Spontaneous Renal Arteriovenous Malformation Presenting with Hematuria: A Case Report

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KEYWORDS
Renal arteriovenous malformation, Hematuria, Therapeutic embolization.

ABSTRACT
Introduction: Rare vascular abnormalities called renal arteriovenous malformations (AVMs) can cause hematuria among other symptoms. Due to overlapping clinical and radiological symptoms, differentiating between renal AVMs and other disorders such renal cell carcinoma (RCC) can be difficult. Here, we report a case of a patient who developed hematuria and was first diagnosed with renal AVM. We go over the difficulties in diagnosing this ailment and the approaches to managing it. Case Presentation: A 34-year-old female patient, reported having hematuria with clots for one week, coupled with fever and left loin discomfort that persisted for four and one day, respectively. Her hemoglobin level was 4.5 g/dL at arrival, which required packed red cell transfusions. A left double-J stent was implanted after an emergency cystoscopy revealed bloody efflux from the left ureteric orifice. A potential arteriovenous malformation in the left kidney's upper pole area was discovered by further imaging. A cirsoid-type AV abnormality arising from the superior polar branch of the renal artery was verified by renal angiography. After therapeutic embolization, the patient's symptoms were relieved and hematuria stopped. Discussion: The case study highlights the need of taking renal AVMs into account when making a differential diagnosis for hematuria, especially if there haven't been any previous urological operations. It also emphasises how important imaging techniques like renal angiography and renal ultrasonography with Doppler are for making the right diagnosis. Distinctive diagnosis from other ailments, such as renal cell carcinoma, highlights the necessity of comprehensive assessment and monitoring to guarantee suitable treatment. Conclusion: Renal arteriovenous malformations are rare but important causes of hematuria. Prompt diagnosis and management are crucial to prevent complications and improve patient outcomes. This case report demonstrates the diagnostic challenges associated with renal AVMs and underscores the importance of a multidisciplinary approach in their evaluation and treatment.

1. Introduction

An uncommon vascular abnormality known as renal arteriovenous malformation (AVM) is typified by aberrant communication between the kidney's venous and arterial systems. (1) Congenital occurrences affect fewer than 1% of the population, although acquired variants, especially arteriovenous fistulas (AVFs), are more prevalent. (1) (2) These malformations can lead to significant morbidity, with complications including recurrent hematuria, renal insufficiency, and hypertension. Prompt recognition and appropriate management of renal AVMs are essential to prevent long-term complications and preserve renal function (3).

Hematuria, the presence of blood in the urine, is a common presenting symptom of renal AVMs, often accompanied by clots and associated with varying degrees of pain. (4) The etiology of hematuria in these cases stems from the abnormal shunting of blood between arterial and venous systems, leading to the leakage of blood into the urinary collecting system. (5) The severity of hematuria can range from microscopic to gross, necessitating urgent medical attention in cases of significant bleeding. (5)

A variety of imaging modalities, such as contrast-enhanced computed tomography (CECT), renal ultrasonography (USG) with Doppler, and traditional renal angiography, are commonly used in the diagnostic assessment of renal AVMs. CECT provides detailed anatomical information and may reveal characteristic findings such as dilated tortuous vessels within the renal parenchyma. Doppler ultrasound aids in the detection of abnormal blood flow patterns, while renal angiography offers direct visualization of the vascular anatomy and allows for therapeutic interventions such as embolization. (6) (7)

Management strategies for renal AVMs aim to control symptoms, prevent complications, and preserve renal function. The choice of therapeutic strategy depends on the size, location, and complexity of the lesion as well as the patient's clinical presentation and comorbidities. Conservative therapy, endovascular embolisation, and surgical excision are the available alternatives. Endovascular embolisation has become a popular minimally

invasive and successful treatment option with low rates of recurrence and high rates of technical success. It uses a variety of agents, including glue, coils, and liquid embolics. (8) (9)

In this report, we describe a 34-year-old female patient who had a renal arteriovenous malformation and was exhibiting hematuria. A thorough description of the patient's clinical history, diagnostic workup, and management strategy will be provided, emphasising the difficulties in identifying and treating this uncommon vascular aberration. We hope that our case report will add to the body of knowledge already available on renal AVMs by highlighting the significance of prompt diagnosis and suitable treatment in obtaining positive results and enhancing patient care.

