal arteries and veins. On the other hand, acquired arteriovenous anomalies, often termed arteriovenous fistulas are characterized by a single vastly dilated artery directly feeding one or more veins. Arteriovenous fistulas with the radiological appearance of an acquired fistula but without an identifiable cause are classified as idiopathic or cavernous AVMs.¹

The prevalence of any type of arteriovenous malformation or fistula is estimated to be less than 0.04%. This figure may exceed the true incidence in the general population as the study was a review of 9,500 arteriograms which were performed in patients with signs suggestive of renal pathology.² In an autopsy study, only 1 renal hemangioma and no arteriovenous malformations were found in more than 30,000 consecutive autopsies.³

Congenital renal AVMs usually present with hematuria, and gross hematuria as the primary complaint was reported in 72% of cases.⁴ Hematuria is thought

Corresponding author;

Radhiana Bt Hassan,
Radiology Department, IIUM,
Hospital Tengku Ampuan Afzan,
Jalan Tanah Putih, 25100 Kuantan,
Pahang Darul Makmur, Malaysia.
E-mail: radhianahassan@gmail.com

to result from minute rupture of the thin-walled veins into the collecting system from an increase in renal venous pressure. This is the reason why even small peripherally located AVMs can cause massive hematuria.⁵ Our patient had microscopic hematuria. There are few reported cases of AVMs in asymptomatic patients with microscopic hematuria.⁶ Hypertension was absent in our patient, but is present in about 50% of AVMs.⁴ Back pain or renal colic may result from obstructing blood clots but our patient never had clinically significant hematuria that may explain the cause of her back pain.

Several imaging modalities have been used to evaluate renal AVMs and AVFs. These include CT scan, Doppler Ultrasound, radionuclide renography and MRI.⁷ However, arteriography is still used as the gold standard to confirm this diagnosis. Moreover in cases of suspected AVMs, selective renal arteriography can be both diagnostic and therapeutic.⁵ Congenital AVMs show numerous tortuous vessels and interconnecting fistulas. Cirroid type is supplied by multiple arteries, usually segmental or interlobar arteries of normal caliber while the cavernous or idiopathic AVMs tend to be supplied by a single dilated vessel.⁶

The goal of treatment is maximal preservation of functioning renal parenchyma and the eradication of symptoms and its hemodynamic effects.¹ Small peripheral AVM without hemodynamic effects and minimal or no symptoms are managed conservatively. Transcatheter arterial embolization has become the management of choice replacing open surgery.⁵ Various agents available for embolization include coils, balloons, alcohol, autologous blood clots, gelfoam, absorbable gelatin sponge, cyanoacrylate and plastic polymers.^{5,8,9}

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 post embolization syndrome may occur, characterized by pain in the embolized area, nausea, vomiting and fever lasting up to 5 days.^{9,10} None of these complications were observed in our patient. Her back pain and microscopic hematuria disappeared after embolization. However, long term follow up is still needed as the malformation may recur due to the rapid development of collateral vessels or recanalization of the abnormal vessels.

CONCLUSION

Renal AVM was found to be the cause of mild back pain and persistent microscopic hematuria in this 45-year-old lady. This case highlights the importance of complete diagnostic work-up in the evaluation of microscopic hematuria in arriving at the correct diagnosis of an uncommon clinical entity.

REFERENCES:

1. Crotty KL, Orihuela E, Warren WM. Recent advances in the diagnosis and treatment of renal arteriovenous malformations and fistulas. *J Urol* 1993; 150:1355-9
2. Cho KJ, Stanley JC. Non-neoplastic and acquired renal arteriovenous malformations and fistulas. *Radiology* 1978; 129:333-43
3. Bell ET. A classification of renal tumors with observations on the frequency of the various types. *J Urol* 1938; 39:238-40
4. Takaha M, Matsumoto A, Ochi K, et al. Intra-renal arteriovenous malformation. *J Urol* 1980; 124: 315-7
5. Sountoulides P, Zachos I, Paschalidis K, et al. Massive hematuria due to a congenital renal arteriovenous malformation mimicking a renal pelvis tumour: A case report. *J Med Case Reports* 2008; 2:144-8
6. Wakefield MR. Renal arteriovenous malformation. In: e-medicine [online]. Available at: www.emedicine.com. Accessed February 2009.
7. Takebayashi S, Aida N, Matsui K. Arteriovenous malformations of the kidneys: Diagnosis and follow-up with color Doppler sonography in six patients. *Am J Roentgenol* 1991; 157:991-5
8. Tarif N, Mitwalli AH, Al Samayer SA, et al. Congenital renal arteriovenous malformation presenting as severe hypertension. *Nephrol Dial Transplant* 2002; 17:291-4
9. Takebayashi S, Hosaka M, Ishizuka E, et al. Arteriovenous malformations of the kidneys: Ablation with alcohol. *Am J Roentgenol* 1988; 150:587-90
10. Nakamura H, Uchida H, Kuroda C, et al. Renal arteriovenous malformations: Transcatheter embolization and follow-up. *Am J Roentgenol* 1981; 137:113-6

