Renal Artery Stenosis With Severe Hypertension: A Case Report

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ABSTRACT

Background: Renal artery stenosis (RAS) is found in 77% of hypertensive patients and is responsible for 1-2% of systemic hypertension. Arteriosclerotic renal artery disease is commonly seen in older patient and rarely seen below 50years, while fibromuscular dysplasia is seen in young adult female with age range of 15-30years.
Objectives: A case of incidentally diagnosed congenital RAS with severe hypertension in a 28-year old lady and the role of radiology in diagnosis of RAS is reported.
Case: A 28-year old lady who presented to the general outpatient department of Aminu Kano Teaching Hospital with 3days history of headache, dizziness and malaise. The patient was not a known hypertensive or diabetic. Examination revealed high blood pressure of 160/120mmHg. Abdominal ultrasound revealed a shrunken right kidney, Intravenous urography showed delayed nephrogram and delayed excretory phase with decrease density in the collecting systems, computed tomogram confirmed shrunken right kidney. Digital subtraction angiography (DSA) showed the stenosed right renal artery involving the ostium and the proximal one-third.
Conclusion: Renal artery stenosis should be suspected in young people with unexplained hypertension and radiologic investigations are key in confirming diagnosis.

KEYWORDS: Renal artery stenosis, young female, radiologic investigations, incidental diagnosis

Introduction

Renal artery disease is usually caused by atheromatous lesion and fibromuscular dysplasia[1]. It has also been found to be associated with neurofibromatosis[2]. Renal artery stenosis (RAS) is found in 77% of hypertensive patients and is responsible for 1-2% of systemic hypertension. Arteriosclerotic renal artery disease is commonly seen in older patient and rarely seen below 50years, while fibromuscular dysplasia is seen in young adult female with age range of 15-30years[3].

Arteriosclerotic renal artery disease frequently involves the proximal 2cm of the main renal artery. It is usually demonstrated on angiography as eccentric stenosis in proximal 2cm of renal artery and involving the orifice mostly. Fibromuscular dysplasia on the other hand is seen mainly in the mid and distal main renal artery with proximal 3rd of the main renal artery being spared in 98% of cases.

Other causes of RAS include fibrous band (congenital, retroperitoneal fibrosis, post radiation), arteritis (Buegers disease, polyarteritisnodosa, Takayasu disease, thromboangitis obliterans, syphilitic arteritis), arteriovenous malformations/fistula, thromboembolic disease (atrial fibrillation, prosthetic valve

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thrombi, cardiac myxoma, paradoxical emboli, atheromatous emboli), renal artery aneurysm, extrinsic compression (renal cyst, neoplasm, chronic subcapsular hematoma, middle aortic syndrome (aortic dissection, dissecting aortic aneurysm) posttraumatic renovascular hypertension.

A case of undiagnosed congenital renal artery stenosis and severe hypertension found incidentally in a young lady of 28-years and the value of radiology in establishing the protocol for diagnosis of RAS as well as the occurrence of RAS at a young age prompted this report.

Case Presentation

A. T. is a-28-year old housewife that came to the general outpatient department of Aminu Kano Teaching Hospital with 3days history of headache, dizziness and malaise. No history of fever, cough or vomiting. The patient was not a known hypertensive or diabetic. Clinical examination revealed an anxious young woman, not pale, anicteric, not in obvious respiratory distress. Abdominal examination and central nervous system were essentially normal. Cardiovascular examination however revealed a high blood pressure of 160/120mmHg with normal apex beat and normal first and second heart sounds. No cardiac murmur was heard.

Laboratory investigation revealed normal Full blood count (FBC), Fasting blood sugar (FBS) and electrolytes and urea. Abdominal ultrasound revealed a shrunken right kidney measuring 67mm as opposed to the left which measured 102mm in bipolar lengths. It appeared slightly echogenic with preserved corticomedullary differentiation and normal calyceal caliber (Fig 1). Intravenous urography and contrast CT were then recommended. The IVU showed delayed nephrogram and delayed excretory phase with decrease density in the collecting systems (Fig 2). An impression of renal artery stenosis was made. The computed tomogram showed a shrunken right kidney (Fig 3) with the coronal reconstruction demonstrating it better (Fig 4). The patient then decided to travel to Egypt for further evaluation, where she did renal scintigraphy and digital subtraction angiography (DSA). The angiography showed the stenosed right renal artery involving the ostium and the proximal one-third (Fig 5 and 6). The radionuclide images show a poorly perfused and shrunken right kidney (Fig 7). A decision was taken there to place the patient on six months trial of antihypertensive and to continue follow up.

