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Unmasking the Enigma: A Case Series on Idiopathic Renal Arteriovenous Malformations

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Abstract

Renal arteriovenous malformations (rAVMs) are rare vascular abnormalities with a global prevalence of only 0.04%. Characterized by dilated, tortuous vessels formed by abnormal communications between renal arteries and veins, these malformations lack an intervening capillary network, leading to the shunting of high-pressure arterial blood into low-pressure veins. Although typically affecting individuals aged 30 - 40, our case series from East Malaysia reveals a notable prevalence among older patients, predominantly females. rAVMs can be classified into congenital, idiopathic, and acquired types, with acquired forms accounting for 70% - 80% of cases. Diagnosing rAVMs can be challenging due to their varied clinical presentations; patients may be asymptomatic or present with hematuria, flank pain, or even life-threatening hemorrhage. Imaging techniques such as Doppler ultrasound, Computer topography, and magnetic resonance imaging are essential for visualization and assessment. Management strategies range from conservative observation in asymptomatic cases to endovascular embolization or surgical resection for symptomatic patients. This case series highlights the clinical presentations, demographic factors influencing rAVM prevalence, and management approaches, aiming to enhance understanding and improve treatment protocols for these complex vascular anomalies. Renal arteriovenous malformations (rAVMs) are infrequent vascular anomalies predominantly occurring in the left kidney, often involving the upper pole and typically affecting the collecting system rather than renal parenchyma. In this case series, all patients were female, supporting the known gender predisposition, as rAVMs affect females twice as often as males. Contrary to the expected peak incidence in individuals aged 30 - 40, most patients presented with symptoms in their sixth and seventh decades of life. rAVMs can be classified into congenital and acquired types, with acquired

forms accounting for 70% - 80% of cases, frequently linked to iatrogenic injuries. Congenital rAVMs result from developmental failures and remain asymptomatic until adulthood. This series highlights a notable prevalence of rAVMs in East Malaysia, particularly in Sabah. The underlying mechanisms of rAVM formation are not fully understood, though genetic factors, such as those associated with Hereditary Hemorrhagic Telangiectasia (HHT), may contribute. Patients typically present with hematuria and flank pain, necessitating a thorough diagnostic approach to differentiate from other urological conditions. Emergency management includes transcatheter renal artery embolization (RAE), favored for its minimally invasive nature, while surgical options are reserved for severe cases. This study underscores the importance of early diagnosis and a multidisciplinary approach in managing rAVMs, especially in older females presenting with hematuria, to prevent life-threatening complications.

Subject Areas

Urology

Keywords

Renal Arteriovenous Malformations, Idiopathic rAVMs, Selective RAE

1. Introduction

Renal arteriovenous malformations (rAVMs) are rare, occurring abnormality of the renal vasculature. On a global scale, its prevalence is only 0.04% [1]. Renal AVMs are composed of dilated, enlarged, tortuous vessels which are formed by abnormal communications between renal arteries and veins forming a complex nidus. These connections are devoid of an intervening capillary network, resulting in shunting flow of high pressure arterial blood to low pressure venous blood. Renal AVMs can be categorized into congenital, idiopathic, and acquired types. Of the three, acquired renal AVMS are found to be the most commonly occurring, making up about 70% - 80% of all renal AVMs [2]. On the contrary, idiopathic renal AVMs are the least common and are typically discovered incidentally during imaging, with their etiology remaining unclear. Based on previous reports, individuals in the middle age group are found to be more prone to develop renal AVMS. Furthermore, studies have shown a favourable female gender predisposition [3]. Various theories have been proposed regarding their development. Renal AVMs can pose a diagnostic challenge, due to its diverse clinical presentation. Some patients are completely asymptomatic upon diagnosis. However, others may exhibit worrying symptoms such as hematuria, flank pain, or a palpable mass. Severe cases may present with renal failure or life-threatening torrential hemorrhage. Diagnosis often involves a combination of imaging techniques including Doppler ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) to visualize the intricate vascular structures and determine the extent

of the malformation. This case series illustrates an overview of a range of cases that focuses on the clinical presentations, racial and demographic factors that could influence the prevalence of Renal AVMs in East Malaysia, as well as their diagnostic approaches, and management strategies.

2. Methodology

This retrospective case series was conducted between January 2020 and December 2024 at Queen Elizabeth Hospital, a tertiary referral centre in Kota Kinabalu, Sabah, East Malaysia. The case series comprised 3 cases of incidental renal arteriovenous malformations (rAVMs) managed with endovascular embolization. Inclusion criteria comprised adult patients with confirmed rAVM on CTA (computer tomographic Angiography) or digital subtraction angiography (DSA) regardless of variation of symptoms.

The cases were compared on several factors including, age, gender, race, co morbids, and clinical presentation. Table 1 depicts the demographic distribution and comparison of clinical presentations between the 3 cases.

Table 1. Patient demographic distribution and clinical symptoms.

Case	Age/sex	Race	Co-morbids	Symptoms
1	55/F	Dusun	nil	Flank pain
2	72/F	Dusun	nil	Visible Hematuria
3	48/F	Dusun	nil	VIsible Hematuria

Cases with incomplete imaging data or non-renal AVMs were excluded. Clinical data, radiological findings, treatment modalities, and outcomes—including renal function before and after embolization—were collected and tabulated to establish comparisons and similarities between the cases. The findings have been illustrated systematically as shown in Table 2.

Table 2. Summary of procedural elements and embolization outcomes.

Case	Site of nidus	Size of nidus (cm)	Type of nidus (Yakes classification)	Embolic agent	Complications	Follow up (post embolization)
1	Left lower pole	1.5 × 1.4	IIA	Histoacryl Glue	Nil	CT renal angiogram in 3 months
2	Right midpole	1.2 × 1.7	IIA	Histoacryl Glue, Polyvinyl Alcohol (PVA), Coils	Rebleeding	Renal angiogram in 3 months and CT renal angiogram in 6 months
3	Right midpole	2.0 × 1.8	IIA	Coils	Renal infarction	CT renal angiogram in 3 months

Post embolization, all 3 patients had regular renal function blood tests to monitor renal function. In all 3 patients, serum creatinine and EGFR remained stable post procedure. Table 3 depicts the renal profiles of the 3 cases during the pre and post embolization period.

Table 3. Blood parameter trends and evaluation of pre and post embolization renal function.

Case	Pre embolization platelet (10^3/UL)	Pre embolization coagulation profile	Pre embolization renal profile		Post embolization renal profile	
			Urea (mmol/L)	Creatinine (umol/L)	Urea (mmol/L)	Creatinine (umol/L)
1	261	PT: 10.7s APTT: 27.1s INR: 1.04	3.0	69	3.9	67
2	193	PT: 10.2 APTT: 22.9 INR: 0.99	8.9	113.5	8.0	102.2
3	532	PT: 10.8 APTT: 22.8 INR: 1.05	2.6	79	4.0	78

For each case, the Renal AVM nidus identified was classified in reference to Yakes classification of Renal AVMs as shown in Table 4.

Table 4. Yakes classification for renal AVM.