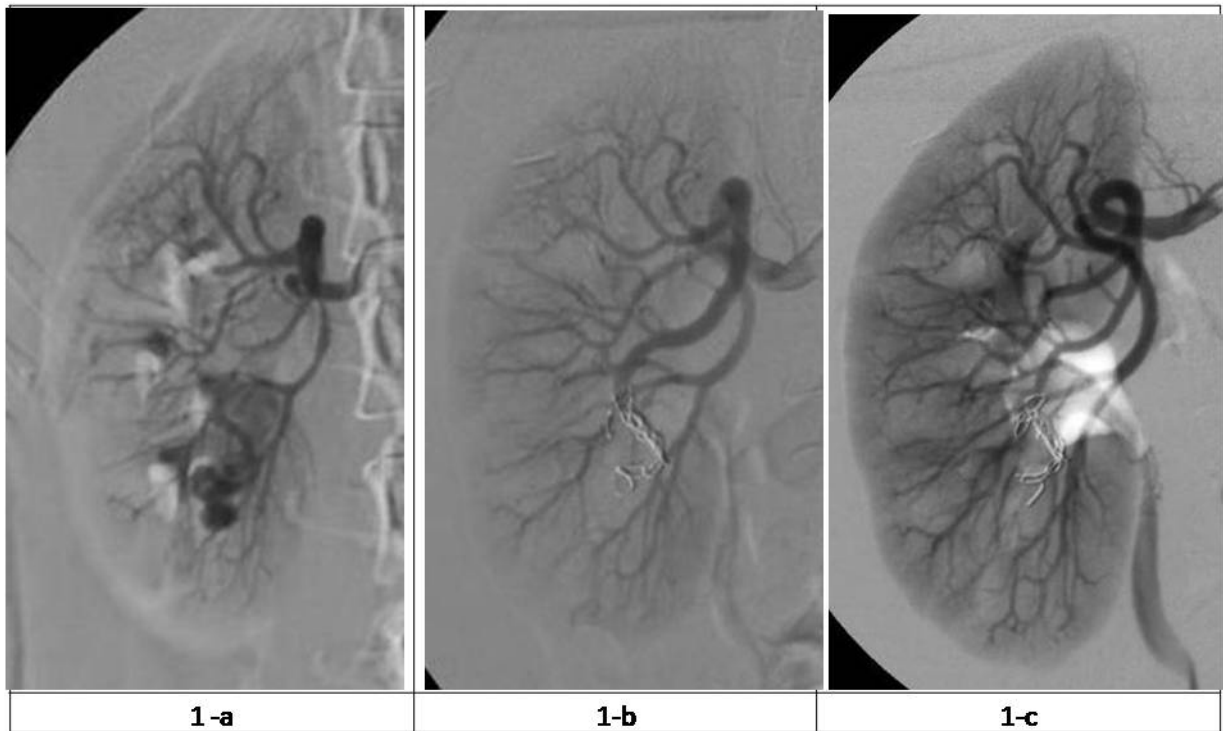


Figure 1 (a-c). Selective right renal arteriogram showing the numerous vascular channels in the lower pole (a). Post embolization showed total occlusion of the arteriovenous malformation (b) and repeat angiogram four months later showed no recurrence of the arteriovenous malformation (c).

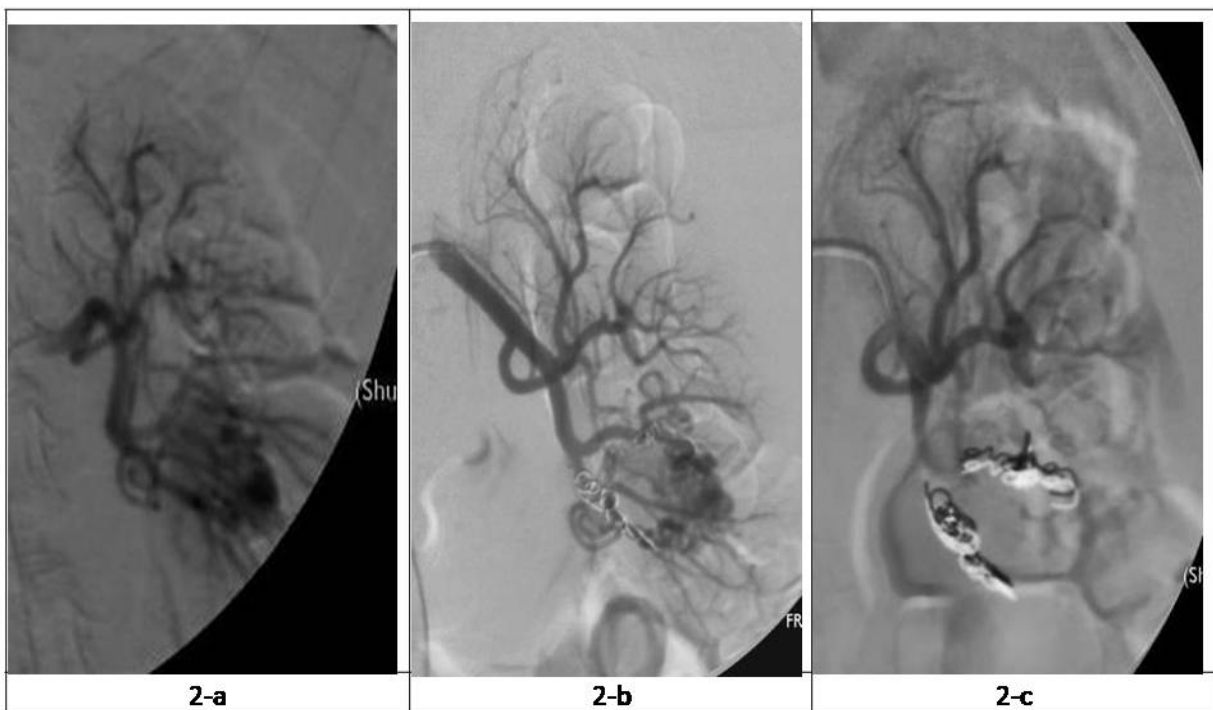


Figure 1 (a-c). Selective right renal arteriogram showing the numerous vascular channels in the lower pole (a). Post embolization showed total occlusion of the arteriovenous malformation (b) and repeat angiogram four months later showed no recurrence of the arteriovenous malformation (c).

