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Case report

A Case of Spontaneous Bilateral Renal Artery Dissection

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Abstract

Introduction. Spontaneous renal artery dissection (SRAD) is defined as dissection of the renal artery without an underlying vascular disease, intervention or trauma. Approximately 200 cases of SRAD have been reported in the literature and its bilateral involvement is much rarer. It is a rare condition and its incidence and clinical findings are not well known. Most common finding is hypertension, and most common symptoms are sudden onset waist or flank pain and hematuria. It is difficult to diagnose as it can often be confused with renal colic or pyelonephritis. Patients who do not undergo computed tomography (CT) are often treated as urolithiasis.

Case presentation. We would like to present here a 38-year old male patient who has bilateral SRAD and has difficulties in diagnosis and treatment.

Conclusion. CT is important in the diagnosis of renal infarction and dissection in cases with flank pain of unknown origin, and CT evaluation should always be considered in cases without concomitant diseases. Anticoagulant and antihypertensive therapy, endovascular procedures, open vascular surgery and nephrectomy are effective treatment options. Although there is no consensus on the choice of treatment options. The diagnostic dilemma and uncertainty of the treatment management in such cases can only be resolved by presenting more case experiences in the literature.

Keywords: Renal artery dissection, renal infarction, renovascular disease, flank pain, hematuria

Introduction

Spontaneous renal artery dissection (SRAD) is defined as dissection of the renal artery without an underlying vascular disease, intervention or trauma. It is a rare condition and its incidence and clinical findings are not well known [1,2]. SRAD was first described in the literature in 1944 [3]. It constitutes 1-2 % of arterial dissections [4,5].

Most common finding is hypertension, and most common symptoms are sudden onset waist or flank pain and hematuria. It is difficult to diagnose as it can often be confused with renal colic or pyelonephritis. It has three different presentations. These are subacute state without serious progression, renal infarction state due to acute occlusion, and chronic state with renovascular hypertension. It is difficult to detect dissection with conventional methods such as ultrasonography or doppler ultrasonography. Patients who do not undergo computed tomography (CT) are often treated as urolithiasis [3,6]. CT, intravenous pyelography, or magnetic resonance imaging may be helpful in diagnosis; but the gold standard diagnostic method is angiography [1].

Anticoagulant and antihypertensive therapy, endovascular procedures, open vascular surgery and nephrectomy are effective treatment options. Although there is no consensus on the choice of treatment options, the general opinion is that an interventional procedure should be planned in cases of failure to control blood pressure with medical treatment or progressive loss of renal function [7].

We here would like to present a case who has bilateral involvement (which is rare) and has difficulties in diagnosis and treatment.

Case Presentation

A 38-year-old male patient who had no previous medical history went to a local hospital's emergency department following a sudden onset right flank pain and nausea. In physical examination he was nonfebrile and normotensive. Except for lactate dehydrogenase (LDH) elevation, complete blood count, kidney and liver function tests and complete urinalysis with microscopy were normal.

A contrast enhanced computed tomography (CT) scan showed right renal parenchymal multifocal infarction and acute occlusion underlining dissection of segmen-

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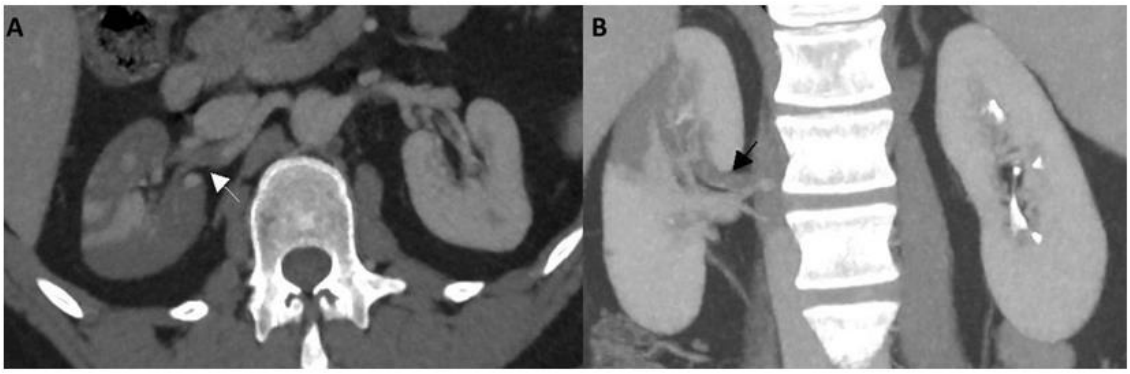