Nidus Types		Angioarchitectures Description	Treatment Approach
Type I		Direct arteriovenous fistula (AVF).	Mechanical occlusion devices (metallic coils, vascular plugs/balloon).
Type II	IIa	Typical AVM nidus.	Transcatheter and direct puncture embolization with ethanol.
	IIb	AVM nidus that drains into dilated/aneurysmal vein.	Similar with Type IIa + coiling of the aneurysmal outflow vein.
Type III	IIIa	Aneurysmal vein with nidus located at the vein wall and has a single outflow vein.	Coiling the single outflow vein.
	IIIb	Type IIIa + multiple outflow veins.	Coiling all the outflow veins.
Type IV		AVM with tissue infiltration.	Transcatheter embolization and direct puncture embolization (50% ethanol and non-iconic contrast mixture).

3. Case Presentation

3.1. Case 1

We report of a 55-year-old female who was referred for hematuria. She claimed

to have been having painful hematuria for 2 months. There were associated symptoms of left flank pain. There were no constitutional symptoms, no history of trauma or renal surgeries previously. The patients physical examination was also unremarkable. A flexible cystoscopy was done which revealed oozing of blood from the left ureteric orifice. However, there was no surrounding lesions visualized, and the overall bladder appeared healthy. A CTA demonstrated a nidus of an arteriovenous malformation at the inferior calyx of the lower pole of the left kidney (Figures 1-3). Her pre-embolization blood investigations were found to be within normal range. Subsequently, super-selective angioembolization of the left lower pole segmental artery AVM was performed successfully resulting in complete obliteration of the nidus (Figures 4-7). Histoacryl glue was used as the embolic material. A Post-embolization CT multiphase showed presence of the embolic material, with no visible nidus (Figure 8). The patient's renal function was monitored via serum creatinine and estimated glomerular filtration rate (eGFR) over a 6-month follow-up period, with no evidence of renal impairment.

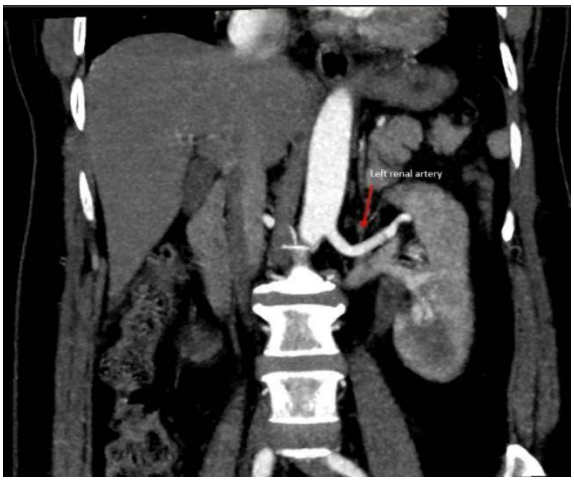

Figure 1. Pre embolization CT renal multiphase.

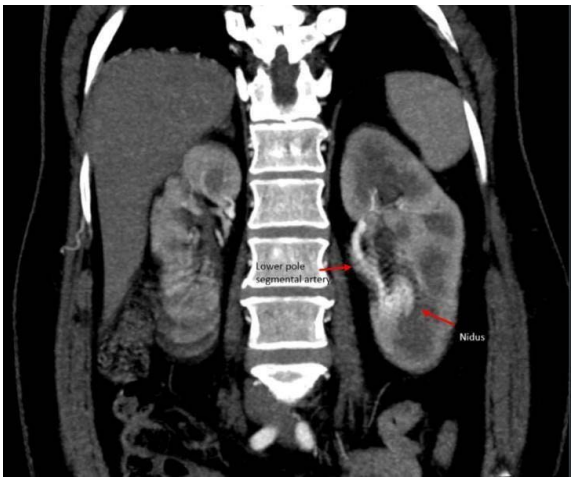

Figure 2. Pre embolization CT renal multiphase. Showing presence of left renal lower pole AVM.

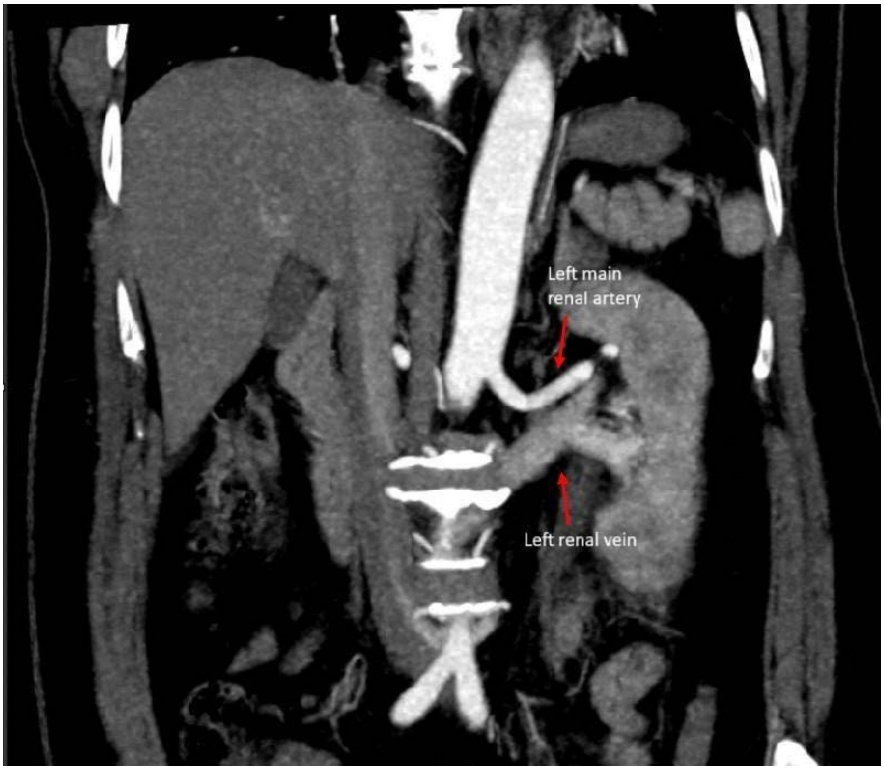

Figure 3. Pre-embolization CT renal multiphase. Opacification of the left renal vein in arterial phase in keeping with arteriovenous shunting.