Understanding the clinical presentation, diagnostic evaluation, and management principles of renal AVMs is crucial for clinicians involved in the care of patients presenting with hematuria and associated symptoms. By elucidating the nuances of this complex vascular entity, we hope to enhance awareness, facilitate timely diagnosis, and improve the overall management of renal AVMs, ultimately leading to better patient outcomes and quality of life.

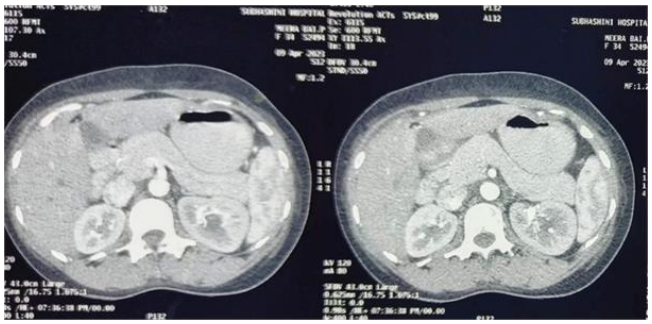
2. Case Presentation:

The case presentation involves a 34-year-old female who sought medical attention due to a one-week history of hematuria characterized by the presence of clots. Additionally, she reported experiencing fever for the past four days and left loin pain for one day. Notably, the patient had no prior history of urological interventions. Upon admission, her hemoglobin level was severely low at 4.5 g/dL, necessitating the administration of three units of packed red cell transfusions.

In response to the critical presentation, emergency cystoscopy was performed to evacuate clots and insert a left double-J stent (DJS). During the procedure, bloody efflux was observed from the left ureteric orifice, prompting the placement of the DJS to alleviate potential urinary obstruction.

Subsequent imaging studies, including CECT of the abdomen (Fig A), revealed a small 6x6 mm left upper calyceal blood clot and mild hydronephrosis (HUN) on the left side, with no intraluminal lesions detected. Urine cytology results returned negative for high-grade malignancy (Paris system Class II). Despite these findings, persistent symptoms and clinical suspicion led to the patient's referral to our institute for further evaluation and management.

Figure A – CECT Abdomen showing small blood clot in left upper calyx



Renal ultrasonography (USG) with Doppler (Fig B) revealed a suspected arteriovenous (AV) malformation in the upper pole region of the left kidney. To confirm the diagnosis, diagnostic retrograde intrarenal surgery (RIRS) was performed, identifying a pulsatile region within the upper pole calyx parenchyma, without the presence of an intraluminal mass.

A cirsoid-type AV malformation arising from the superior polar branch of the renal artery was definitively visualized using conventional renal angiography (Fig C), which was carried out under local anesthesia. Subsequently, therapeutic embolization was carried out using 4 ml of 17% n-butyl cyanoacrylate (NBCA) glue (Fig D), leading to complete occlusion of the malformation. Post-embolization imaging demonstrated no uptake in the lesion (Fig E), and the patient's hematuria resolved following the procedure.

Figure B – USG Doppler revealed Whirlpool Sign in upper calyx

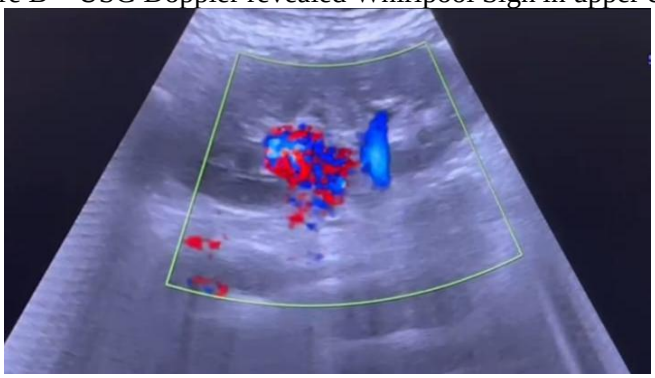


Figure C – Conventional Angiogram revealed AV malformation in upper pole and hilar region