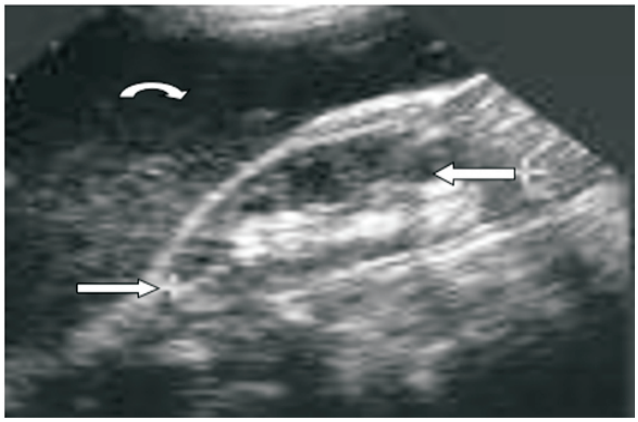

Fig. 1: A sonogram showing a slightly shrunken and echogenic kidney (straight arrows) with the liver superiorly (curved arrow).

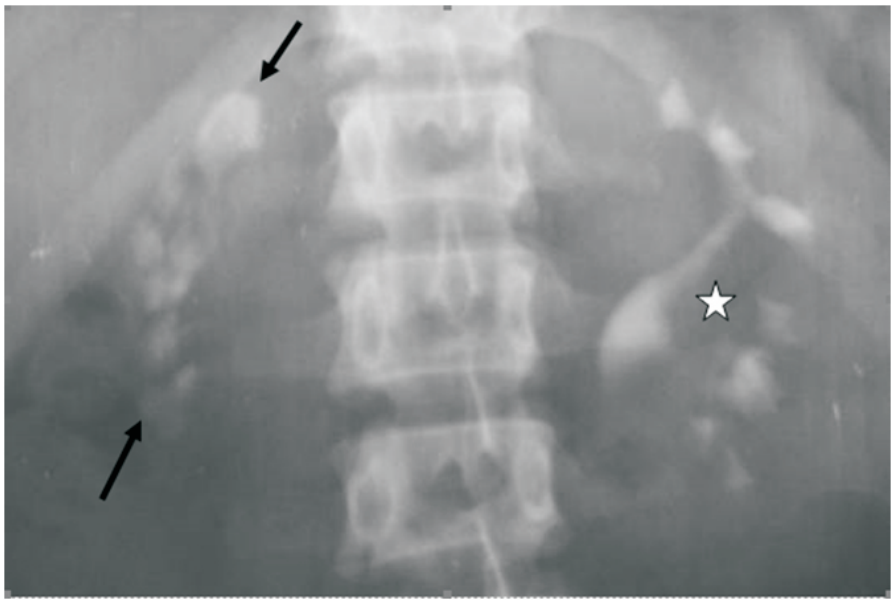

Fig. 2 : A 15min intravenous urography showing decrease density of the right kidney (straight arrows) with delayed appearance of the collecting systems.
The left kidney is normal (star).