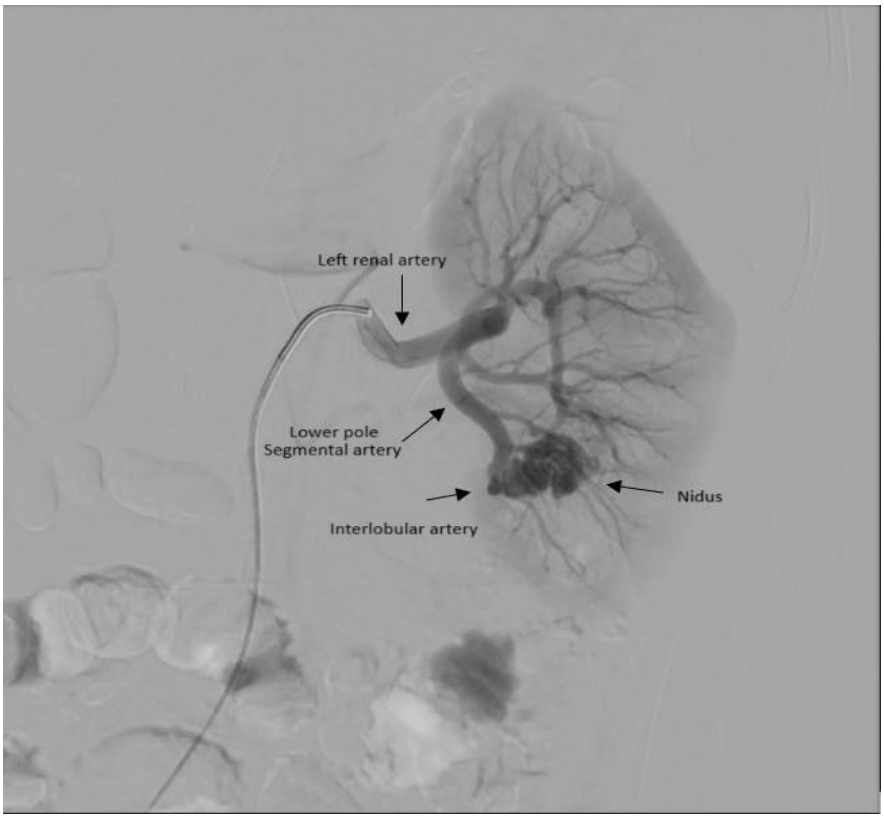

Figure 4. Pre embolization angiogram.

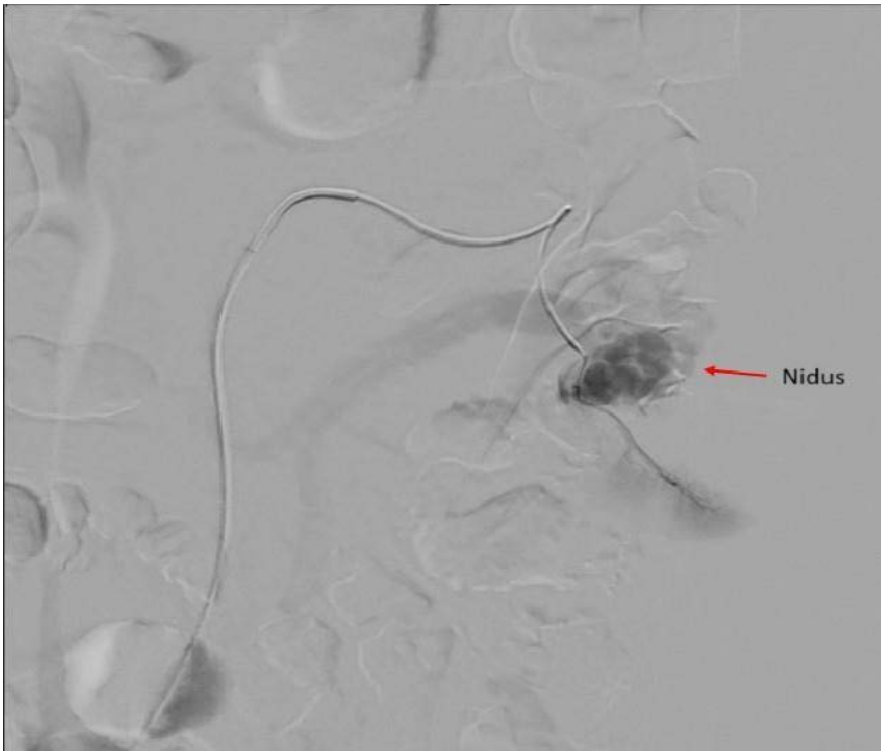

Figure 5. Pre embolization angiogram.

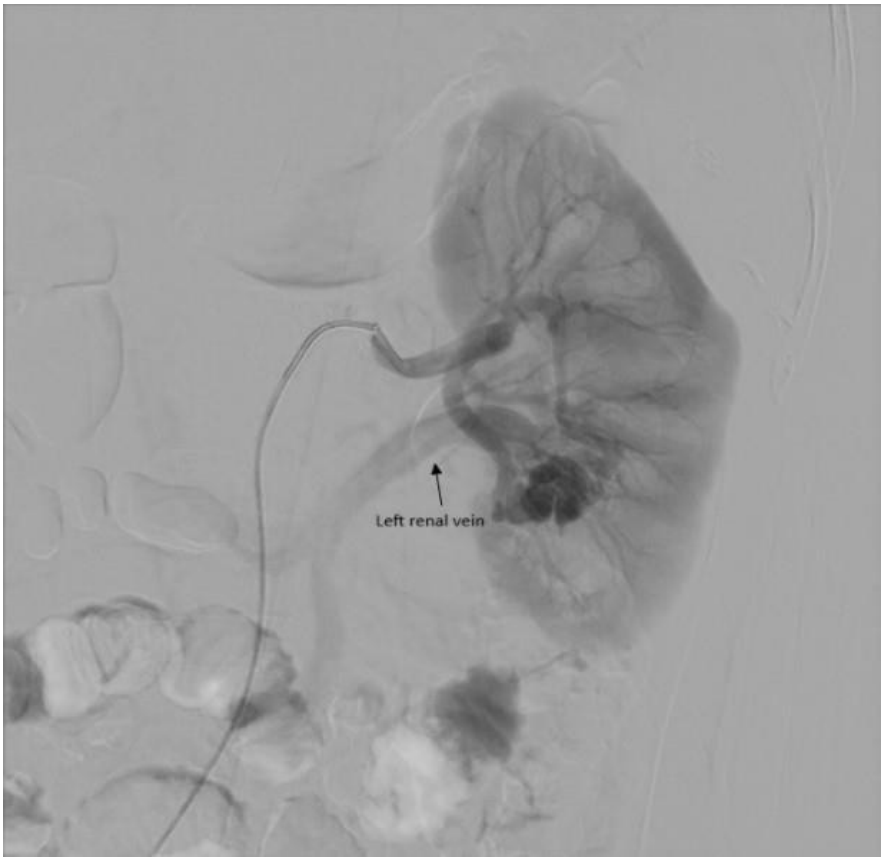

Figure 6. Pre embolization angiogram.

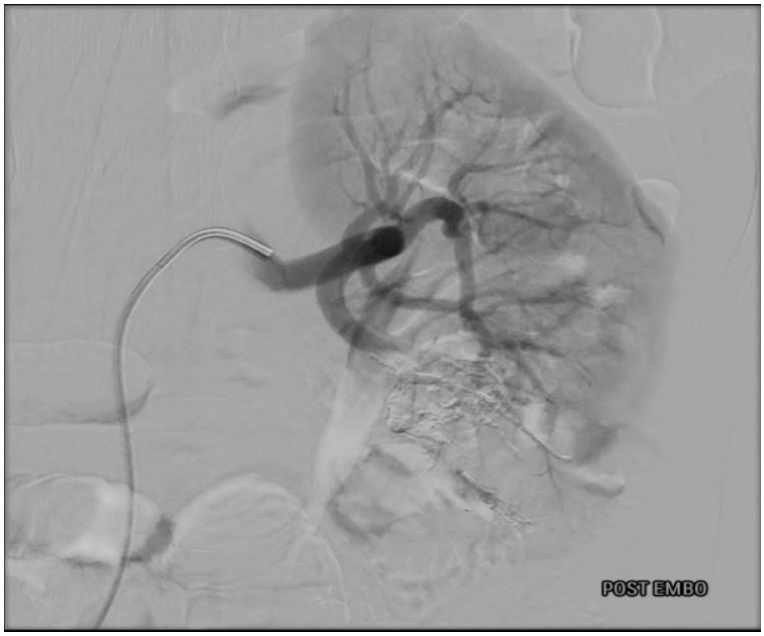

Figure 7. Post embolization of super selective of the left lower pole segmental artery. It shows complete obliteration of the nidus.