Figure D – Injection of NCBA glue

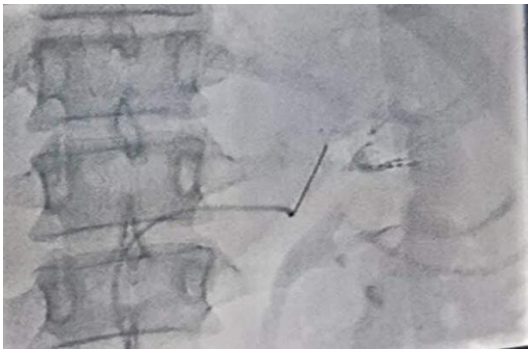
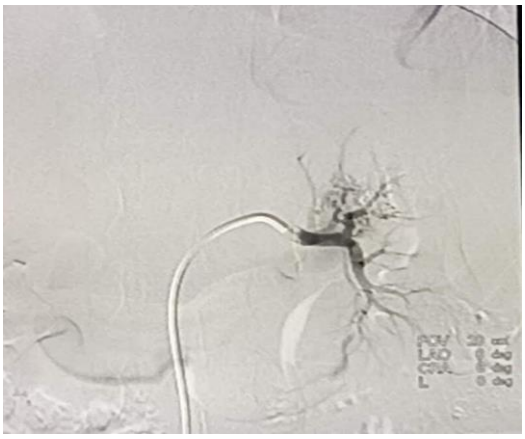


Figure E – Post embolization shows resolution of the AV malformation



During a six-month follow-up period, the patient remained asymptomatic, with no recurrence of hematuria or associated symptoms reported. Regular surveillance will be continued to monitor for potential complications or

recurrence of the AV malformation, ensuring the patient's ongoing well-being and optimal management of the condition

3. Discussion

The presented case of renal arteriovenous malformation (AVM) underscores several key clinical and management considerations. Renal AVMs are rare vascular anomalies characterized by abnormal communications between the arterial and venous systems within the kidney. They can manifest with a wide spectrum of symptoms, ranging from asymptomatic incidental findings to life-threatening hemorrhage. In this discussion, we will delve into the pathophysiology, diagnostic approach, therapeutic options, and prognostic implications of renal AVMs, as demonstrated by the case presented.

The pathogenesis of renal AVMs is multifactorial, with both congenital and acquired etiologies contributing to their development. Congenital AVMs arise from abnormal vascular development during embryogenesis, leading to aberrant connections between arterial and venous structures within the kidney. Acquired AVMs, on the other hand, can result from trauma, inflammation, or iatrogenic injury, such as prior renal biopsy or surgery.

The hallmark feature of renal AVMs is the presence of arteriovenous shunting, wherein arterial blood bypasses the normal capillary bed and directly enters the venous circulation. This abnormal flow pattern can lead to arterialization of the venous system, venous hypertension, and subsequent rupture or hemorrhage.

Imaging investigations and clinical evaluations are used to diagnose renal AVMs. Although some instances may be asymptomatic, patients usually appear with symptoms such hematuria, flank discomfort, or hypertension. Imaging modalities are essential for verifying the diagnosis and defining the lesion's vascular architecture..

A tiny left upper calyceal blood clot and moderate hydronephrosis were seen during the first assessment of the patient using contrast-enhanced computed tomography (CECT) abdomen, which raised concerns about an underlying vascular problem. Additional data from renal ultrasonography (USG) with Doppler suggested the presence of an arteriovenous malformation in the left kidney's upper pole area. Through diagnostic retrograde intrarenal surgery (RIRS) and traditional renal angiography, the cirsoid-type AV malformation arising from the superior polar branch of the renal artery was visualised, providing confirmation of the diagnosis.

The management of renal AVMs aims to control symptoms, prevent complications, and preserve renal function. Therapeutic options include conservative management, endovascular embolization, and surgical resection. The choice of approach depends on various factors, including the size, location, and complexity of the lesion, as well as the patient's clinical presentation and comorbidities.

In the case presented, therapeutic embolization was performed using 17% n-butyl cyanoacrylate (NBCA) glue, resulting in complete occlusion of the AV malformation. Embolization serves as a minimally invasive and effective treatment modality, offering high rates of technical success and low rates of recurrence. By occluding the abnormal arteriovenous shunts, embolization effectively interrupts the aberrant blood flow and reduces the risk of hemorrhage.

