

Spontaneous renal artery dissection: Current perspective

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Abstract

Spontaneous Renal artery dissection is a rare entity that may remain clinically silent or present with non-specific signs and symptoms, which makes it a diagnostic challenge. It may be associated with certain underlying vascular pathologies, but its occurrence remains idiopathic in the majority of cases. While there are no evidence-based guidelines for its management, blood pressure control and preservation of renal function remain the cardinal goals of therapy.

Keywords: Renal artery, Dissection, Review, Stent, SRAD.

Introduction

Since the first report in 1944 and the first angiographic description in 1956, almost 200 cases of Spontaneous Renal Artery Dissection (SRAD) have been reported in medical literature, with a quarter of them diagnosed at autopsy.¹ Smith et al have classified the clinical presentation of renal artery dissection as acute or chronic; acute is further classified into spontaneous, iatrogenic (guidewire, catheter, angioplasty balloon) and agonal (sepsis, malignancy, stroke, chronic renal failure, cirrhosis) while chronic is classified as functional and silent.² Out of all artery dissections, renal artery dissection accounts for only 1-2% of cases, and in a normal healthy individual with no predisposing factors, renal artery dissection occurring spontaneously is an even rarer phenomenon.³ Prior to 1980, due to the nonspecific nature of SRAD's clinical manifestations, the diagnosis was almost always made on autopsy and only rarely by angiography. With advances in imaging techniques such as computed tomography (CT), cases of SRAD became more readily identifiable.⁴ However, the management and long term follow-up protocols still remain controversial.

Etiology:

SRAD usually occurs in individuals during their fourth to sixth decades of life, with the male to female

ratio being 4:1.^{1,5,6} It has no predilection for either side and in 10-15% of cases it may be bilateral.^{5,7-9} Spontaneous renal dissection may be accompanied by dissection of other peripheral arteries.¹⁰

The recognized risk factors for SRAD include malignant hypertension, severe atherosclerosis, Marfan syndrome, fibromuscular dysplasia, cystic medial necrosis, Ehlers-Danlos syndrome, subadventitial angioma, syphilitic arteritis, polyarteritis nodosa, positive anti-phospholipid antibody, tuberculosis and extremes of physical exertion.¹¹⁻¹³ In most cases of SRAD, however, no such associations are found; cases of otherwise completely healthy individuals presenting with SRAD have been reported in literature.¹⁴⁻¹⁷

In addition to the afore-mentioned risk factors, there are also certain pathophysiological factors known to influence the risks of development of SRAD, including blood pressure, heart rate, sympathetic activity, basal vascular tone, circadian rhythms, plasma viscosity, endogenous vasoconstrictor hormones, platelet aggregability, haematocrit as well as drugs like cocaine and amphetamine.^{3,18,19}

Although the exact pathogenesis of SRAD remains to be explained, the ultimate event is understood to be either a haemorrhage in the vessel wall through bleeding vasa vasora, extension of a primary intimal tear or endothelial dysfunction.^{12,13,16,20}

Clinical Presentation:

Depending on the degree of vascular occlusion as a result of the dissection, presentation can range from a clinically silent phenomenon to a full-blown picture of renal failure. As renal artery dissection can be mild and resolve spontaneously, it is likely that the number of reported cases of SRAD is an under-estimation of its true incidence.

SRAD can present clinically with severe hypertension, haematuria, proteinuria, flank pain, abdominal bruits and/or acute renal failure.²¹ The most

common symptom of SRAD is unilateral flank pain radiating to the epigastrium (reported in as many as 92% of the patients in one retrospective study)⁵ which if accompanied by haematuria can erroneously point towards a diagnosis of urolithiasis. The most common sign on clinical examination is hypertension;² one study reported that 40% of patients with pre-existing hypertension had a rise in blood pressure.²² Because of this vague clinical presentation, SRAD can only be confirmed by renal imaging.^{1,5}

Diagnostic Procedures:

Multiple imaging modalities including computed tomography (CT), magnetic resonance imaging (MRI) with gadolinium contrast, CT angiogram, intravenous pyelogram (IVP) and intravascular ultrasound (IVUS) can be used to visualize the dissection or an area of renal hypoperfusion.