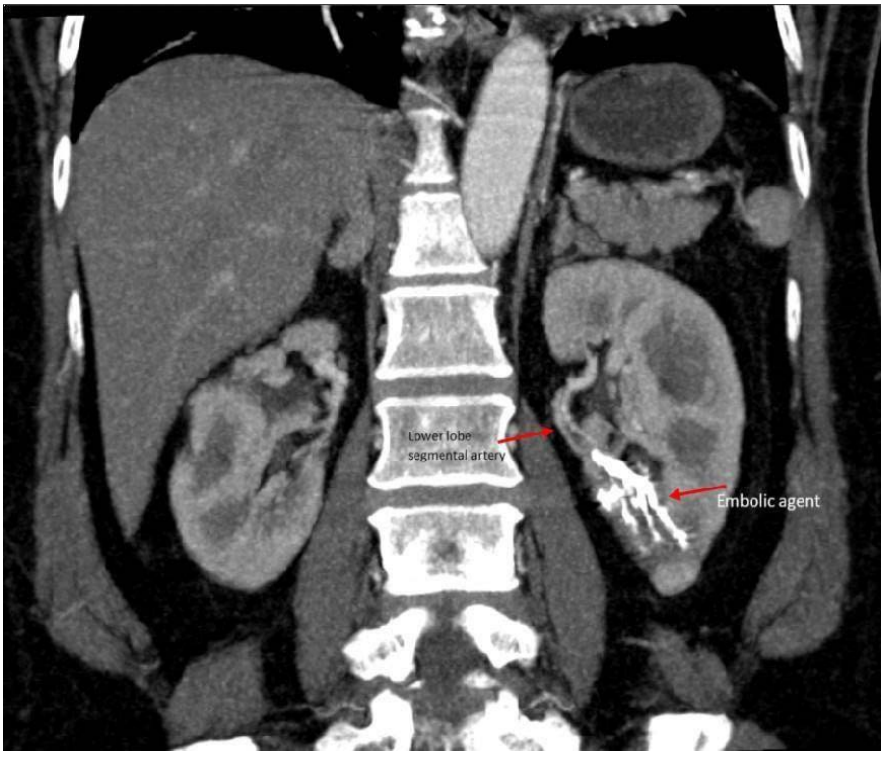

Figure 8. 2 months post embolization CT renal multiphase shows hyperdense embolic material seen at the left lower pole segmental and interlobar renal artery. No obvious nidus seen in this region.

3.2. Case 2

We report a 72-year-old female, a nonsmoker, who presented with intermittent

painless hematuria. She initially underwent a flexible cystoscopy, which revealed large amounts of clots from the right ureteric orifice. An ultrasound scan showed that the Right kidney had sediments within pelvicalyceal system, with features of pyelonephritis, right mild hydronephrosis. On Plain CT imaging, there were hyperdensities observed within renal pelvis and ureter in keeping with blood product as well as the presence of hydronephrosis and hydroureter (Figure 9). The arterial phase of the CT imaging demonstrated the presence of arteriovascular malformation nidus within the midpole of the right kidney with its supply arising from the segmental branches of the renal artery. She underwent a superselective angioembolisation using Nester coil ×10 (Figure 10). However, 2 weeks later she presented again with complaints of hematuria. A repeat CTA showed persistence of the right midpole segmental renal artery AVM nidus with presence of contrast extravasation suggestive of active rebleeding of right renal artery AVM (Figure 11). A pre embolization angiogram demonstrated contrast extravasation around the nidus seen at the midpole further confirming the diagnosis of bleeding AVM (Figure 12). Based on this, she was then subjected to a second angioembolization using histoacryl glue as the embolic agent instead. Renal function tests remained stable post-procedure, and no long-term impairment was detected on subsequent follow-up.

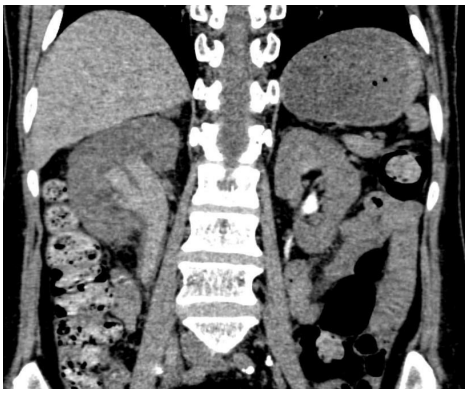

Figure 9. CT renal multiphase (plain). Hyperdensities within renal pelvis and ureter in keeping with blood product. Presence of hydronephrosis and hydroureter.

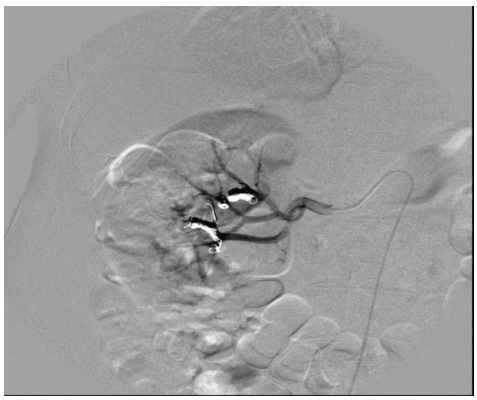

Figure 10. Post coiling, small residual right renal interpole AVM.

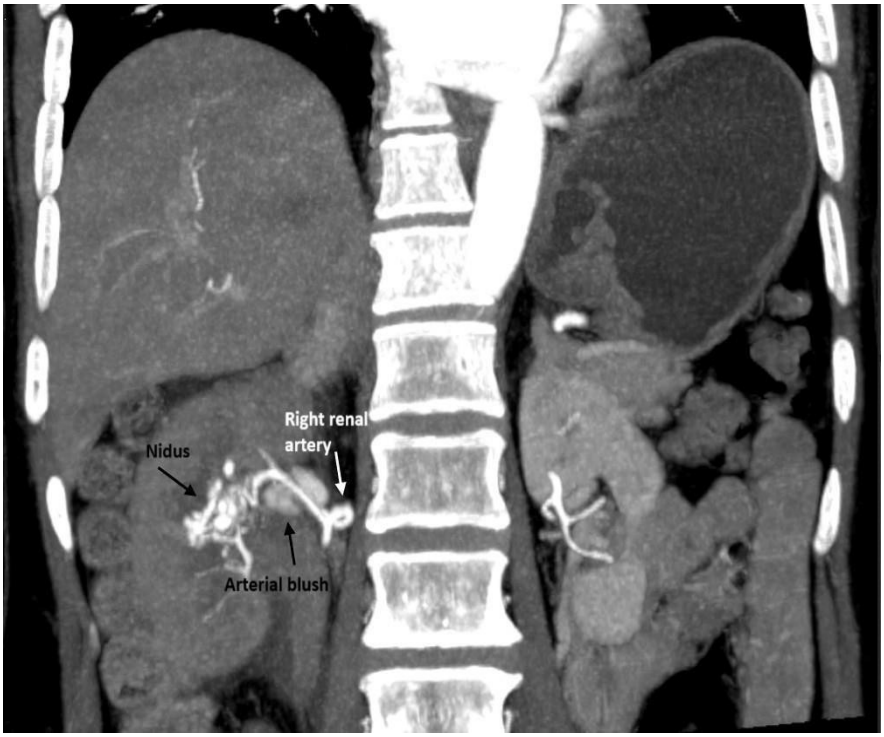

Figure 11. AVM nidus seen within the midpole of right kidney with its supply arising from the segmental branches of the renal artery. Presence of arterial blush in keeping with active bleed.