Fig. 1. Axial (A) and coronal (B) abdomen BT images show wedged shaped hypoattenuation of renal parenchyma due to ischemia and acute occlusion underlining dissection of segmental renal artery (Arrows in A and in B)

tal branch of renal artery that supply superolateral part of right kidney (Figure 1).

Table 1. Patient's first biochemical and hematologic tests

Patient's first biochemical and hematologic tests	Results	Referance range
White blood cell count	13500/mm³	4000-10000
Serum creatinine	1.34 mg/dL	0.75-1.35
Blood urea nitrogen	30 mg/dL	6-24
Lactate dehydrogenase	1775 U/L	105-233
Urine nitrite	Negative	Negative
Urine protein	2+	Negative
Urine erythrocyte	1	0-3
Urine leukocyte	1	0-5
Antithrombin III activity	93.4 %	80-120
Protein C activity	63.8 %	70-140
Protein S activity	110.1 %	60-130
C3 complement	125 mg/dL	75-135
C4 complement	45.7 mg/dL	12-72
Fibrinogen	859 mg/dL	170-420

He was referred to a tertiary center and he was consulted to hematology department in terms of conditions that may predispose to thrombosis. Antithrombin 3, protein S antigen, C3, C4 levels were normal. Protein C activity was slightly low (63,8 %, N: 70-140 %), fibrinojen was elevated (859 mg/dl, N: 170-420 mg/dl). In echocardiogram his ejection fraction was 65 % and there was no intracardiac thrombosis. Oral vitamin K antagonist therapy was started to the patient and he was discharged. He was asymptomatic and monthly follow-ups continued.

6 months later he had left flank pain that woke him up at night. When he admitted to the hospital, his blood pressure was 147/86 mm Hg, heart rate was 110/min, respiratory rate was 22/min and he was afebrile. His laboratory values were white blood cell 13500/mm³, serum creatinine 1.34 mg/dl, blood urea nitrogen 30 mg/dl, LDH 1775 U/L. His liver functions were normal and urinalysis with microscopy showed nitrite negative, 2+ protein, 1 erythrocyte and 1 leukocyte (Table 1). He was consulted to infectious diseases and infective endocarditis was suspected. He was started empirical ceftriaxone 1x2 gr/day treatment. In his transesophageal echocardiogram there was no intracardiac vegetation or thrombosis. There was no microorganism in his blood and urine cultures.

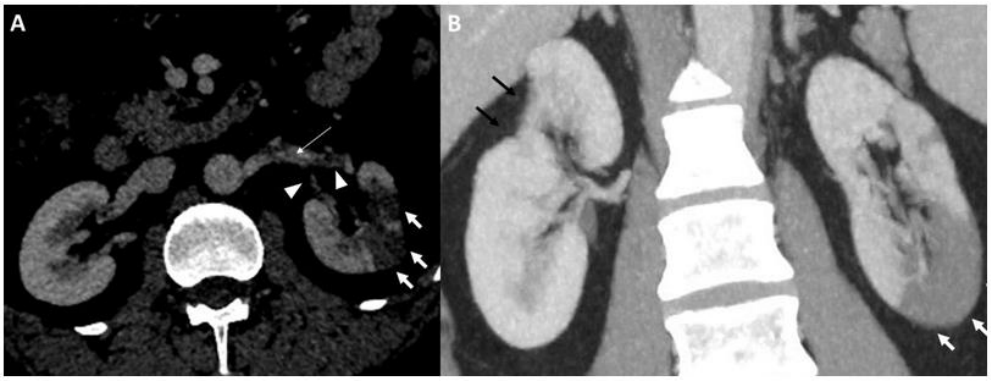

Fig. 2. Axial (A) and coronal (B) abdomen BT images show wedged shaped hypoattenuation of renal parenchyma due to ischemia (thick white arrows in A and B), left renal artery dissection with intimal flap (thin arrow in A) and thrombosis of segmental branches (arrowheads in A). Also, note that there is right parenchymal scarring (black arrows in B) due to previous episode

