

Massive Hematuria Due to Congenital Renal Arteriovenous Malformation Successfully Treated by Renal Artery Embolization

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Abstract

Congenital renal arteriovenous malformations (AVMs) are rare benign vascular lesions and a rare cause of massive hematuria in females predominantly involving right kidney. Clinical presentation in a male with involvement of the left kidney is very rare. Only a few case series describing the outcome of congenital renal AVMs have been reported in the literature. We report a challenging case of a male patient with life threatening massive hematuria with congenital renal AVMs in left kidney. Successful embolization was performed using coils and gel foam.

Introduction

Renal arteriovenous malformations (AVMs) are rare benign vascular lesions which may be acquired or congenital. Congenital renal AVMs are usually small and asymptomatic and close spontaneously.¹ Congenital renal AVMs are more common in females in the third or fourth decade of life and involve the right kidney.² Acquired renal AVMs also known as arteriovenous fistulae, account for about 70% to 80% of all AVMs and may occur as a result of renal biopsy, blunt or penetrating trauma, inflammation, malignancy, or renal surgery.³ Hematuria is the most common symptom. Other clinical manifestations include hypertension, left ventricular hypertrophy, high output cardiac failure and abdominal pain.⁴ The usual treatment of AVMs is nephrectomy but transcatheter arterial embolization (TAE) can be considered as an alternative. We present a rare case of massive life threatening hematuria

in an adult male with congenital AVMs involving left kidney which was successfully treated using TAE.

Case Report

A 46 year old male patient presented to the urology outpatient department of our hospital with repeated episodes of massive hematuria and urinary retention for the last 2 months. Patient had no significant past medical history involving renal injury, renal biopsy or known nephrolithiasis. He denied history of bleeding disorder or any medications. His physical examination revealed blood pressure of 100/70 mm Hg, heart rate was 96 beats/minute, body temperature was 37.4 °C and pallor and no continuous bruit could be appreciated over flanks. Biochemical,

hematological and coagulation parameters revealed creatinine of 1.1 mg/dl, blood urea of 29 mg/dl and eGFR of 105.56 ml/minute, hemoglobin of 7.8g/dl, platelet count of $178 \times 10^3/\text{mm}^3$, aPTT of 31.7 sec and PT of 13.6 sec. For severe anemia patient required multiple packed red blood cell transfusions. Urine analysis showed erythrocytes but malignant cytology was negative. Ultrasonography of abdomen revealed no bladder or kidney stones. CECT and MRI abdomen revealed thickened wall of left pelvis and upper ureter and since these findings may represent urothelial malignancy, cystoscopy and retrograde studies were conducted but were negative. Retrograde study of the left pelvicaliceal system revealed active bleeding. Based on these findings renal arteriography was planned.

Renal arteriography was performed using a right transfemoral approach and a 6- French sheath and initially an aortogram was taken to delineate the renal arteries using the 6-French pigtail catheter. Left renal artery was then cannulated with 6-French judkins right catheter and selective left renal angiogram revealed AV malformation involving the posterior segmental branch of the left renal artery. It was also selectively cannulated with

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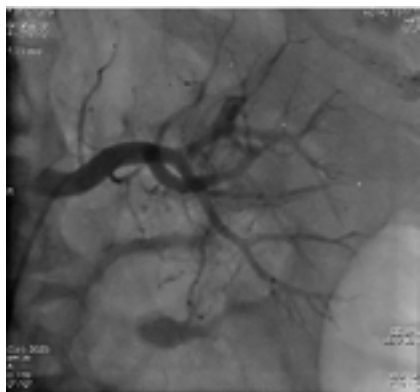


Fig. 1: Selective cannulation of posterior segmental branch of left renal artery with 5FJR4 catheter revealing the AVM

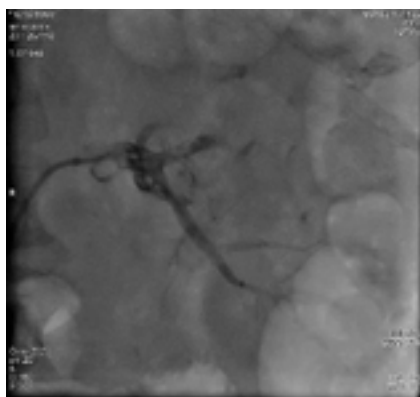


Fig. 2: Complete occlusion of the AVM by embolization coils using 6FLCB catheter

5-French judkins right catheter with terumo wire support and selective angiogram revealed AV malformation arising from the posterior segmental branch of the left renal artery (Figure 1). Digital subtraction arteriography (DSA) demonstrated the feeding artery to the AVMs. The lesion was selectively catheterized with 6-French left coronary bypass catheter with terumo wire support. After confirming the feeding vessel of AV malformation and positioning the catheter into its origin, coil embolization of feeding vessel was done using three MREye embolization coils (Cook Medical Inc. Bloomington, In, USA) – two 5mm and one 4mm coils. At the end of the procedure complete occlusion of the AVM was confirmed using DSA with no procedural complications (Figure 2).