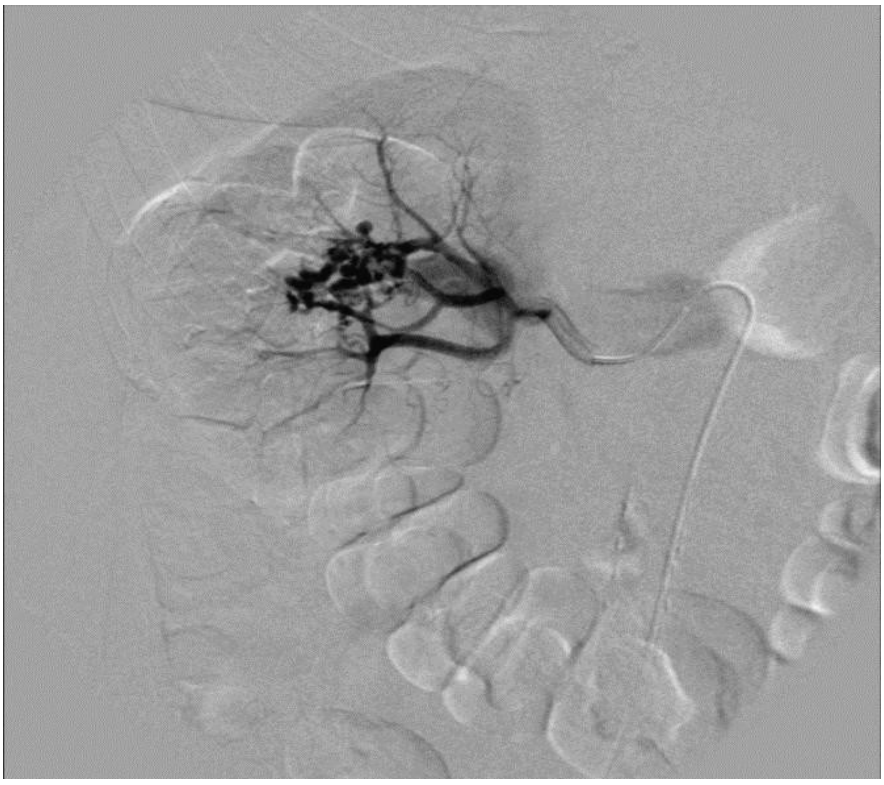

Figure 12. Pre embolization angiogram, AVM nidus seen at the midpole with contrast extravasation in keeping with bleeding AVM.

3.3. Case 3

A 48-year-old female, a nonsmoker, with no known medical illness previously, presented with acute urinary retention. Further history revealed she had hematuria for 5 days prior to developing urinary retention. It was associated with right flank pain. A formal ultrasound was performed showing right middle pole minimally complex renal cyst with right hydronephrosis. A CT Urography revealed right mild hydronephrosis and proximal hydroureter secondary to intraluminal hyperdensities in the right pelvicalyceal system and along the right proximal tubule. A CT renal Multiphase confirmed a right arteriovenous malformation nidus with blood products in the collecting system causing obstructive uropathy. It described a tortuous vessel with a nidus of 1.2 × 1.7 cm. The feeding artery arises from the anterior division, middle segmental artery and drains into the right renal vein (**Figure 13**) Early opacification of the right renal vein further reaffirmed the diagnosis of AVM (**Figure 14**). Following this, she underwent a superselective renal artery angioembolisation of the right midpole artery (**Figures 15-18**). Cook coils were the embolic agent used. Post embolization renal angiogram showed 20% renal perfusion with no other abnormal vessels (**Figure 19**). She is currently planned for a post embolization CT. Her renal function has remained stable to date, with no decline in eGFR.

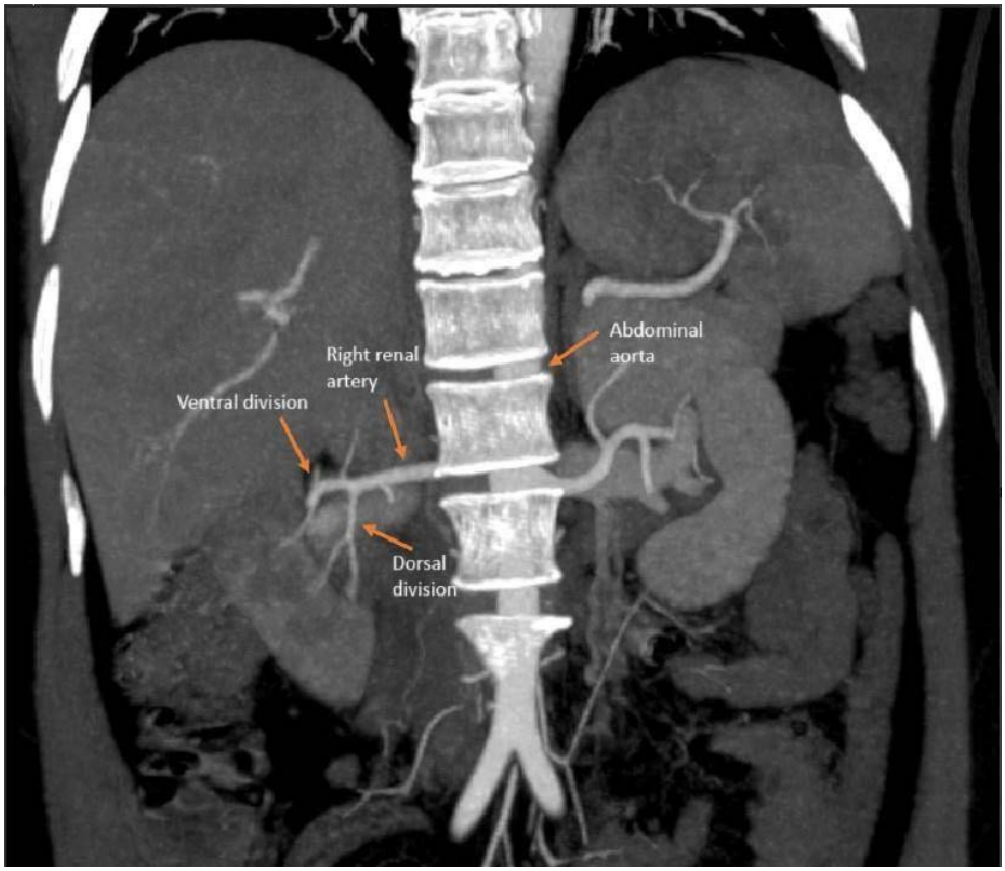

Figure 13. CT renal multiphase. Coronal view.