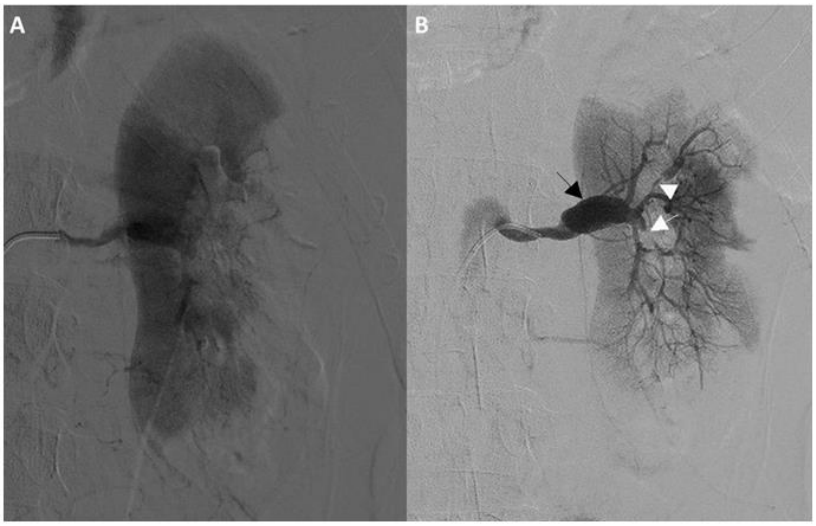

Fig. 3. A) Renal DSA image showing contrast filling defect in peripheral inferolateral part of the left kidney due to ischemia. B) Renal DSA image showing fusiform dilatation of false lumen (black arrow) due to known renal artery dissection, occlusion of segmental branch (white arrow), involvement of interlobar artery (arrowhead)

The patient underwent abdomen CT scan that showed wedged shaped renal parenchymal ischemia and left renal artery dissection and thrombosis of segmental renal artery branches which could be related to the underlying dissection (Figure 2). Ten days after the CT scan, the patient underwent renal digital substraction angiography (DSA) to better delineate renal vasculature. Renal DSA showed parenchymal filing defect due to ischemia, fusiform dilatation of distal renal artery and its segmental branches, and occlusion of inferolateral segmental branch after fusiform dilatation (Figure 3). No interventional procedure was recommended to the patient.

He was consulted to rheumatology department for etiological evaluation and his rheumatological markers were all negative (Table 2). Cranial CT showed no intracranial aneurysm. He was consulted to hematology department for anticoagulant therapy. His oral vitamin K antagonist therapy was switched to novel oral anticoagulants for being more efficient for intraabdominal thrombosis.

Table 2. Patient's rheumatologic and hematologic tests

Rheumatologic and hematologic tests	Results	Referance range
Antithrombin III activity	83.3 %	80-120
Protein C activity	93.4 %	70-140
Protein S activity	103.4 %	60-130
LA verification	33.4 min	30-38
ANA (anti-nuclear antibody)	Negative	Negative
Anti dsDNA	Negative	Negative
ANCA (IFA)	Negative	Negative
ANCA profile (elisa)	Negative	Negative
C3 complement	115 mg/dL	75-135
C4 complement	35.4 mg/dL	12-72
Anti cardiolipine Ig M, Ig G	Negative	Negative
Beta-2 glycoprotein-1 Ig M, IgG	Negative	Negative
Homocysteine	6.0 µmol/L	5-15
PAI-I 4G/5G mutation	Heterozygous	Negative
MTHFR677C mutation	Heterozygous	Negative

As comprehensive investigations for etiology were inconclusive, the patient was considered to have spontaneous bilateral renal artery dissection. His blood pressure was controlled with ramipril 2,5 mg/day. He is currently under regular follow-up, his blood pressure is normal, his renal functions are stable, and no new renal artery embolism has developed in the last 2 years.

