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Segmental arterial mediolysis presenting as spontaneous bilateral renal artery dissection

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ABSTRACT

The commonest site of primary dissection involving the visceral vessels is renal arteries; however, spontaneous bilateral renal artery dissection is an extremely rare entity. Spontaneous renal artery dissection (SRAD) is rarely a cause of renovascular hypertension. Segmental arterial mediolysis is a rare arteriopathy of unknown etiology which is a nonatherosclerotic and noninflammatory condition. We report a case of a 51-year-old male patient with spontaneous dissection of bilateral renal arteries with clinical, laboratory, and angiographic findings consistent with segmental artery mediolysis. Early diagnosis and treatment of this condition will decrease morbidity and mortality.

Introduction

The commonest site of primary dissection involving the visceral vessels is renal arteries; however, spontaneous bilateral renal artery dissection is an extremely rare entity [1]. Spontaneous renal artery dissection (SRAD) is rarely a cause of renovascular hypertension [2]. The incidence of SRAD is unknown and original reports were based on autopsy findings. A quarter of the published reports are single case reports or reports of small series. A few cases of spontaneous dissection affecting the visceral, peripheral, and intracranial arteries have been reported [3]. The underlying etiology of dissection is not clearly known. However, fibromuscular dysplasia (FMD), segmental arterial mediolysis (SAM), and connective tissue disorders are known to predispose spontaneous dissection. We report a case of a 51-year-old male patient with spontaneous dissection of bilateral renal arteries with clinical, laboratory, and angiographic findings consistent with segmental artery mediolysis. SAM is a rare arteriopathy of unknown etiology which is a nonatherosclerotic and noninflammatory condition [4]. It was first described by Slavin et al. in 1976 as autopsy diagnosis [5]. Since then a few case reports have been published. Early diagnosis and treatment of this condition will decrease morbidity and mortality.

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Case report

A 51-year-old man with significant medical history of smoking, hyperlipidemia, and hypertension presented to our hospital with an acute onset right flank pain. He denied nausea, vomiting, or hematuria. A week prior to this admission, he presented to an outside hospital with a similar episode involving the left flank and was evaluated as presumed diverticulitis. On further investigation, computed tomography of the abdomen showed an infract involving the lower pole of the left kidney. He was treated with anticoagulation. At our facility, diagnostic workup including complete blood count, urinalysis, complete metabolic profile, echocardiography, and coagulation workup revealed no abnormality. Laboratory markers of inflammatory vasculitis (including erythrocyte sedimentation rate, antinuclear antibodies, and antineutrophil cytoplasmic antibodies) were negative. Radiological workup with ultrasound of the kidneys demonstrated hypoechoic cortex, with avascularity in the lower pole of left kidney on color flow imaging consistent with an infarct (Fig. 1). Magnetic resonance angiography (MRA) revealed an irregular beaded appearance with focal dilatations of left main renal artery and a lower pole infarct. There were 2 right renal arteries—a main renal artery and an inferior accessory renal artery arising from the aorta. The mid portion of right main renal artery showed fusiform dilatation with diffuse narrowing affecting its distal segment and the branches. Inferior accessory showed diffuse narrowing affecting its distal segment with nonvisualized parenchymal arteries (Fig. 2). To further evaluate the renal arterial pathology, catheter angiography of the renal arteries were performed. This study demonstrated segmental dilatation of left artery with irregular outline. This represented focal dissection with dilated false lumen. The lower pole renal artery was seen proximally with diffuse tapered distal segment (Fig. 3). Angiogram of right main renal artery showed fusiform dilatation of the mid segment. Angiogram of the accessory right renal artery demonstrated diffuse wavelike configuration to the distal segment and the branching vessels. These findings were consistent with dissection of the accessory right renal artery and its branches (Fig. 4). Considering the age, male predilection, negative laboratory workup for inflammatory vasculitis, and the radiological findings of dissection, we diagnosed this case as SAM affecting both renal arteries.

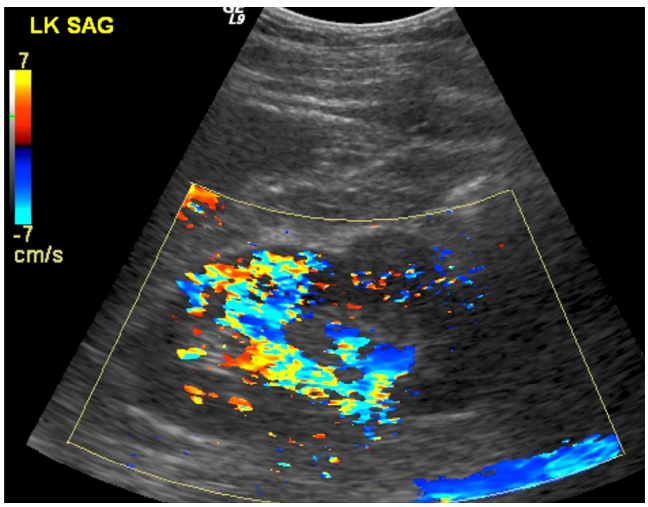

Fig. 1 – Color Doppler ultrasound shows lack of vascularity within the hypoechoic region in the middle lower pole of the left kidney.