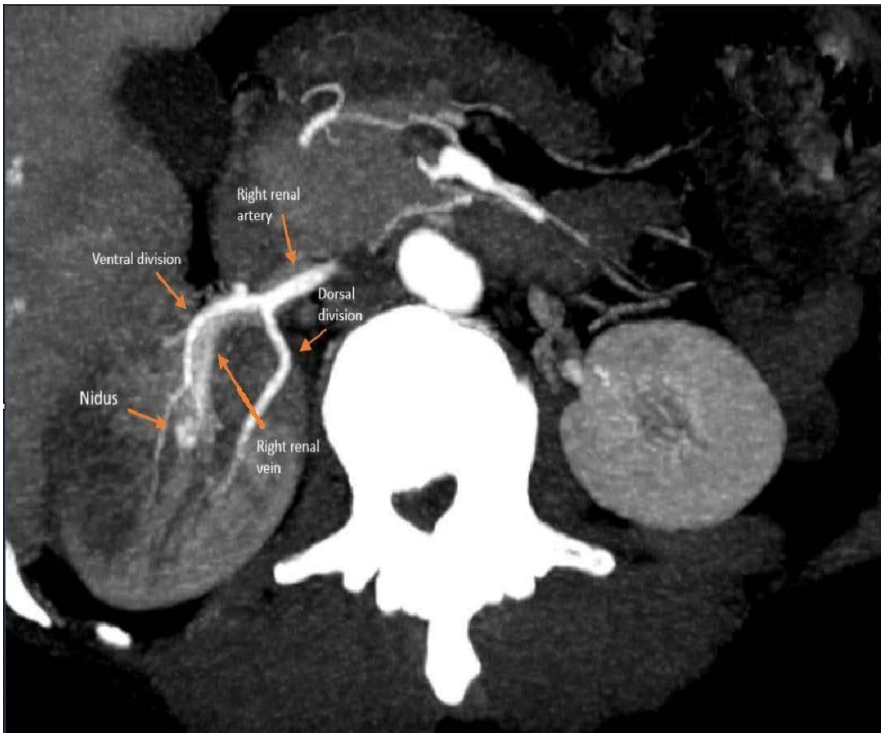

Figure 14. CT renal multiphase. Axial view. Presence of nidus arising from anterior/ventral division of right renal artery. Early opacification of the renal vein in keeping with AV malformation.

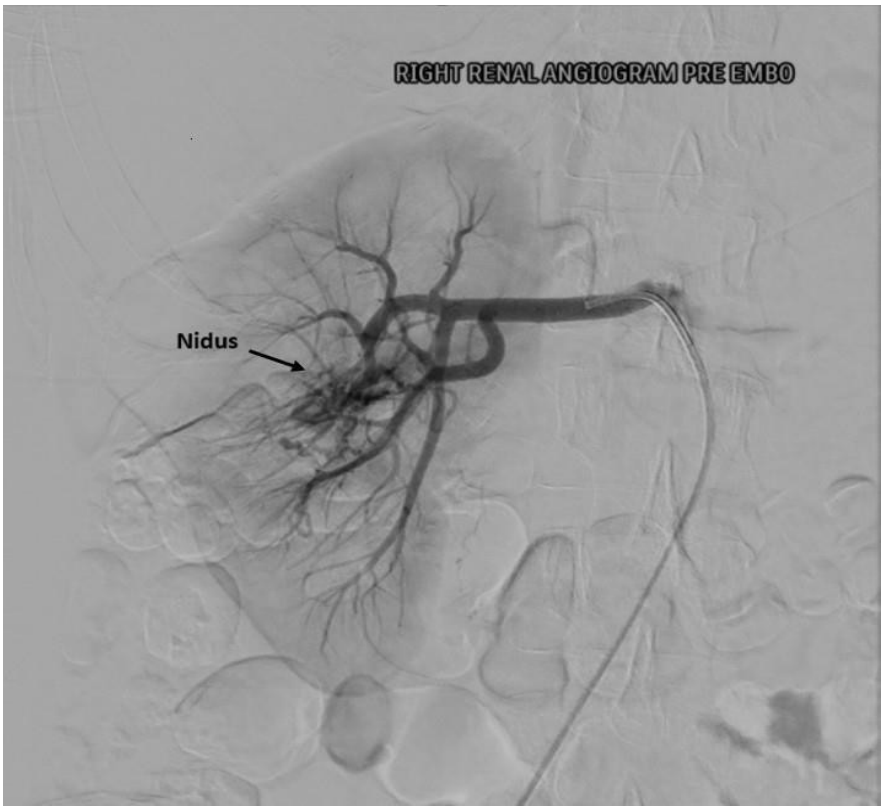

Figure 15. Pre embolization angiogram shows nidus at the midpole of the right renal artery.

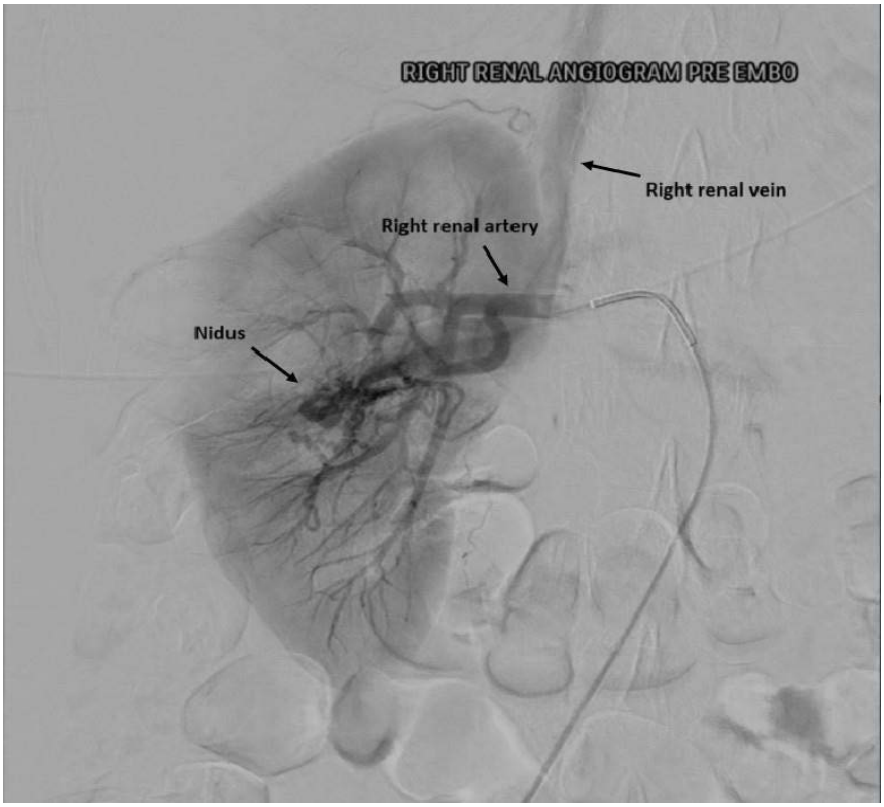

Figure 16. Early opacification of the right renal vein in keeping with the AV malformation.

