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Unilateral renal artery stenosis clinically mimicking scleroderma renal crisis : Case report

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Abstract

Scleroderma renal crisis (SRC) is a life-threatening complication of systemic sclerosis (SSc) that is characterised by new-onset malignant hypertension and progressive acute renal failure, often associated with microangiopathic haemolytic anaemia and thrombocytopenia. SRC was at one time almost fatal, with death often occurring within a few weeks. With the development of angiotensin-converting-enzyme inhibitors (ACE-I), survival has improved dramatically, but death rates still remain unacceptably high. About 20% of SRC cases occur prior to making a diagnosis of SSc. In some cases of SRC, there is no evidence of skin sclerosis. We report a 48-year-old systemic sclerosis malewith accelerated hypertension and deranged renal function.

Keywords

acute renal failure; hypertension; scleroderma renal crisis

Introduction

SSc is an autoimmune disease of uncertain aetiology. It is characterized by inflammation and fibrosis in the skin and internal organs. The most important renal complication in SSc is scleroderma renal crisis (SRC). Abrupt onset of a accelerated hypertension and acute renal failure in a scleroderma patient should evoke the possibility of SRC [1]. SRC is associated with a high degree of mortality [2]. Early treatment with angiotensin-converting enzyme (ACE) inhibitors has reduced this to an estimated 24% [3].

We report a 48-year-old systemic sclerosis male with accelerated hypertension and deranged renal function.

Case Report

A 48-year-old man with a 2-year history of systemic sclerosis (SSc) presented with occipital headache. There was associated nausea and one episode of vomiting. He denied fever and nocturnal headaches. There was no history of blurring of vision, diplopia, seizures or loss of consciousness.

On examination, the patient was conscious and oriented. Eye movements were normal in all directions. Pupils were equal and reacting to light. There was no nystagmus. His blood pressure was

200/110 mmHg. There were no hemorrhages, no exudates and no evidence of papilloedema on fundus examination. He was moving all his limbs with grade 5 power. There was no sensory loss. Deep tendon reflexes were brisk with extensor plantars. He did not have any cerebellar signs or signs of meningeal irritation.

All his laboratory parameters were within the normal range except for serum creatinine, which was progressively increasing. Our patient did not have anaemia and thrombocytopenia. The patient was doing well these 2 years and was on immunosuppression with Mycophenolate mofetil, low dose of a steroid and vasodilator.

During the course of his illness, his blood pressure was normotensive and serum creatinine was within the normal range. But now, hypertension (blood pressure 200/110 mmHg) and progressively increasing serum creatinine (from 1.3 mg/dL to 1.8 mg/dL) led to the clinical suspicion of scleroderma renal crisis (SRC). Initial eGFR (estimated glomerular filtration rate) was 65mL/min/1.73m^2 (CKD-EPI Creatinine equation). Later on eGFR dropped to 44mL/min/1.73m^2. Thus, there was more than 60% reduction in eGFR. Renal biopsy was suggested, but the patient was not willing.

In regard to high blood pressure and Scleroderma Renal Crisis, he was started on angiotensin converting enzyme inhibitor (Ramipril). But his blood pressure was persistently high and serum creatinine progressively increased even after stepping up of the ACE I dosage. Hence, brain imaging was done, which was normal. Further, CE-MRA (3D FLASH) was done to evaluate the renal arteries. Multiplanar reconstruction and Maximum Intensity projection was processed after subtraction .There was significant stenosis of right main and accessory renal arteries, approximately 70% (Figure 1 a, b, c). Left renal artery ostium was normal. No accessory renal artery was seen on the left side. Axial MPR also shows the osteal narrowing clearly on the right side (Figure 2 a, b).

T1 and T2 FSE was obtained before Gd enhanced MRI to evaluate the morphological status of kidney, which showed normal size and parenchymal thickness .Post contrast sequences revealed symmetric contrast perfusion and excretion on both sides (Figure 3). ACE inhibitor was stopped and creatinine came back to normal. But blood pressure was persistently high. Calcium channel blocker was initiated after stopping ACE inhibitors. His response to calcium channel blocker was good and is under regular follow up. CCB gradually lowered his blood pressure over several days to 130/70mmHg.

Discussion

SRC constitutes a new onset of significant systemic hypertension (>150/85 mmHg) and decreased renal function [>=30% reduction in estimated glomerular filtration rate (eGFR)]. A minority of patients may appear normotensive. But their blood pressure will invariably be increased from baseline and accompanied by other manifestations characteristic of SRC [4].

Headaches, hypertensive retinopathy associated with visual disturbances and encephalopathy, seizures, fever and general malaise may occur in SRC.

Pulmonary oedema is also common, resulting from water and salt retention due to fluid overload and oliguria. Arrythmia, myocarditis and pericarditis, if present, may indicate poor prognosis [5].