Discussion

This patient with renal infarction due to renal artery dissection had no underlying comorbid conditions and had bilateral involvement. As spontaneous renal artery dissection related renal infarction is a rare entity, it obviously created some difficulty in the diagnosis and management. Without a prompt diagnosis and an appropriate follow-up and anticoagulation strategy, he may have some adverse consequences.

There are many causes of renal infarction. The most common cause is cardiogenic thromboembolism. Atrial fibrillation, cardiomyopathy, valvular heart diseases and infective endocarditis are some of the underlying causes [8]. Rare underlying causes are trauma, angiographic procedures, thrombophilic diseases and hematological diseases [9].

In renal artery dissection, which is one of the rare conditions that may lead to renal infarction, fibromuscular dysplasia (FMD), malignant hypertension, severe atherosclerosis, Marfan Syndrome, Ehler Danlos Syndrome, heavy physical activity and cocaine use may be the underlying causes [2,7,10].

Three mechanisms are thought to be effective in the pathogenesis of renal artery dissection. These are shear stress, vaso vasorum rupture and segmental arteriolar mediolysis. Connective tissue diseases, heavy physical activity, trauma, cocaine use and uncontrolled hypertension cause dissection with shear stress. FMD and

mid-vessel vasculitis lead to dissection by causing vasa vasorum rupture and intramural hematoma [11].
In some cases, no etiological cause could be found and these cases were defined as SRAD. Although SRAD is a rare cause of renal infarction, it was shown that 21 (4,79%) of 438 patients with renal infarction had SRAD in the study by Oh *et al*. [8]. In the case series by Yoon *et al*., of 35 patients with renal infarction, SRAD was detected in 6 patients (17,1 %). SRAD was the second most common cause of renal infarction in these series. It has been suggested that this incidence is high because high spatial resolution of current multidetector CT (MDCT) was used in these case series. Therefore, these results may more accurately reflect the incidence of SRAD [5].
More than 200 cases of SRAD have been reported in the literature, one quarter of which are presented in autopsies [12]. It is more common in men than women, and prevalence ratios between 4:1 and 10:1 were reported in the literature. It is mostly seen in the 4th and 6th decades of life [13,14]. Bilateral involvement has been seen in 10-15 % of SRAD cases [7].
The underlying mechanism in SRAD cases is unknown. One hypothesis is that physical exercise causes tension in the arterial wall in these cases, and the other hypothesis is that SRAD cases are a clinical variant of FMD or atherosclerosis [15]. However, in the 6-patient SRAD case series of Renaud et al. no cardiovascular risk factors suggestive of arterial dysplasia or atherosclerosis were found.
The clinical presentation of our case is mainly consistent with previous cases. SRAD usually presents in fit and 30- or 40-year-old males with an abdominal, flank or low back pain [10]. Hypertension is seen in almost all patients with SRAD [1]. Our case was observed to be hypertensive at the time of admission. It has been determined that the incidence of new-onset hypertension in patients with SRAD is higher than in patients with embolism-related renal infarction [5]. Blood tests often show a raised white cell count and renal functions may deteriorate acutely with an elevation of lactate dehydrogenase. Urinalysis sometimes reveals microscopic hematuria, but in our case urinalysis was normal.
There are many options for treatment, from conservative treatment to endovascular and surgical intervenetions. Although there is no consensus on treatment, endo\vascular revascularization is often recommended in cases of hypertension that cannot be controlled with medical treatment or progressive renal dysfunction [7]. Antihypertensive treatment and anticoagulant treatment are medical treatment options to prevent further thrombus formation after endothelial damage. Most of the experience published so far has been with vitamin K antagonists. The optimal duration of anticoagulation is unclear and there is a lack of data on the use of the oral anticoagulants [10].

Conclusion

In conclusion, CT is important in the diagnosis of renal infarction and dissection in cases with flank pain of unknown origin, and CT evaluation should always be considered in cases without concomitant diseases. The diagnostic dilemma and uncertainty of the treatment management in such cases can only be resolved by presenting more case experiences in the literature.

Conflict of interest statement. None declared.

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