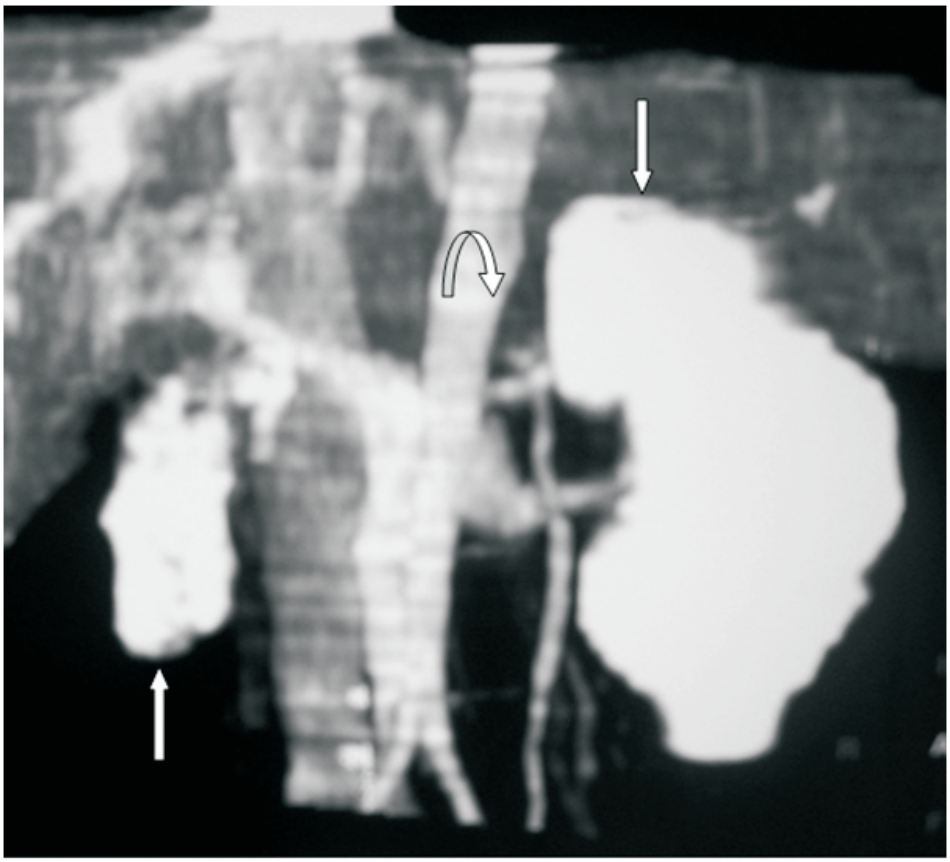

Fig. 4 : Coronal Reformatted Computed Tomographic Image demonstrating shrunken right kidney (arrow up) with a normal left kidney (arrow down), the aorta (curved arrow down).
(Image from Nasser Institute Hospital Cairo, Egypt)

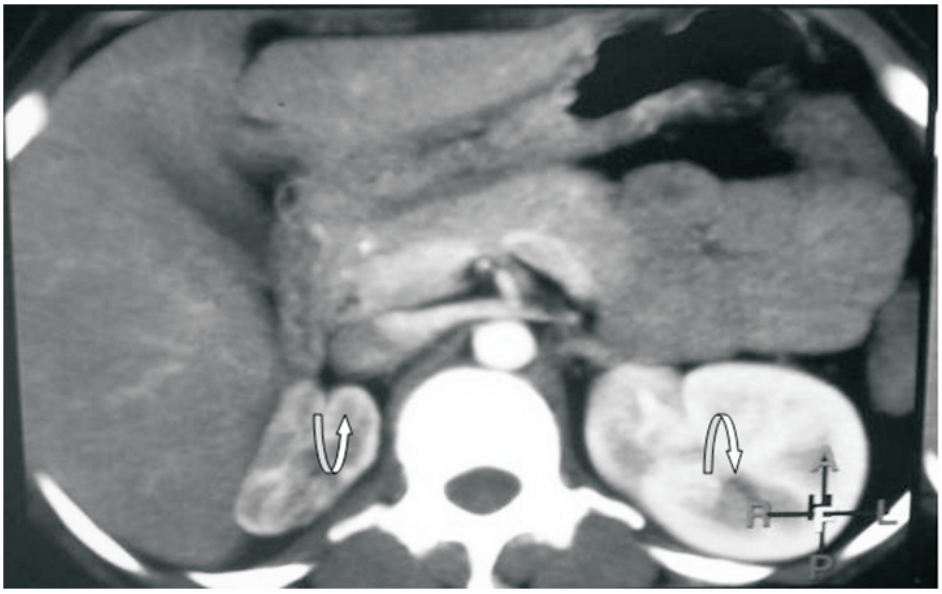

Fig. 3 : Axial contrast enhanced CT image of the upper abdomen, showing enhancing but shrunken right kidney *(curved arrow up)* and normal left kidney *(curved arrow down).*