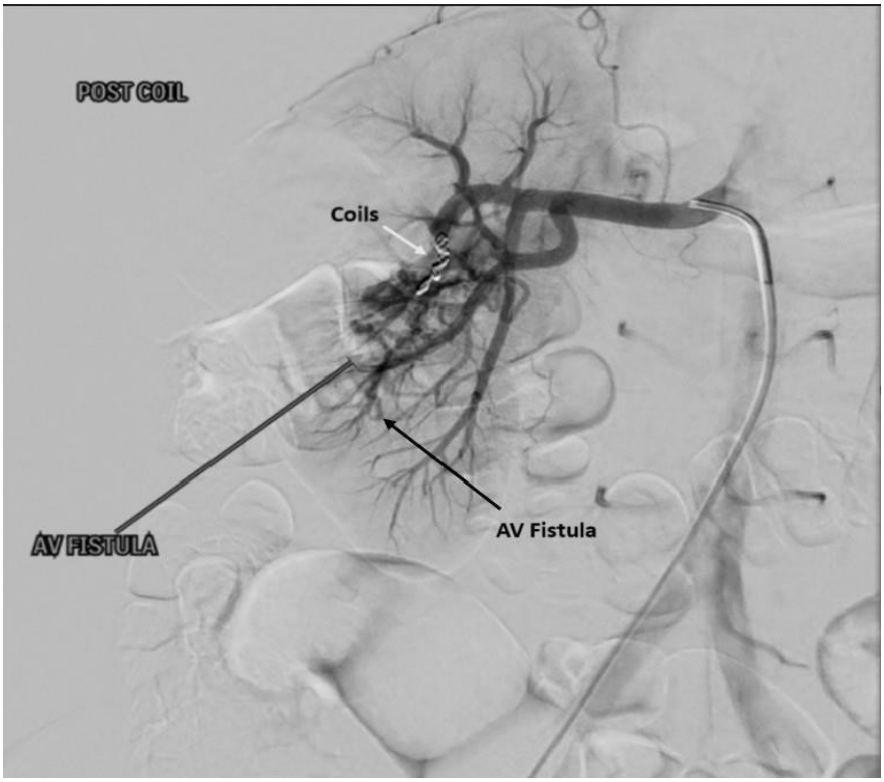

Figure 17. Post coiling shows multiple serpiginous tortuous arteries with early venous opacification from mid and lower pole arteries.

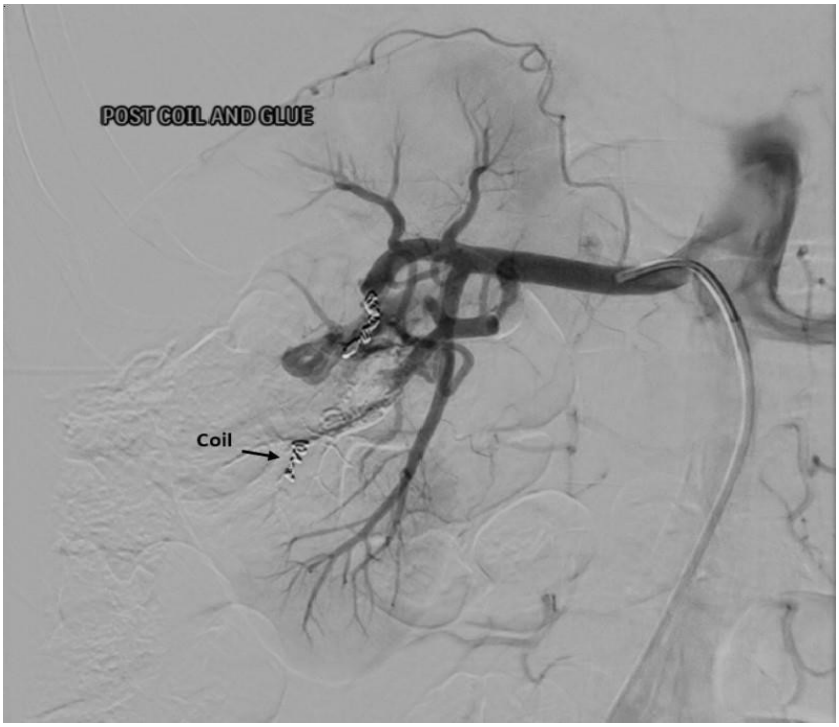

Figure 18. Post coiling and embolization of right pole renal artery branch.

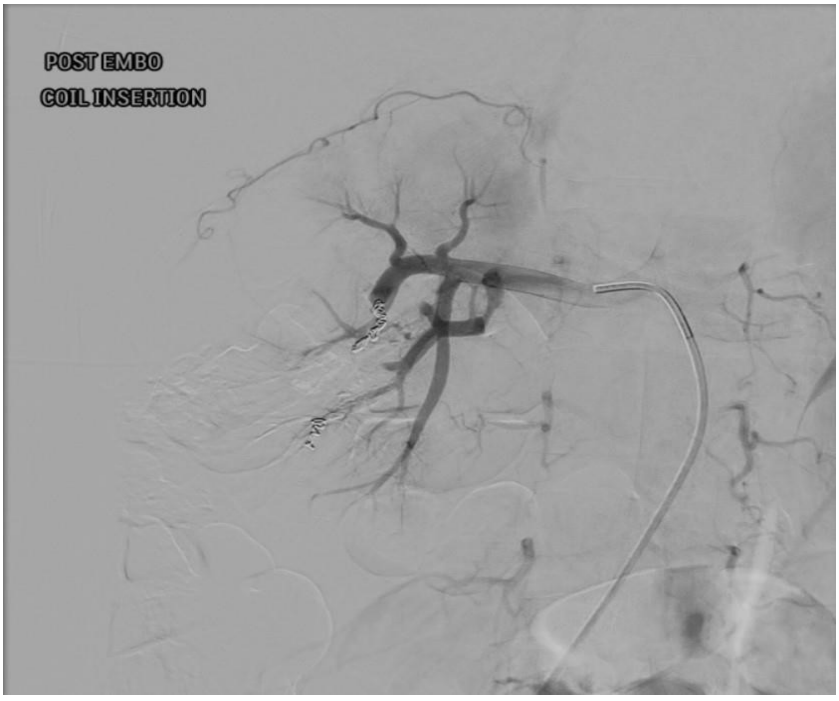

Figure 19. Post embolization and coiling angiogram of right main artery shows no abnormal vessels seen. Approximately only 20% renal perfusion demonstrated post embolization in keeping with severe right renal infarction post embolization.

4. Discussion

Renal Arteriovenous malformation is considered an uncommon pathology in the

human population. With a prevalence of only 0.04%, it is very often underdiagnosed or misdiagnosed [1]. Renal AVMs by definition are a configuration of abnormal connections between renal arteries and veins that are devoid of an intermediating capillary bed. The result of these connections is the formation of a vascular nidus which appears as composition of dilated and tortuous vessels [2]. In terms of risk factors, Gender predisposition plays an important factor as rAVMs tend to affect females twice as much than men. In our case series, we can see gender predispostition as a favourable factor for renal AVMs as all our patients in this case series are females. However, contrary to the reported peak incidence age group of renal AVMs which is usually during the 3rd decade of life, all the patients reported in this case series were ranging between 50 - 70 years old at the time of diagnosis [3].

Renal AVMs can be divided into congenital, acquired and idiopathic renal AVMs. Of the three, acquired renal AVMs are the most common making up 70% - 80% of all renal AVMs. They usually manifest as arteriovenous fistulas occurring as a result of iatrogenic injuries such as renal biopsies, penetrating or blunt trauma, or renal malignancy or inflammation [4]. The congenital type representing about 25% of renal AVMS, usually occur during 4th to 10th week of intrauterine fetal development due to failures in focal spontaneous vascular development. This type of AVM usually only becomes symptomatic during the 3rd and 4th decades of life [5].