On 7th postembolization day, massive hematuria recurred with hypotension requiring inotropes and hence patient was again taken up for an emergency selective renal arteriography and

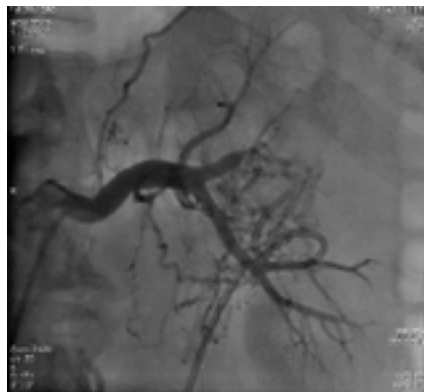


Fig. 3: Multiple feeding vessels to the AVM from left renal artery and its anterior segmental branch responsible for recurrence of hematuria

embolization. Left renal artery was selectively cannulated using 6-French judkins right catheter and left renal angiogram revealed multiple feeding vessels from anterior segmental artery as well as vessels arising directly from the left renal artery which were not present in previous angiogram (Figure 3). Initially selective embolization of the anterior segmental artery feeders was attempted with coils but despite occlusion of these feeding vessels left renal AVM was seen filling through the feeders from the left main renal artery. Hence, coil embolization followed by gel foam embolization was done to left main renal artery. Repeat contrast injection revealed no refilling of left renal AVM (Figure 4). Blood supply of the left kidney had to be compromised to save the life of the patient from the catastrophic bleeding. TAE avoided an inevitable emergency laparotomy and nephrectomy. Patient was observed for recurrence of hematuria, hypertension and postembolization syndrome. No recurrence of hematuria was observed but patient developed postembolization syndrome (PES) with flank pain, fever, nausea and vomiting and was managed conservatively with parenteral opioid analgesics (morphine and fentanyl), antiemetics (ondansetron), antipyretics and intravenous fluids. PES resolved over the next 3-4 days. Subsequent hospital course was uneventful with no worsening of renal function with creatinine of 1.1mg/dl and eGFR of 105.56 ml/minute on discharge.

Discussion

AVMs, first described by Varela in 1928⁵ are rare vascular lesions.

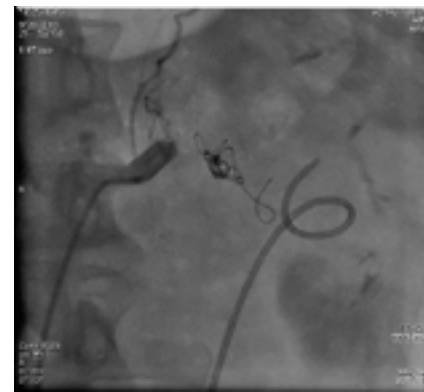


Fig. 4: No refilling of the AVM after coil and gel foam embolization of the left renal artery

Congenital renal AVMs are considered to represent focal spontaneous failures of vascular development occurring between the 4th and 10th week of life.⁶ However they usually remain asymptomatic until the 3rd or 4th decade of life.

On angiographic examination depiction of renal veins in the early stages of the arterial phase confirms the diagnosis of renal AVMs. Congenital AVMs may be classified as cirroid and aneurysmal. Cirroid type AVMs consists of multiple small and dilated varix like arteriovenous communications with multiple feeding arteries and draining veins and they represent a truly congenital form of arteriovenous malformation. Cirroid type AVMs develops from a nidus in the submucosa of renal pelvis. Aneurysmal type consists of a single feeding artery and a single draining vein. Acquired AVMs are primarily aneurysmal. Acquired AVMs etiology may be idiopathic or secondary. Idiopathic renal AVMs present later in life and develop when pre-existing renal aneurysms form shunts with adjacent renal segmental vein. Secondary renal AVMs are caused by iatrogenic injuries, penetrating renal trauma or blunt renal injury. Renal biopsy is the most common cause of secondary AVMs. Renal malignant tumors may cause shunts between pseudoaneurysms and renal segmental veins resulting in AVMs. Life threatening hematuria is more characteristic of the congenital AVMs and is present as the primary symptom in 3 out of 4 patients. Aneurysmal type of congenital AVMs is generally asymptomatic and is often incidentally detected on CT abdomen or ultrasonography.

AVMs are complex lesions and are almost never entirely cured and may recur. The goal of treatment should be to control the symptoms and perform repeated embolizations if needed. Patients should be counseled that treatment may be life-long and that currently there is no cure. Indications for treating AVMs are increase in the size of the fistula, recurrent or persistent hematuria, hemodynamic effects such as hypertension and high output cardiac failure. TAE has become the treatment option of choice for the management of severe hematuria caused by renal AVMs, even in case of AVMs complicating pregnancies.⁷ Embolic materials used for TAE include gelatin sponge, metallic coil, absolute alcohol, lipiodol, and n-butyl 2-cyanoacrylate. In our case platinum coils and gel foam was used for successful treatment of AVMs. TAE is a safe procedure with a relatively low rate of complications. The most common complication is PES affecting 90% of patients.⁸ PES usually present with flank pain, fever, nausea, vomiting, paralytic ileus, and/or leukocytosis for 1 to 3 days after renal artery embolization (RAE).⁹ Treatment is supportive, consisting of analgesics, antipyretics, and antiemetics as needed, until symptoms resolve. For this reason, it is recommended that patients be observed in a hospital after RAE for monitoring and control of symptoms.

Coil migration is an unusual, but

serious complication, occurring in less than 2% of cases.⁷ It is commonly detected at the end of the procedure and can be rectified using endovascular grasping device (snare). Non-target embolization can result in spine, lower extremity, and bowel infarction.¹⁰⁻¹² Reflux of embolization material can result in loss of renal function and subsequent hypertension. Other complications include access site hematoma, infection and contrast induced nephrotoxicity.

Conclusions

A middle aged male with life threatening hematuria after ruling out common causes of hematuria underwent renal angiogram to reveal features of congenital renal AVM in the left kidney and subsequently an emergency embolization was successfully performed using coils and gelfoam. Clinical presentation of congenital renal AVM's in a male with involvement of the left kidney is very rare. Our case also highlights the fact that symptoms, history and imaging may be misleading in renal AVM's. In cases of suspected renal AVMs, selective renal arteriography and DSA can be both diagnostic and therapeutic modality. The patient is in our follow up with no recurrence of hematuria or worsening azotemia.

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