A CT scan can be used early on if there is suspicion of SRAD because it can show an area of infarction even if the dissection is not yet apparent, thus pointing towards a vascular pathology.⁴ An example of that is shown in Figure-1.

Rarely if CT scan fails to visualize the renal infarct, we can use Diffusion-Weighted Magnetic Resonance Imaging. DWI can easily detect ischaemic areas early and show the infarct as high-intensity areas.²³

The gold standard for the past six decades has been the angiogram, which not only enables the extent

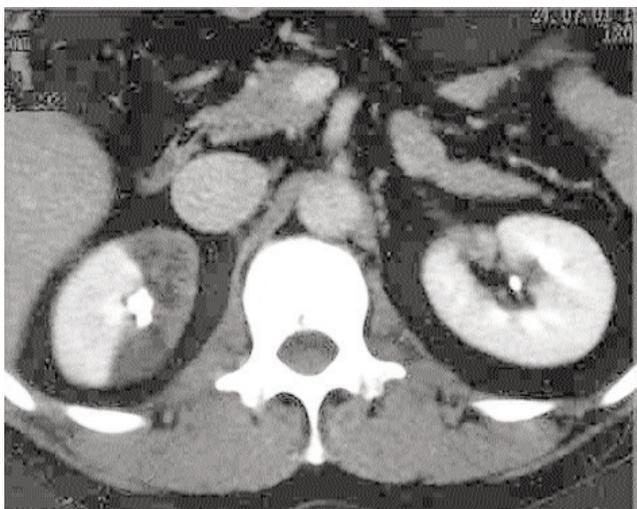


Figure-1: A CT Scan without contrast done on a 47 year old patient who presented to the emergency room with flank pain. The CT scan showed hypoperfusion of the superior segment of the right kidney thus pointing towards a vascular pathology (Reprinted by permission of Reference. 3: E. Guérin, E. Vandueren, E. Dubois, C. Delcour, J. C. Wautrecht, G. Verhelst. Spontaneous Renal Artery Dissection in an Otherwise Healthy Male. *Acta chir Belg*, 2006, 106, 703-706).

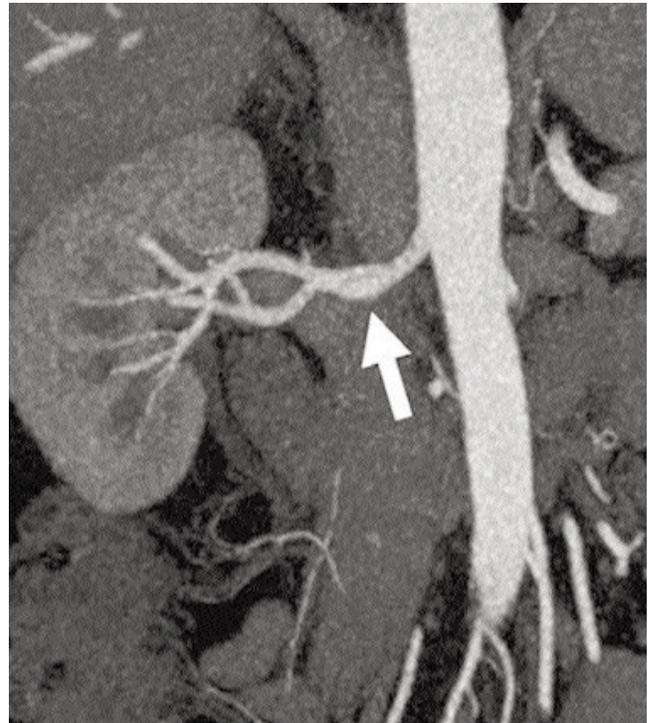


Figure-2: CT angiography showing double lumens at the distal truncal right main renal artery (arrow) and 1-cm long focal renal artery dissection. (Reprinted by permission of Reference. 18: Pernicioglu B, Piskinkaya S, Ozer C, et al. Isolated Spontaneous renal artery dissection: diagnosis and endovascular management. *Diagn Interv Radiol* 2011; 17: 101-104.).