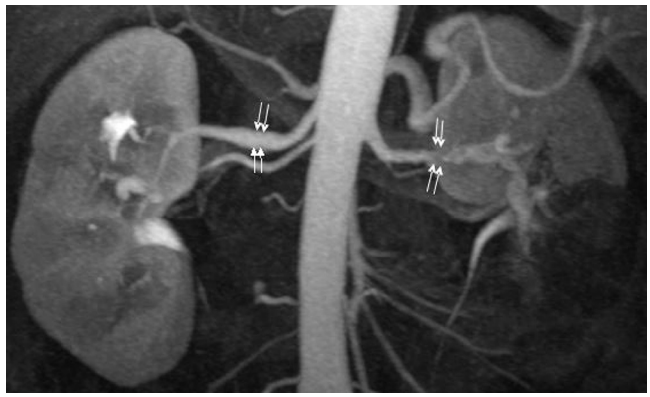

Fig. 2 – Contrast-enhanced magnetic resonance angiography shows focal diffuse dilatation of right main renal artery, distal tapering of inferior accessory right renal artery, and irregularity and dissection of left main renal artery.

Discussion

SAM is a nonatherosclerotic, noninflammatory arteriopathy. The etiology and disease progression are not yet fully known.

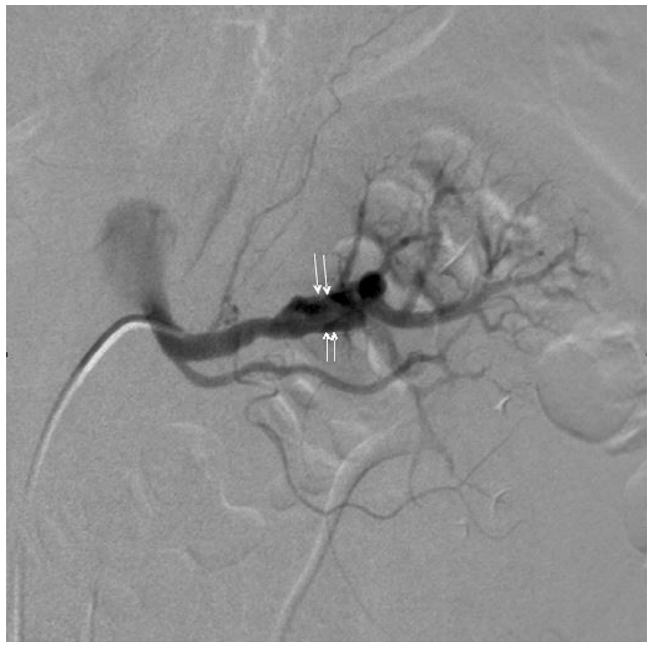

Fig. 3 – Left renal angiography shows focal dissection of left main renal artery with dilated false lumen. Also note diffuse narrowing of lower pole branch vessel secondary to dissection extending into the lower pole branch resulting in an infarct seen on magnetic resonance imaging.

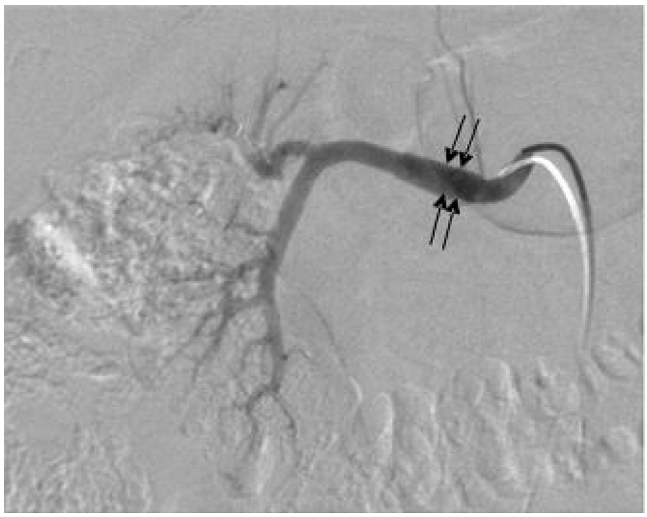

Fig. 4 – Right renal arteriogram shows diffuse fusiform dilation of mid segment of right main renal artery.

