

# RENAL ARTERIOVENOUS MALFORMATIONS- REVIEW OF LITERATURE

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### Introduction

Renal arteriovenous malformations are abnormal communications between the intrarenal arterial and venous systems. The condition was first described by Varela in 1928<sup>1</sup>. Renal arteriovenous malformations are uncommon and usually identified during the evaluation of gross haematuria<sup>2</sup>. The estimated rate in large autopsy series is less than 1 case per 30,000 patients. But incidence of 1 per 1000-2500 patients was found to those undergoing evaluation for urologic or vascular imaging techniques. They may cause hypertension, peripheral embolization, high output cardiac failure and haematuria<sup>3</sup>. Renal arteriovenous malformations are congenital, acquired and idiopathic<sup>2</sup>. Congenital arteriovenous malformations account for less than one third of renal arteriovenous malformations. Two types of congenital renal arteriovenous malformations are described. The cirroid arteriovenous malformation is the most common type, a dilated corkscrew appearance, much like a varicose vein. The cavernous congenital arteriovenous malformation with single dilated vessels is less common<sup>4</sup>. With the increased incidence of percutaneous renal surgery, renal biopsies, penetrating renal injuries, incidence of acquired renal fistulas are increased in recent days. Acquired arteriovenous fistulas represent as many as 75-80% of renal arteriovenous malformations. Acquired renal arteriovenous anomalies are often termed renal arteriovenous fistulas. Idiopathic renal arteriovenous fistulas have the radiographic characteristics of acquired fistulas, may be associated with renal artery aneurysms, represents less than 3% of renal arteriovenous malformations<sup>4</sup>. Treatment can be tailored to the individual patient. Options for therapy range from observation to embolization to nephrectomy.

### Etiology

The etiology of congenital arteriovenous malformations is unknown. The cause of acquired arteriovenous malformations is usually known. Percutaneous renal biopsy is the most common known cause of acquired renal arteriovenous fistula<sup>5-7</sup>. Trauma is another important, although uncommon, cause of acquired renal

fistulas<sup>8</sup>. In patients with hypertension following renal trauma, renal arteriovenous malformations may occur in one third of patients. In patients with penetrating trauma, arteriovenous fistulas may affect as many as 80% of patients with post-traumatic hypertension. Trauma during ureteroscopy has recently been described as a cause of intrarenal arteriovenous fistula<sup>9</sup>. Idiopathic arteriovenous fistulas are thought to arise from the spontaneous erosion or rupture of a renal artery into a nearby renal vein. Arteriovenous malformations may also occur in the setting of malignancy. Angiogenic tumor factors have been implicated and may explain the development of arteriovenous malformations within renal tumors.

### Pathophysiology

In the cirroid congenital arteriovenous malformation, multiple communications exist between the arteries and veins. These communications develop multiple coiled channels, forming a mass within the renal parenchyma<sup>2</sup>. The communicating vessels are tortuous, dilated, and located beneath the lamina propria of the renal urothelium. This cluster of vascular channels forms a mass, with the arterial supply arising from one or more segmental or interlobar renal arteries. Its nearness to the collecting system may explain the high prevalence of hematuria<sup>2</sup>. The less common cavernous congenital arteriovenous malformation is characterized by a single artery that feeds into a single cystic chamber, with a single draining vein. Acquired arteriovenous malformation results from traumatic disruption of renal vessels. A fistulous connection between the arterial and venous systems occurs as a result of the trauma. Any renal arteriovenous malformation may result in renin-mediated hypertension.

### Presentation

Patient with arteriovenous malformations may remain asymptomatic<sup>10</sup>. They may be discovered incidentally on image studied performed on other reason. Usual mode of presentation is Gross hematuria (as many as 75% of cases)<sup>2</sup>. Sometimes the diagnostic evaluation of

patients with microscopic hematuria also may lead to the discovery of an arteriovenous malformation. Flank pain with ureteric colic from obstructing blood clots may lead to the diagnosis of arteriovenous malformation, which may be voided as wormlike masses. A significant percentage of patients with renal arteriovenous malformations are hypertensive. Cardiomegaly, congestive heart failure, or both also may be present among patients evaluated for renal arteriovenous malformations<sup>2</sup>. Rarely, a patient may present with hypotension from hemorrhage caused by an arteriovenous malformation. A physical evaluation may demonstrate findings of a flank bruit. A palpable mass is usually present in those patients with renal tumors as the cause of the fistula.