and nature of the dissection to be precisely visualized but also provides an avenue for therapeutic intervention (Figure-2).⁵

Intravascular ultrasound (IVUS) has also been used in some cases. It can be particularly helpful in guiding accurate stent placement and in distinguishing between true and false lumens (Figure-3).¹⁹

Management:

Treatment options vary and may be selected on the basis of the severity of a patient's condition as well as the specific co-morbidities. In general, a conservative approach is favoured for mild cases, while surgical modalities are reserved for more complex scenarios.

Non-Surgical Management:

Non-surgical management aims on achieving optimum blood pressure control and adequate renal function through anti-hypertensives and anti-coagulation.

For blood pressure control, calcium channel blockers, angiotensin converting enzyme inhibitors and ACE inhibitors have been successfully used. In many

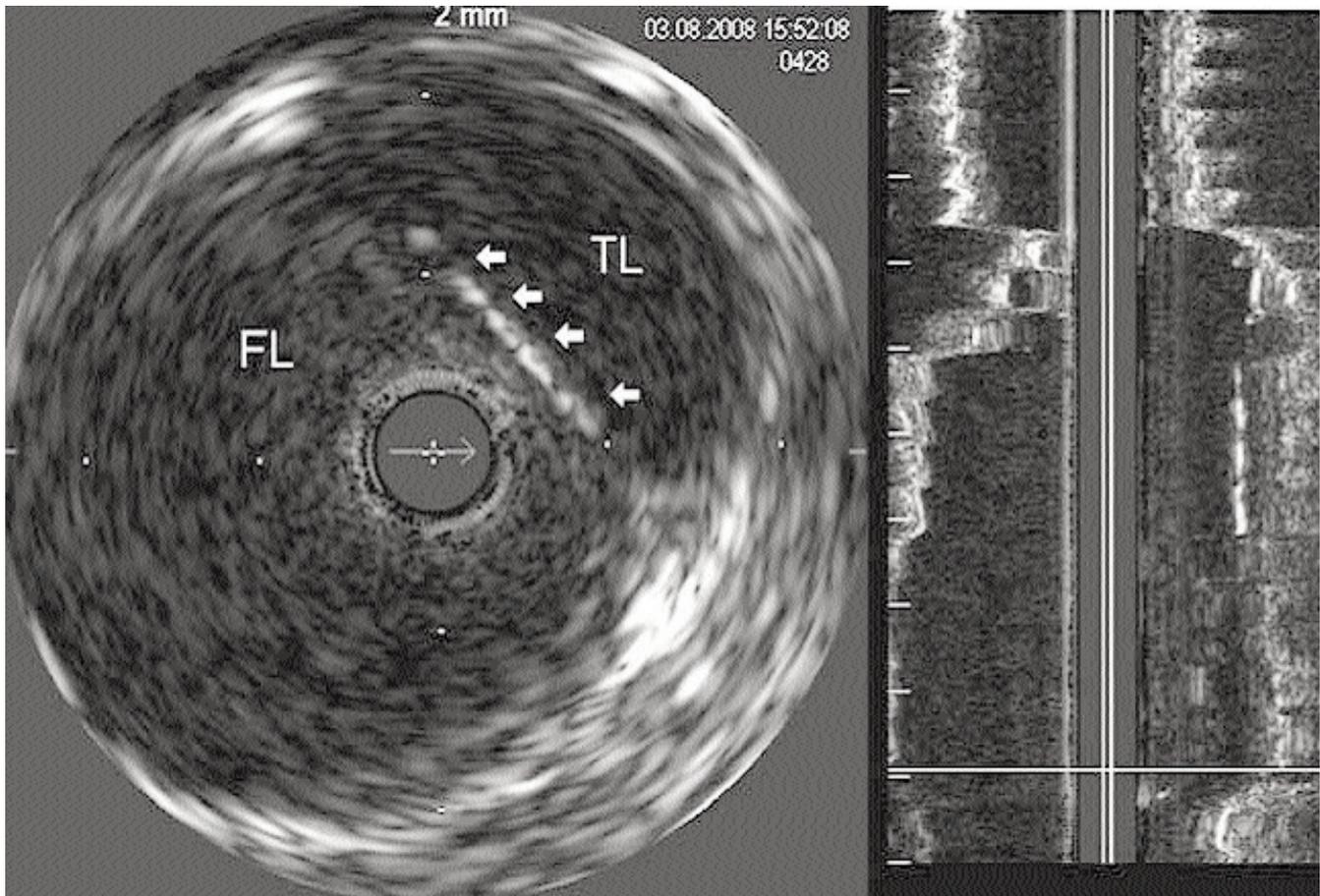


Figure-3: Intravascular ultrasound image showing the caudal lumen (FL, falselumen) suggested by absence of intimal echogenic reflection and revealing the intimal flap with a single layer of bright intimal reflection (arrows) only at the side of the cranial true lumen (TL). (Reprinted by permission of Reference. 18: Pernicioglu B, Piskinkaya S, Ozer C, et al. Isolated Spontaneous renal artery dissection: diagnosis and endovascular management. *Diagn Interv Radiol* 2011; 17:101-104.)

cases, a multi-drug regimen may be necessary for optimizing blood pressure control.^{21,23,24}