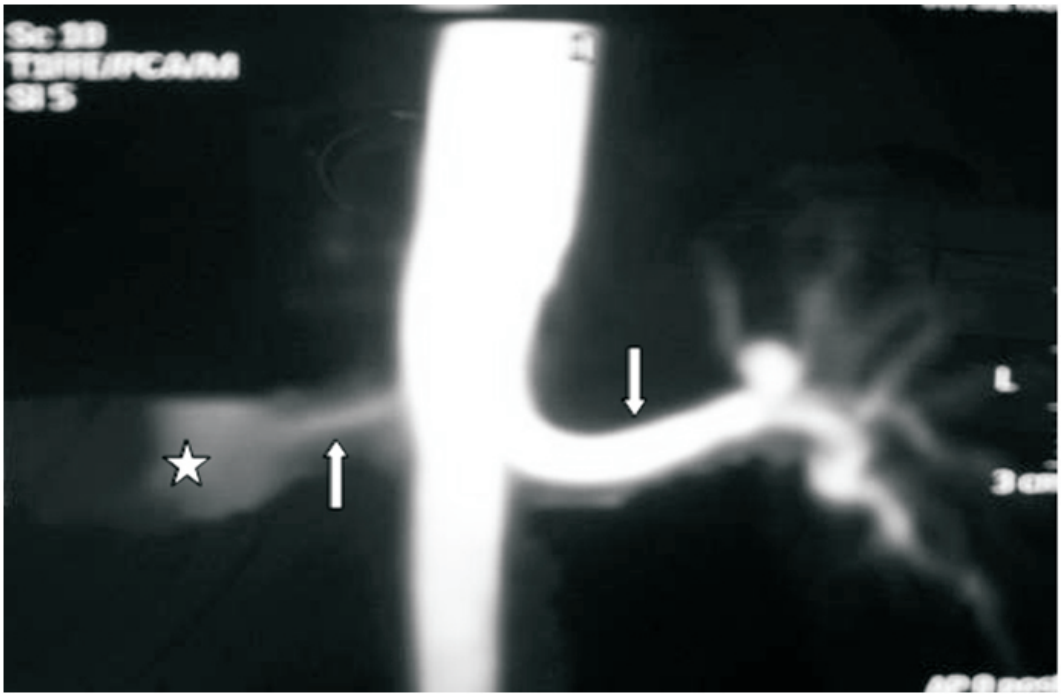

Fig. 5: Renal Angiogram showing stenosis of ostial and proximal 1/3 of the right renal artery (arrow up), ipsilateral post stenotic dilatation (star) and normal left renal artery (arrow down). (Image from Nasser Institute Hospital Cairo, Egypt**)**

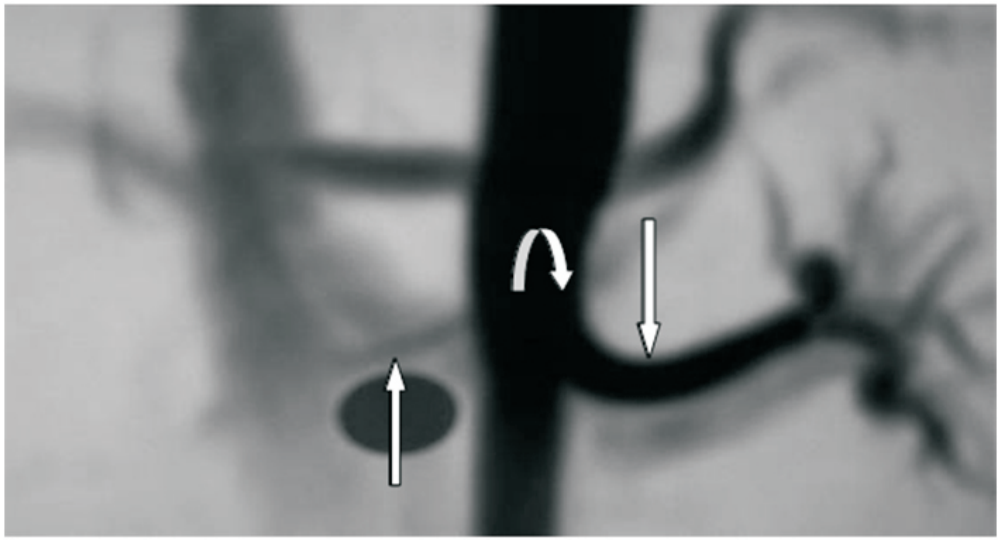

Fig. 6: Digital Subtraction Angiography demonstrating the stenosed right renal artery (arrow up) with normal left renal artery (arrow down). The aorta is seen centrally (curved arrow). (Image from Nasser Institute Hospital Cairo, Egypt)