Idiopathic aneurysms though are the rarest form renal AVMs, comprising of only 5% of all renal avms cases [6]. True to its terminology, idiopathic renal avms have no identifiable cause. In this case series, we identified a growing prevalence of Renal AMVS in East Malaysia, particularly the state of Sabah. Previous literature describes that most cases of rAVMs are found to be occurring in the left kidney, more commonly involving the upper pole, but can occur in mid pole and lower pole as well in equal proportions. Moreover, they predominantly occur in the renal parenchyma rather than the collecting system [7].

Renal AVMs can present in a wide range of clinical scenarios, from asymptomatic instances to severe symptoms like hematuria, flank pain, perinephric hematoma, abdominal mass, abdominal bruit, hypertension, and in severe cases, renal failure or life-threatening hemorrhage. Hematuria is a common symptom of renal AVMs, but ironically renal AVMs are not a common cause of hematuria. This is clinically evident in the cases reported, in which the majority of them presented with a main complaint of hematuria. On a histological level, the absence of the elastic lamina beneath the renal calyx or renal pelvis mucosa makes the vessels fragile and more prone to damage. Increased vascular pressure causing rupture of these dysplastic subepithelial vessels explains the pathogenesis for the development of hematuria in renal AVMS [8]-[10]. This occurs in response to increased vascular pressure as high-pressure arterial blood is directly shunted into low-pressure veins causing blood to enter the collecting system. In cases where hematuria is severe, blood clots may form, and may result in blockage of the renal pelvis and

ureters. As a result, the patient may develop flank pain and abdominal pain which was also evident in some of the cases discussed in this case series [11].

The growing prevalence of renal avms in east malaysia, raises the concerns to determine the potential causes or risk factors in hopes of finding a common causative link for predisposition. In this study, we observed that all patients were female, aged between 50 and 72 years, which is well established as known risk factors favouring renal AVMS. Another similarity we found was that all 3 patients were of Kadazan-Dusun ethnicity. While this may suggest a potential demographic clustering, we emphasize that such findings are hypothesis-generating and limited by the small sample size. Statistical inference regarding racial predisposition cannot be drawn from this limited case series.

The molecular mechanisms for the pathogenesis renal arteriovenous malformation (rAVM) formation aren't completely understood, but emerging evidence implicates genetic and angiogenic dysregulation similar to that described in hereditary vascular disorders such as hereditary hemorrhagic telangiectasia (HHT) and capillary malformation–arteriovenous malformation (CM-AVM) syndrome [12]. Mutations in genes such as *ENG* (endoglin) and *ACVRL1* (ALK-1), both key regulators of the transforming growth factor-β (TGF-β) signaling pathway, result in endothelial dysfunction and aberrant vascular remodeling. Some studies suggest that the loss of endothelial endoglin function, and the role vascular endothelial growth factor (VEGF) as an angiogenic stimuli may promote direct arteriovenous shunting [13].

As of now, not much of molecular studies has been done on Renal AVM, however most of these mechanisms are already well established in cerebral and systemic AVMs. Therefore, it is plausible that renal AVMs share a similar pathophysiological basis. Somatic mutations involving the RAS-MAPK signaling pathway (including *KRAS* and *MAP2K1*) have also been reported in sporadic AVMs, promoting endothelial proliferation and vessel wall instability [14] [15]. Further molecular characterization of renal AVMs is therefore warranted to determine whether comparable genetic or angiogenic mechanisms contribute to their formation [16].

Moreover, the formation of AVM can be attributed to blood flow and shear stress on the vasculature. Studies have shown that under normal circumstances, healthy Endothelial cells (EC) migrate against the direction of blood flow which helps to maintain normal vasculature. However, in cases where there is loss of endoglin, ECs become dysfunctional and tend to migrate in the direction of blood flow. This inevitably leads to accumulation of ECs on the venous side of capillaries, thus supporting the notion that the formation of AVMs in Hereditary hemorrhagic telengectasia might originate at the venous side [17]. The histological appearance of arteriovenous malformations (AVMs) consists of tortuous arteries, irregular in caliber, with fragmented and thickened elastic laminae. Conversely, the veins commonly exhibit intimal hyperplasia, thickening of the perivascular smooth muscle, and adventitial fibrosis as a result of the high-pressure, turbulent

arterial flow [18]. Therefore, it could be postulated that the pathogenesis of renal avms could develop as a result of endothelial dysfunction and overactivation of pro angiogenic pathways,

Diagnosing Renal Avms can be challenging task. Therefore, the initial workup usually focuses on ruling out associated pathologies such as urological malignancies and renal colics. In the event these pathologies have been excluded, a targeted vascular imaging would be warranted. The main modalities of establishing a diagnosis is via a computed tomography (CT) angiogram or digital subtraction angiography (DSA) [19].

Emergency management for rAVM includes surgery or transcatheter renal artery embolization (RAE). RAE has become the preferred treatment option due to the advancements in endovascular embolization [20]. Its advantages include less invasiveness, greater effectiveness, avoidance of general anesthesia, shorter hospital stays [21], reduced perioperative and postoperative morbidity, and preservation of unaffected renal parenchyma. Despite these benefits, potential risks remain, such as renal infarction, pulmonary embolism, recanalization, and incomplete occlusion. Successful embolization aims for the total occlusion of the malformed or shunted vessels while preserving normal renal arterial branches and preventing embolic material migration or reflux. Recent studies have demonstrated high technical success and long-term safety of embolization for renal AVMs, with minimal impact on renal function [22]. Our findings are consistent with these reports, as none of the patients in this series experienced post-procedural renal impairment.

In the event of failed embolisation or ruptured AVMs which render the patient hemodynamically unstable, Surgical intervention may be the only alternative of treatment. A total or partial nephrectomy would be warranted to achieve optimal patient outcome. Renal artery ligation can also be considered an option although these are less favorable options as they are more invasive, require longer hospital stays, involve slower recovery, carry higher morbidity due to the risk of complete loss of renal function, involve complications from general anesthesia, and carry postoperative complications [23]. Nonetheless, surgery remains a treatment option in life-threatening conditions where embolization has failed or is technically difficult. The choice of embolic agents depends on the type of rAVM, the cause (traumatic vs. non-traumatic), and the angioarchitecture of the malformed vessels (flow, feeder, size of the fistula, and accessibility to the target lesion).

5. Conclusion

The rare presentation of renal AVMs amongst patients particularly in East Malaysia, Sabah highlights a need for further epidemiological investigation. While demographic or racial predisposition may be suspected, these observations should be viewed as exploratory and not conclusive due to the limited sample size. The non-specific symptoms of this disease make it a diagnostic challenge for clinicians. Prompt recognition of rAVM as a differential diagnosis for unexplained hematu-

ria is essential, as delayed diagnosis may risk rupture and torrential hemorrhage. Endovascular embolization remains the gold standard treatment for renal AVMs. Selection of embolic agents should be tailored to the lesion's angioarchitecture to minimize recurrence. Long-term follow-up is recommended to ensure sustained vessel occlusion and preservation of renal function. Finally, given the complexity of this disease, a multidisciplinary approach involving a urologist, radiologist, and interventional radiologist is crucial to achieve optimal patient outcomes.

Conflicts of Interest

The authors declare no conflicts of interest.

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