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SCLERODERMA AND A COMPLICATED CASE OF HYPERTENSIVE CRISIS, RENAL FAILURE AND RESPIRATORY FAILURE: A CASE REPORT

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ABSTRACT

Background: Scleroderma renal crisis (SRC) is a life-threatening complication of systemic sclerosis characterized by abrupt malignant hypertension, rapidly progressive acute kidney injury, and microangiopathic hemolysis. Although traditionally associated with diffuse cutaneous disease, patients with limited cutaneous systemic sclerosis and high-risk serologic profiles—particularly RNA polymerase III antibodies—remain at substantial risk. Early recognition and uninterrupted renin–angiotensin–aldosterone system (RAAS) blockade are critical to improving outcomes, yet management is often complicated by overlapping cardiopulmonary disease and hemodynamic instability. **Objective:** To describe a complex case of scleroderma renal crisis presenting with hypertensive emergency, acute renal failure, and respiratory failure in a patient with limited systemic sclerosis and RNA polymerase III antibody positivity, highlighting diagnostic challenges and pathophysiology-guided management. **Case Presentation:** A 68-year-old man with limited systemic sclerosis, severe chronic obstructive pulmonary disease, and strongly positive RNA polymerase III antibodies presented with acute respiratory distress, malignant hypertension (217/143 mmHg), and severe acute kidney injury (creatinine 486 µmol/L; baseline 103 µmol/L). Initial management addressed respiratory failure and possible infection; however, evolving microangiopathic hemolysis and persistent severe hypertension shifted the working diagnosis toward SRC. Intravenous enalaprilat was initiated and transitioned to high-dose oral captopril with rapid titration. Blood pressure remained markedly labile, requiring multimodal antihypertensive therapy including nitrates, hydralazine, and calcium channel blockade. Renal function worsened despite RAAS inhibition, and brief continuous renal replacement therapy (CRRT) was required before interruption. Aggressive diuresis subsequently improved pulmonary edema and respiratory status. Renal recovery remained incomplete at discharge planning. **Conclusions:** This case underscores the diagnostic complexity of SRC in patients with limited cutaneous systemic sclerosis and high-risk serology. Extreme blood pressure lability, overlapping cardiopulmonary disease, and apparent hypovolemia can obscure early recognition. Prompt initiation and continuation of short-acting ACE inhibitors, even in the setting of rising creatinine, remain central to management. Multidisciplinary coordination and pathophysiology-driven hemodynamic strategies are essential in stabilizing this high-risk phenotype.

KEYWORDS: Systemic Sclerosis; Scleroderma Renal Crisis; RNA Polymerase III Antibodies; Hypertensive Emergency; Acute Kidney Injury; Microangiopathic Hemolysis; Renin–Angiotensin System; Critical Care; Case Report; Multisystem Disease.

INTRODUCTION

Systemic sclerosis (SSc) is a chronic, immune-mediated connective tissue disease characterized by immune dysregulation, progressive fibroblast activation with

excessive collagen deposition, and a distinctive obliterative vasculopathy affecting small arteries and arterioles.[1] This pervasive vascular injury underlies much of the morbidity and mortality associated with the disease, leading to ischemic tissue damage and fibrosis of the skin and multiple internal organs, including the lungs, kidneys, gastrointestinal tract, and heart.[2] Clinically, SSc is traditionally classified into limited cutaneous systemic sclerosis (lcSSc) and diffuse cutaneous systemic sclerosis (dcSSc) based on the extent of skin involvement, a distinction that has historically guided prognostication and surveillance strategies.[2,3]

Limited cutaneous SSc typically follows a more indolent course, with skin involvement confined to the distal extremities and face and frequent involvement of the gastrointestinal tract, whereas diffuse cutaneous disease is characterized by rapid, proximal skin thickening and a substantially higher risk of early, life-threatening visceral complications.[3,4] These include interstitial lung disease, pulmonary arterial hypertension, cardiac involvement, and scleroderma renal crisis. However, accumulating evidence has demonstrated that serologic profiling may be as prognostically important as skin subtype, with certain autoantibodies conferring organ-specific risk independent of the extent of cutaneous disease,[4,5]

Among these, antibodies directed against RNA polymerase III (RP11 and RP155) define a particularly high-risk SSc phenotype.[6] Their presence has been strongly associated with rapidly progressive disease, malignancy-associated SSc, and, most notably, a markedly increased risk of scleroderma renal crisis in as many as 50% of these patients.[7] This observation has reshaped traditional risk stratification, as patients with otherwise limited cutaneous involvement may still harbor substantial risk for abrupt and severe renal and vascular complications when these antibodies are present.