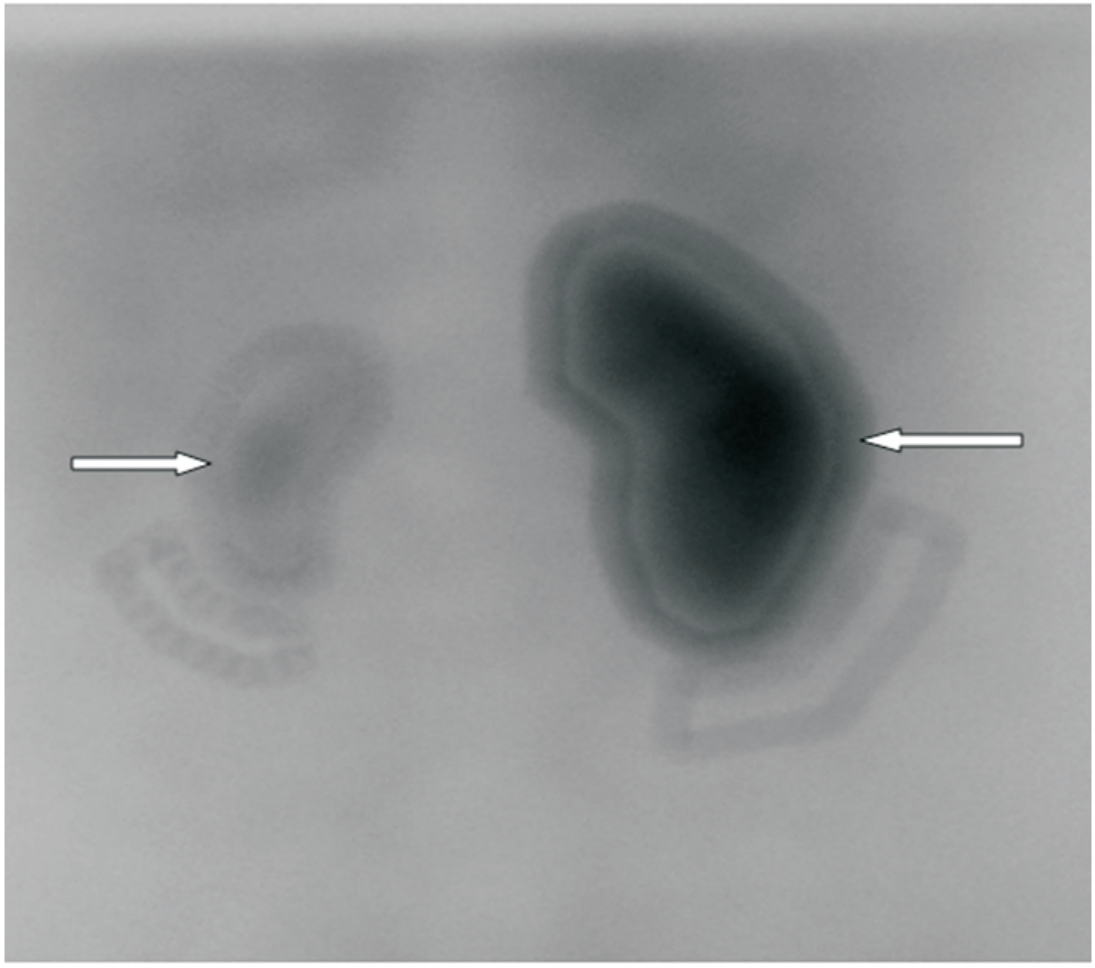

Fig. 7 : Renal Scintigraphy show a reduced activity in the right kidney (right arrow) with a normal left kidney (left arrow). (Image from Nasser Institute Hospital Cairo, Egypt)

Discussion

Disease of the renal artery is a well-known cause of secondary hypertension and it is also increasingly being recognized as the cause of renal failure in the elderly due to a condition known as ischaemic nephropathy[4].

Overall the most common cause is atheromatous narrowing which is seen in older patients and is frequently associated with degenerative vascular disease[1]. The index case being a young (28 years old) woman with right kidney affectation makes the diagnosis of fibromuscular dysplasia more likely since it is the commonest cause in this age.

Fibromuscular dysplasia (FMD) is the congenital band of fibrous tissue around an artery, which as the patient grows, causes progressive narrowing of the vessel[5].