Anti-coagulation was first used in SRAD, based on its proven success in resolving small carotid artery dissections over a period of several months.³ Anti-coagulation is aimed at pre-empting the development of thrombosis at the site of endothelial injury. A continuous intravenous infusion of heparin at a dose of 25000UI/day has been typically used. After a week-long therapy with intravenous heparin, the patient can be switched to long-term oral anti-vitamin K therapy.³

If serial angiography is performed and indicates stability of the dissection with no worsening in renal function, a conservative approach may be appropriate.¹⁶ However, follow-up in some cases has shown failure of the condition to stabilize in the long term.^{20,25} In case a conservative approach fails or there is acute deterioration in renal function, urgent surgical intervention may be necessary.⁵

Surgical Management:

Various surgical procedures can be employed but there is little evidence guiding their relative merits, demerits and indications.

Usually, vascular reconstruction via stenting or coiling is considered if there is enough residual renal function. Various case reports demonstrate successful return of optimum renal blood flow along with achievement of blood pressure control after stenting.^{17,25-29} One study showed that out of 22 patients who underwent revascularization procedures, 90% reported significant improvement in blood pressure control in the short-term, but up to 35% required either a partial (9%) or total (26%) nephrectomy eventually. In contrast to this according to one study 16 patients were treated with endovascular stent placement for SRAD and followed up for a mean duration of 8.6 ± 3.4 years. Seven of these patients did not need any antihypertensive medication while nine had to use single or multiple anti-