Biochemical changes in SRC include increased plasma creatinine, microangiopathic haemolytic

anaemia (MAHA), thrombocytopaenia and hyperreninaemia. Hypercreatinaemia may affect >=96% of the patients. Prevalences of MAHA and thrombocytopaenia are estimated to be 60 and 50%, respectively [6]. Our patient did not have MAHA and thrombocytopenia.

High levels of renin may induce hypertension, which may be alleviated following nephrectomy [7]. Urinalysis commonly demonstrates mild proteinuria and haematuria, with granular casts evident on microscopy.

Pathological changes comprise mucin accumulation in arcuate and interlobular arteries, mucoid intimal thickening and fibrinoid necrosis of arterioles with fibrin thrombi [7]. Subsequent manifestations include hypertensive vascular damage, glomerular ischaemia, thrombotic vascular occlusion and fibrosis, and proliferation of intimal cells.

ACE inhibitors significantly improve blood pressure for many patients and, in some cases, may also lead to regression of skin manifestations [8].

Gradual reduction of blood pressure is aimed in SRC as sudden decrease in blood pressure can further diminish renal perfusion and increase the risk of acute tubular necrosis. The addition of calcium channel blockers may be beneficial for patients with inadequate blood pressure reduction on ACE inhibitor therapy alone. Plasma exchange is considered if there is substantial thrombotic microangiopathy. Renal function is supported by intermittent haemodialysis or continuous venousvenous haemofiltration.

Ischemic nephropathy is kidney failure following stenosis or an obstructive lesion in the main renal arteries due to atherosclerosis [9]. It is suspected in the following clinical settings; acute renal failure caused by antihypertensive medication, especially ACE inhibitors, progressive azotaemia in patients with known renovascular hypertension, and acute pulmonary oedema superimposed upon poorly controlled hypertension [10]. MRA is a recommended imaging method for diagnosis.

Imaging is indicated when there is worsening of renal failure after the administration of ACE inhibitor or Angiotensin receptor blocker. The first approach involves direct scanning of main renal arteries with colour Doppler USS followed by analysis of renal artery velocity and pattern with spectral Doppler. However USS is operator dependent and accuracy ranges from 60-90% [11]. Accessory renal arteries and bowel gas are the major limiting factors.

CT Angiography is done when the e-GFR is more than 60ml/mt and CE-MRA is recommended when e-GFR is between 30 and 60%, though renal angiography is the gold standard. Angiographically, RAS is graded as mild (<50%), moderate (50-70%) and severe stenosis (>70%) based on reduction in vessel diameter at the renal ostium. More than 70% is considered hemodynamically significant.

Most studies have shown excellent correlation between Conventional angiography and MRA with sensitivity more than 95% and specificity more than 98 % [12]. MRA should therefore be reserved for patients with high clinical suspicion of RVH; avoiding unnecessary diagnostic angiography in patients with renal failure. Scleroderma renal crisis is a recognized complication of systemic sclerosis. ACE inhibitor is the drug of choice for SRC and majority of the patients do respond well. Uncontrolled hypertension and deterioration of the renal status after the administration of ACE inhibitor (Ramipril) in our patient was unusual. MRA showed 70% occlusion of both the right renal arteries. This would

probably explain the cause for uncontrolled hypertension and renal status deterioration after initiation of Ramipril, which is usually seen in bilateral renal artery stenosis. Its occurrence in unilateral renal artery stenosis makes our case unique. Hypertension was controlled and serum creatinine came to the normal range, when ACE inhibitor was stopped and CCB was initiated.

The therapeutic options for ischemic nephropathy include conservative medical therapy, percutaneous transluminal angioplasty, and surgical revascularization. Interventional therapy should be performed when the medication fails to control blood pressure and to cease the deterioration of renal function. Acute renal failure in association with ACE inhibitor therapy typically reverses with discontinuation of ACE inhibitor, although occasionally, recovery is delayed or does not occur. Furthermore, the administration of calcium channel blocker (CCB) was effective for the control of blood pressure in the present case. CCB has been shown to improve endothelial-dependent relaxation and reverse the vasoconstrictive response to nitric oxide inhibitors, and is effective in preserving kidney function by end organ protection [13].

ACE inhibitor/ARB is the first drug of choice in scleroderma renal crisis. When renal failure is exacerbated after administration of ACE inhibitor/ARB or if hypertension not responding to ACE inhibitor/ARB, renal artery stenosis should be considered, either bilateral or unilateral (as in our patient). Ideally an ultrasound doppler should be done and ACE inhibitor/ARB should be discontinued in such cases.