### Diagnostic workup

The laboratory evaluation is dictated by the clinical presentation of the patient. Haemoglobin and hematocrit estimation is important for correction of Anemia. Anaemia may contribute to the severity of heart failure in some patients with renal arteriovenous malformations.

Serum creatinine values are measured to assess the renal function before contrast-enhanced radiographic studies are performed. Renal function may also dictate the type and timing of surgical intervention. Prothrombin time, activated partial thromboplastin time, bleeding time should be done to exclude any coagulopathies. Urinary tract infections should be excluded before intervention is pursued.

### Imaging Studies

The initial study for the evaluation of gross hematuria depends on several factors, including location, urologist and radiologist preference, and patient factors. The successful treatment of renal arteriovenous malformations relies on definitive localization of the lesion. Meticulous radiographic evaluation is needed because some lesions are subtle. The characteristics of renal arteriovenous malformations on, ultrasound, IVU and CT scan images are described.

Ultrasound is a noninvasive means for evaluating renal causes of haematuria. On the other hand many patients incidentally diagnosed during ultrasound study. Color-duplex Doppler ultrasound studies increases the sensitivity for vascular lesions. The lesions are identified as arteriovenous malformations based on the turbulent blood flow within a cystic mass<sup>11</sup>.

CT scan is now becoming a next modality for evaluation of haematuria or renal mass lesion detected on

ultrasound examination. CT urography has replaced IVP in many centers for the initial evaluation of hematuria. With modern spiral CT scanners and IV contrast, detailed anatomic and functional information can be obtained and can lead to the accurate diagnosis of renal arteriovenous malformations. With proper equipment and oversight, CT urography, angiography, or both can provide information about renal function, as well as detailed definition of the anatomy, including the vascular and collecting systems<sup>12</sup>.

Intravenous pyelography still being used commonly for the evaluation of haematuria. Arteriovenous malformations have several characteristics on IVP images. A mass lesion may be observed on the nephrotomogram images, especially in the medullary region, with compression of the collecting system. Hypo perfusion distal to the arteriovenous malformation may be present, which manifests as a wedge-shaped defect or segmental nonvisualization. Filling defects of the collecting system may also be present. The arteriovenous malformation may cause an irregular impression on the collecting system, and clots may fill and obscure a calyx or the renal pelvis. However IVP results may be normal in patients with an arteriovenous malformation.

Magnetic resonance angiography (MRA) is a promising technology for the evaluation of renal masses<sup>13</sup>. MRA is especially useful in those patients who cannot tolerate iodine-based contrast. Several reports have confirmed the diagnostic usefulness of MRA for the diagnosis of renal arteriovenous malformation<sup>14</sup>.

Angiography remains the standard for the clinical diagnosis of arteriovenous malformation<sup>15</sup>. Angiography of an arteriovenous malformation demonstrates rapid contrast visualization in the inferior vena cava within seconds of contrast injection because of the rapid shunting of blood from the arterial system to the venous system. Decreased density on the nephrogram also may appear distal to the arteriovenous malformation. The actual malformation may be a subtle blush if the arteriovenous malformation is small, or the multiple small tortuous vessels may be easily visualized. Cirroid arteriovenous malformations are supplied by multiple arteries, while the cavernous arteriovenous malformations and arteriovenous fistulas tend to be supplied by single vessels.

Cystoscopy should be performed to evaluate for coincidental lower tract pathology. Cytological evaluation of the urine is also useful for screening for carcinoma in

situ of the bladder, which can be missed during diagnostic Cystoscopy.