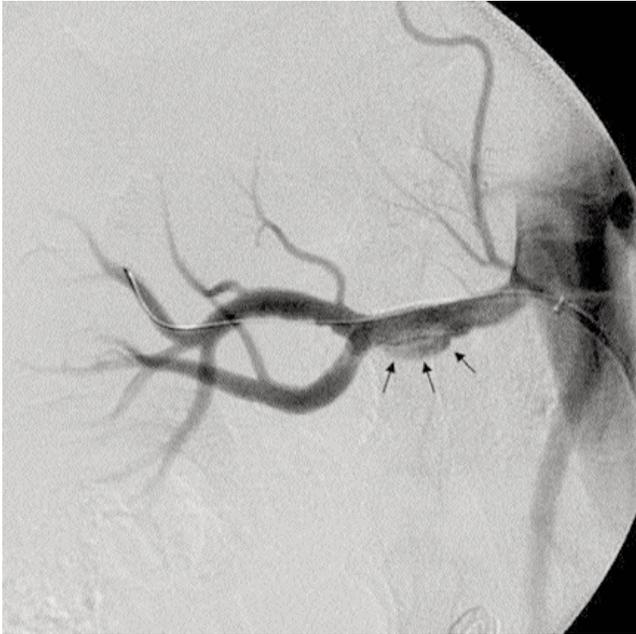


Figure-4: Digital subtraction angiography image after the stent placement showing a patent cranial true lumen with compressed false lumen just below the stent only partially filled with contrast medium (arrows). (Reprinted by permission of Reference. 18: Pernicioglu B, Piskinkaya S, Ozer C, et al. Isolated Spontaneous renal artery dissection: diagnosis and endovascular management. *DiagnIntervRadiol* 2011; 17:101-104.).

hypertensives but none of the 16 patients had restenosis on repeated imaging.³⁰ Figure-4 illustrates angiographic stent placement in one patient.

Nephrectomy is preferred over vascular reconstruction procedures if the kidney cannot be saved due to size of the infarct, severely compromised renal function on isotope renography or if the revascularization would be extremely tough due to involvement of a branch artery.¹⁹ One can choose partial over total nephrectomy if the infarct size is small. For patients with bilateral lesions, preoperative blood urea nitrogen, serum creatinine and renal isotope scanning is used to decide whether residual renal function would be sufficient enough after unilateral nephrectomy.

In cases where the lesion is progressively worsening, especially if the hypertension remains uncontrolled despite multi-drug treatment, another approach that could be used is segmental embolization. Mudrik et al have reported success in halting the evolving dissection through this technique.⁵

Finally, skilled vascular surgeons can also operatively repair the dissection. This can be done either in situ or ex-vivo, and requires expertise. In situ repair is more feasible if the dissection does not involve the main renal artery and ex-vivo repair is more practical in

dissections involving the main renal artery. Although this procedure can achieve satisfactory results, a number of post-operative complications including fibrotic stenosis, thrombosis, partial atrophy and renal failure can ensue.²⁰

Outcome and Complications:

The prognosis of SRAD remains poorly defined due to dearth of sufficient data, which in turn stems from a low incidence, frequently silent clinical presentation and inadequate longitudinal follow-up to date. The main long-term complication of SRAD is recalcitrant hypertension, which requires lifelong therapy with multiple antihypertensive agents. Approximately 30-40% of patients do require chronic management of hypertension in the long run. The main cause of mortality in patients with SRAD is renal failure. The chances of SRAD being fatal have been shown to increase with bilateral lesions.^{1,2}

There are also some documented cases of SRAD leading to a spontaneous resolution. Such a phenomenon can be putatively explained by re-entry of the false lumen back into the true lumen or due to a complete obliteration of the dissected segment by thrombosis and organization.⁹

Arterial rupture as a complication of the dissection is fortunately rare but may be fatal. Two cases have been reported where such a dissection did not result in mortality.⁶

Conclusion

Isolated SRAD is a rare phenomenon and often presents as a diagnostic and therapeutic challenge. Although advanced imaging modalities like CT are helpful in making the first diagnosis, angiography remains the definitive study and also enables concurrent therapeutic intervention. The short and long-term management of patients with this condition is poorly understood and controversial but anti-coagulation and adequate blood pressure control is the cornerstone of management in all cases. In complex or recalcitrant cases, various angiographic and surgical interventions have been tried and have produced mixed results. Long-term follow-up is required in order to control hypertension and prevent renal compromise.

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