It is most commonly seen affecting the medium-sized muscular arteries (celiac, mesenteric, and renal arteries) of the abdomen [6]. A few cases affecting the cerebral and pulmonary arteries have been reported [6,7]. The first case of SAM was described by Slavin et al. in 1976. Gruenwald described similar morphology in epicardial coronary arteries of newborn infants in 1949 [8]. This condition was initially coined as segmental mediolytic arteritis; however, as there was lack of evidence to suggest an inflammatory etiology in both clinical and laboratory data, it was later changed to SAM, describing its pathologic findings as it involves skip lesions in segmental fashion and lysis of the outer medial layer of the artery [9]. Recent literature from Slavin et al. proposed the etiology as vasospastic phenomenon which favors the morphology and structural features [9,10]. Similar findings of SAM were demonstrated when ractopamine (beta adrenergic agonist) was administered in dogs [11]. This also suggested the possibility of norepinephrine-mediated vascular structural changes. Recent data suggest SAM as a disorder of peripheral sympathetic nervous system and alpha 1 adrenergic receptor. However, more studies and data are needed to support this hypothesis. Some studies proposed the role of endothelin –1 receptors as they are demonstrated in the adventitia and granulation tissue during the reparative phase of the disease [12]. The disease occurs in 2 phases—injury phase and reparative phase [13]. Both mediolysis and arterial tear that separates the adventitial layer from outer medial muscular wall occur during the injury phase. The most common radiological presentations on the angiography are arterial dilatation, single or multiple aneurysms giving an appearance of string of beads, dissecting hematomas, stenosis of the involved arteries, and occlusion [14]. Of these, aneurysm is the most frequent presentation. Dissection of the arteries can happen in both injury and reparative phases. Bleeding in the injurious phase occurs from gap junctions and in the reparative phase it occurs from fragile vessels of the granulation tissue between the outer media and adventitia [5,13]. Stenosis in the injurious phase is due to thrombus formation and overgrowth of the granulation tissue and plaques in the reparative phase [5,11,13].

Differential diagnosis of the renal infarcts and dissection include congenital conditions (Ehlers-Danlos, Marfan, Loeys-Dietz), vasculitis (Behçet disease, polyarteritis nodosa, neurofibromatosis), embolism, coagulation disorders, renal trauma (iatrogenic), infection (mycotic aneurysm and endocarditis), aortic dissection, atherosclerosis, FMD, and SAM [15]. The workup for coagulation disorders, vasculitis, embolic phenomenon, and infection was negative in our patient. Our patient did not have history of renal trauma and no signs of congenital disorders were present. MRA and catheter angiography demonstrated no evidence of atherosclerosis. FMD was excluded based on the male sex and age of the patient. In addition, FMD presents more often with hypertension and less likely with acute dissection. Unlike FMD, SAM presents as profuse bleeding and dissection. Asymptomatic cases of SAM with no gaps and reparative phase may evolve into medial fibroplasias and perimedial dysplasias, the most common types of FMD [10,12,15]. A definite diagnosis can only be made with the histopathology. We did not attempt to obtain a biopsy of the renal artery as it would cause more harm than benefit. Due to the rarity of this condition there are no standard guidelines for management.

Management

For life-threatening emergencies such as massive hemorrhage, endovascular embolization or surgery could be beneficial [16]. Balloon dilatation can also be considered if the patient has uncontrolled hypertension and ischemia. Stenting should not be attempted in the acute phases as the vessels are extremely friable. We believe conservative approaches such as smoking cessation, strict blood pressure control, and aspirin intake were appropriate in our patient. Treatment with endothelin 1 antagonists and norepinephrine antagonists awaits the confirmation of their role played in the pathogenesis [12]. Due to the rarity of the condition, patient follow-up after acute resolution is not well defined. Few small short-term studies showed full resolution or stability of the imaging findings. A clinical and imaging follow-up study conducted by Kalva et al. demonstrated that 50% of the patients had abdominal pain as presenting complaint [17]. Among 14 patients, presenting findings on angiography included dissection in 10, fusiform aneurysm in 6, arterial wall thickening in 2, and artery occlusion in 1. Follow-up imaging was available in 10 patients; of these, 3 had stable findings, 1 showed improvement, 1 had the disease resolved, and 1 had progression of disease. New imaging findings like dissection and aneurysms were noted in 4 patients. Due to varied progression of the disease, yearly follow-up with computed tomography or MRA is recommended.

Conclusion

SAM is a nonatherosclerotic noninflammatory angiopathy of medium-sized arteries. The lack of reliable criteria poses a diagnostic challenge. We present this case to increase the

awareness of the condition. The rarity and the nonspecific presentation of the SAM often lead to delayed diagnosis. Clinicians should consider SAM as a possible cause of renal infarctions in patients presenting with acute flank pain when most common intra-abdominal pathologies are excluded.

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