## **Treatment**

### *Medical Therapy*

Conservative therapy can be used safely in some cases. Acquired arteriovenous fistulas tend to resolve spontaneously<sup>4</sup>. A recent report describes spontaneous resolution of an arteriovenous malformation. Asymptomatic small lesion without any hemodynamic complications may warrant observation. Medical management is also essential to optimizing outcome before surgical intervention. In addition to relieving pain, hypertension should be treated. Heart failure must be controlled. Blood transfusions may be needed for the patient with hemorrhage from an arteriovenous malformation.

### *Intervention*

The initial therapy for treatment of arteriovenous malformations is usually angiographically guided embolization of the malformation (16-19). Numerous substances have been injected in an effort to ablate the arteriovenous malformation. Initial attempts at embolization were complicated by recurrence of the arteriovenous malformation. This was thought to be due to the type of material used for embolization. Materials that have been used for embolization include steel coils, autologous blood clots, gelatin sponges and foams, and synthetic polymers. Super selective embolization with coils and microspheres has also been described<sup>17</sup>.

The most effective material for embolization appears to be absolute alcohol, which is relatively inexpensive. Injection through the catheter lumen is also easier than with many of the synthetic materials<sup>19</sup>. Balloon catheters are used to occlude the feeding artery to prevent retrograde migration of the alcohol. The alcohol denatures the proteins within the wall of the arteriovenous malformation, thereby inducing thrombosis and occlusion of the malformations. Repeat treatments may be needed to completely ablate the arteriovenous malformation. Epinephrine injection before embolization may make the procedure more effective by inducing vasospasm, thereby concentrating the injected material within the arteriovenous malformation. Contrast-induced nephropathy and allergic reactions may occur during embolization technique. The agent used for embolization may migrate or be misdirected and thus cause damage to normal renal tissue or other organs. A recent case description noted coil and guidewire erosion into the

colon<sup>20</sup>. Alcohol may cause transient headaches and mild intoxication. Recurrence or persistent fistulas are possible. Hematomas and pseudo aneurysm at the puncture site are not uncommon.

### *Surgical Therapy*

The treatment most likely to cure an arteriovenous malformation is total or partial nephrectomy. Total nephrectomy is indicated for large cirroid arteriovenous malformations<sup>21</sup>. In most cases, nephrectomy is reserved for patients in whom more conservative therapy has failed. If the fistula is due to malignancy, then radical nephrectomy is usually indicated<sup>2</sup>.

Partial nephrectomy has been accepted as a safe treatment for small, polar lesions<sup>2</sup>. In many centre's laparoscopic partial and total nephrectomy have also been used with increasing frequency to treat selected renal arteriovenous malformations<sup>22</sup>. Recurrence arteriovenous fistulas may occur following partial nephrectomy which may be silent and discovered incidentally during subsequent imaging studies. They also may manifest with signs or symptoms similar to the original arteriovenous malformation.

Small malformations located in the peripheral aspect of the kidney may be treated by ligation of feeding vessels. The dissection of the feeding vessels may be technically difficult. Bench surgery with auto transplantation may facilitate the successful treatment of large and/or centrally located malformations<sup>23</sup>. This degree of renal reconstruction is rarely necessary but may preserve enough functional renal tissue to avoid dialysis in select cases.

## **Conclusion**

Renal arteriovenous malformations remain an uncommon clinical problem. High index suspicion is necessary while evaluation a renal lesion with gross haematuria or hypertension. Several case reports describe clinical situations in which a renal arteriovenous malformation was classified incorrectly as a malignant tumor or as hydronephrosis. Specific CT scan protocols seem especially promising as a minimally invasive way to improve the classification of renal masses. Further, improvements in MRI, MRA, and Doppler ultrasound may decrease the need for the use iodinated contrast agents. Angiographic embolization treatment is now the usual first line of therapy because it can be accomplished at the time of diagnosis, with little morbidity. However nephrectomy remains the criterion standard for treating renal arteriovenous malformations.

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