Figures

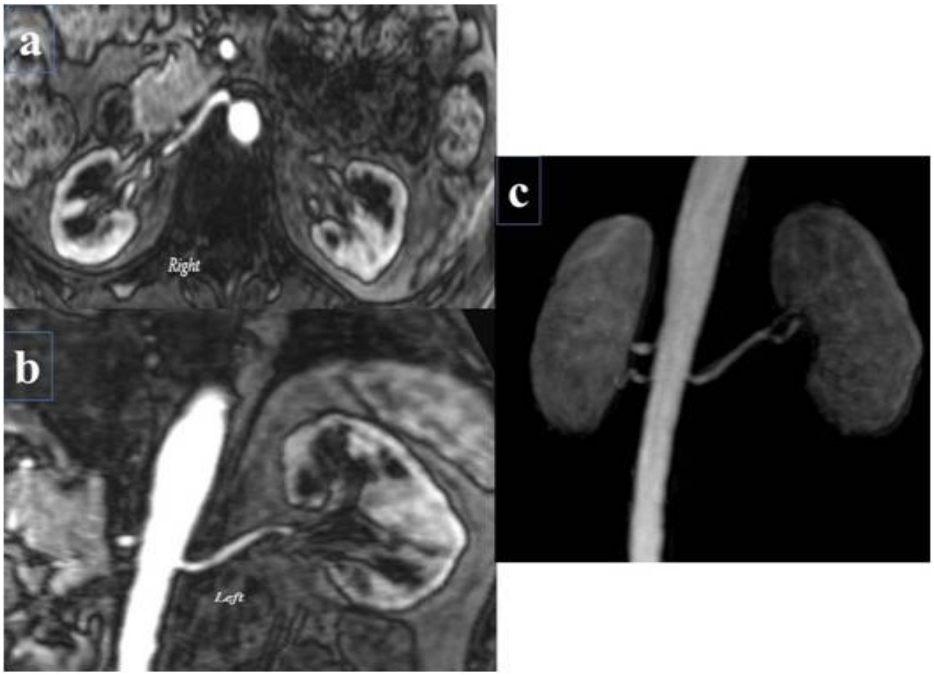

Figure 1: Post contrast (a) Axial 3D FLASH demonstrates significant narrowing at the right renal ostium (b) Coronal images show normal left ostium (c) Coronal 3D MIP images were created to provide an angiographic image. There is significant (more than 70%) narrowing at the right main and accessory renal artery ostium.Left ostium is normal.

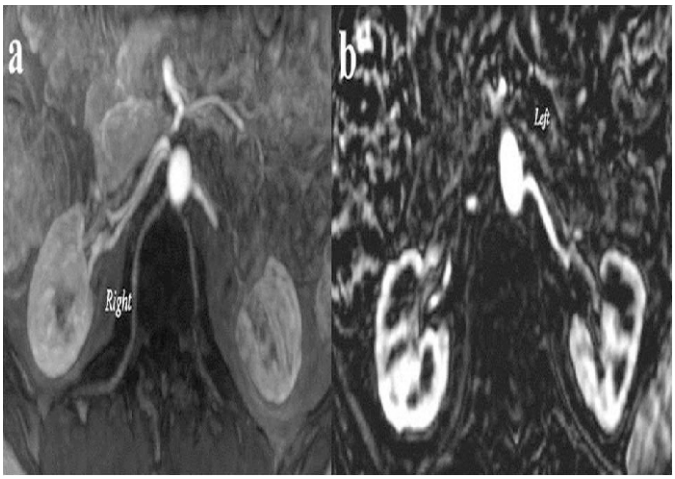

Figure 2: (a) Axial MPR showing renal artery stenosis on the right side. (b) Subtracted image shows normal left ostium.

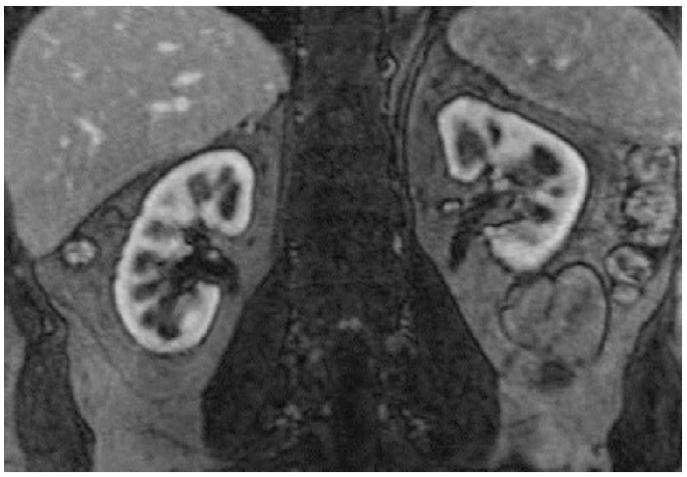

Figure 3: Coronal post contrast images showing normal renal size, parenchymal thickness and enhancement on both sides.

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