The prognosis of renal AVMs following treatment is generally favorable, with resolution of symptoms and preservation of renal function observed in the majority of cases. Post-embolization imaging demonstrated no uptake in the lesion, and the patient's complaints of hematuria ceased. Additionally, the patient remained asymptomatic during six months of follow-up, highlighting the efficacy of therapeutic embolization in achieving long-term symptom relief and preventing recurrence.

Renal arteriovenous malformations (AVMs) pose diagnostic challenges due to overlapping clinical presentations with other conditions, including renal cell carcinoma (RCC) and urinary tract infections (UTIs). Several case reports and studies shed light on the diverse manifestations and management strategies associated with renal AVMsThe study "Gross hematuria: Renal cell carcinoma mimicking a renal arteriovenous malformation describes a case in which an 80-year-old man was diagnosed with renal cell carcinoma (RCC) after first receiving an arteriovenous malformation (AVM) diagnosis. This underscores the difficulty in differentiating between the two conditions and highlights the significance of short-term follow-up to confirm diagnosis or identify any changes that may indicate an underlying tumour". (10)

"A Case of Right Renal Arteriovenous Malformation Mimicking the Symptoms of Urinary Tract Infection in the Emergency Department: This study describes a situation in which a patient who initially had symptoms similar to a urinary tract infection was ultimately found to have renal AVM. This highlights the need for

emergency physicians to be aware that renal AVM can present with symptoms similar to UTI, particularly in young female patients". (11)

"Recurrent hematuria in renal angio-venous malformation, delay diagnosis and endovascular treatment, a case report: The difficulties in identifying and treating congenital renal AVMs are covered in this paper, with a focus on the changing approaches to management—from nephrectomy to catheter embolization—and the critical role renal vascular angiography plays in achieving an accurate diagnosis". (12)

"Congenital renal arteriovenous malformation: a rare cause of visible haematuria: This paper highlights the need of early detection and intervention to prevent serious problems associated with renal AVMs by reporting a case of renal AVM, describing its angioarchitecture, and outlining endovascular therapy". (13)

"Renal arteriovenous malformation mimicking hydronephrosis—hidden danger: This study highlights the significance of colour flow by presenting a case where renal AVM was first misdiagnosed as hydronephrosis. Doppler ultrasonography is used in precise diagnosis to prevent misdiagnosis and potentially dangerous outcomes". (14)

"Congenital Renal Arteriovenous Malformation: Diagnostic Clues and Methods: This paper explores diagnostic hints and suggests an algorithm for improved renal AVM detection, highlighting the significance of taking AVMs into consideration in patients whose hematuria is not found by standard workup, especially in mid-aged or pregnant patients". (15)

"Congenital renal arteriovenous malformation with cirsoid and cavernosal-type characteristics: This paper highlights the significance of early detection through radiologic testing for timely treatment and successful outcomes. It also presents a case of congenital renal AVM with distinctive morphological characteristics". (16)

To summarise, the examination of several case studies and reports highlights the range of clinical manifestations, difficulties in diagnosis, and available treatments related to renal AVMs. Increased awareness among medical professionals, especially emergency room doctors, along with the prudent application of sophisticated imaging modalities are necessary for precise diagnosis and prompt initiation of suitable treatment, which will ultimately improve patient outcomes and lower the risk of complications from these uncommon vascular anomalies.

4. Conclusion

In conclusion, renal arteriovenous malformations represent rare but clinically significant vascular anomalies that can present with hematuria and associated symptoms. A comprehensive diagnostic approach involving imaging studies and invasive procedures is essential for accurate diagnosis and treatment planning. Therapeutic embolization emerges as a safe and effective treatment modality, offering high rates of success and low rates of recurrence. Through prompt recognition and appropriate management, favorable outcomes can be achieved, leading to symptom resolution and preservation of renal function. This case report contributes to the existing literature on renal AVMs, emphasizing the importance of early intervention and multidisciplinary collaboration in optimizing patient care.

Acknowledgment:

We acknowledge the contributions of all healthcare professionals involved in the diagnosis and management of the patient described in this case report.

Conflict of Interest:

The authors declare no conflicts of interest related to this study.

Consent Declaration:

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

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