Stenosis in RAS is usually classified as ostial, proximal or distal according to the part of the vessel affected, and is quantified in terms of the degree of narrowing[3]. Stenosis of less than 50% is not usually considered hemodynamically significant[6]. Arteriosclerotic renal artery disease frequently involves proximal 2cm of the main renal artery usually demonstrated on angiography as eccentric stenosis in proximal 2cm of renal artery and involving the orifice mostly. Fibromuscular dysplasia on the other hand is seen mainly in the mid and distal main renal artery with proximal 3rd of the main renal artery being spared in 98% of cases as seen on angiography[3]. The area of affectation in this case which involved almost the entire main renal artery is however atypical. There is post stenotic dilatation in the angiography of the patient which is seen in the two types of renal artery stenosis and it's seen distal to the narrowed area[4].

Patients with RAS usually present with abdominal or flank pain, hematuria, hypertension, oliguria or anuria, low urine sodium concentration and normal or decrease renal size (greater than 2cm)[5]. This patient however presented with renovascular hypertension.

The patient also presented with classical radiological features of renal artery stenosis (RAS) which include normal or decreased renal size (67cm in this patient). Some patients present with features of ischemic nephropathy whereby the kidneys will be echogenic, shrunken but smooth in contour with loss of the normal corticomedullary differentiation[3]. In this case the patient has shrunken kidney with grade I renal parenchymal disease.

Doppler ultrasound usually demonstrates RAS as increased peak systolic velocity of >150cm/sec for angles $<60^0$ or 180cm/sec for angles >70 with measurement taken at the stenosed site[7]. However false positivity may be due to suboptimal Doppler angles. There may also be a post stenotic spectral broadening with or without flow reversal. Absence of flow during diastole in those with more than 50% stenosis is usually seen. Indirect sign include tardusparvus pattern which is seen as a result of slowing and attenuation of Doppler wave form during diastole in regions distal to the stenosis[3,7,8].

Intravenous urography has a 60% true positive rate in demonstrating the radiological features of RAS[3]. Delayed wash out of contrast medium, lack of distension of collecting system and generalized attenuation of contrast density are some of the IVU findings reported[9,3]. This patient's IVU shows a delay in appearance (nephogram phase) of contrast. Increase concentration of contrast medium is usually seen as a result of reabsorption of water by the ischaemic kidney but not in this case[3]. There was a slight delay in excretion of contrast medium on the

affected side when compared with the normal side at the early films because of the reduced blood flow.

Conventional angiography, Computed Tomography (CT) and Magnetic resonance angiography (MRA) are some of the very important diagnostic tools for RAS. However, conventional angiography is the gold standard[3] followed by digital subtraction angiography. This patient's angiography demonstrated stenosis of the main right renal artery with post stenotic dilatation.

This is as oppose to RAS due to arteriosclerosis which usually demonstrate eccentric stenosis in the ostia and proximal 2cm of the renal artery[1, 10]. The computed tomography showed the shrunken right kidney with stenosed vessel. MRI was however not done without any reason being given. It has the advantage of multiplanar imaging and is better for all soft tissues imaging.

Radionuclide study has 44% sensitivity and 95% specificity in diagnosing RAS.
It showed markedly reduced perfusion of the patient's right kidney.

Poor visualization or non-visualization on delayed images are some of the findings on scintigraphy.

Management of renal artery stenosis is divided into medical, radiological/surgical treatments.

The medical treatment includes the use of antihypertensive therapy, low dose aspirin and lipid lowering drugs if appropriate[5]. Renal artery angioplasty has 60% success rate for non-ostial lesions. Resections of the diseased segment with end to end anastomosis or replacement with a vein graft are some of the other treatment options. Transluminal balloon angioplasty has a success rate of 90% with very low re-stenosis rate[11].

This patient had medical management where she was placed on antihypertensive drugs.
Patient with arteriosclerotic renal artery stenosis (RAS) in about 40-45% undergo progression of the atherosclerotic lesion leading to renal atrophy, arterial occlusion and eventual ischaemic renal failure. Fibromuscular dysplasias on the other hand undergo progression of the lesion in 20% of cases causing decline in renal function[3].

Some of the complication of RAS include azotemia in case of arteriosclerotic type especially in bilateral or compromised contralateral renal function. Giant aneurysm and arteriovenous fistula formation between renal artery and vein are some of the complication encountered in fibromuscular dysplasia RAS.[3]Radiological investigations are key to diagnosis of Renal Artery Stenosis (RAS) and where available conventional CT angiography isparamount.