Scleroderma renal crisis (SRC) remains one of the most feared and life-threatening manifestations of SSc. It classically presents with the abrupt onset of malignant hypertension, rapidly progressive acute kidney injury, and features of thrombotic microangiopathy, often accompanied by acute heart failure, hypertensive encephalopathy, and flash pulmonary edema.[8] Although SRC most commonly occurs within the first several years of disease onset and is more prevalent in diffuse cutaneous SSc, it can also develop in patients with limited or indeterminate disease, particularly in the presence of high-risk serologic markers.[9] Known precipitants include systemic corticosteroid exposure, intravascular volume depletion, acute vasospasm, and progression of underlying vasculopathy.[3,10]

The pathophysiology of SRC reflects profound renal ischemia caused by proliferative narrowing of renal arterioles, leading to intense activation of the renin–angiotensin–aldosterone system.[10,11] This results in extreme blood pressure lability, further vascular injury, and a self-perpetuating cycle of renal hypoperfusion and hypertension. Importantly, patients may appear clinically hypovolemic at presentation, creating diagnostic and therapeutic tension between cautious volume resuscitation and the urgent need for renin–angiotensin system blockade (Figure 1).[11] Since the introduction of angiotensin-converting enzyme inhibitors, outcomes have improved substantially, and early, uninterrupted ACE inhibition using short-acting agents remains the cornerstone of therapy, even in the setting of rising creatinine. Differentiating SRC from pre-renal azotemia, cardiorenal syndrome, or other causes of acute kidney injury can be particularly challenging in patients with overlapping pulmonary and cardiac disease.[12]

In this report, we describe a 68-year-old man with longstanding systemic sclerosis, strong RNA polymerase III antibody positivity, severe chronic obstructive pulmonary disease, esophageal dysmotility, digital ischemia, and active tobacco use who presented with acute respiratory failure, malignant hypertension, and severe acute kidney injury. His presentation highlights the diagnostic complexity of scleroderma renal crisis in the setting of multisystem disease and apparent hypovolemia, the extreme hemodynamic lability driven by renal vasculopathy, and the need for pathophysiology-guided blood pressure management that balances urgent antihypertensive therapy with preservation of renal perfusion. This case underscores the importance of serologic risk stratification, early recognition of SRC physiology, and coordinated multidisciplinary care in managing this high-risk systemic sclerosis phenotype.

CASE PRESENTATION

A 68-year-old man with a history of limited systemic sclerosis presented to the emergency department on admission day 1 with severe dyspnea, increasing work of breathing, intermittent chest pressure, and purulent sputum for four days. Emergency Medical Services (EMS) reported that the patient became acutely dyspneic shortly before arrival, experiencing “three-word dyspnea” and mottling over his abdomen. Crackles were noted on auscultation during transit and he was started on bi-level positive airway pressure (BiPAP) due to worsening respiratory distress. Blood pressure was markedly elevated at 217/143 mmHg prior to treatment. He was clinically volume-depleted with dry mucous membranes and minimal peripheral edema. Lung examination showed good air entry without overt wheeze or crackles. His abdomen was soft and scaphoid. Cardiovascular exam revealed distant but normal heart sounds.

His past medical history was significant for limited systemic sclerosis with features of sclerodactyly, telangiectasias, Raynaud’s phenomenon, Barrett’s esophagus, and esophageal dilation noted on esophagogastroduodenoscopy EGD (last month prior to admission). Serologies revealed antinuclear antibody

(ANA) positivity (1:320), strongly positive RNA polymerase III antibodies (RP11 and RP155), and negative anti-centromere and anti-topoisomerase I (SCL-70) antibodies. He also had COPD with severe obstructive impairment (FEV1 48% predicted), anxiety/depression, a history of thymic cyst resection, and active daily cigarette and cannabis use. Outpatient rheumatology notes documented recurrent digital ulcers, persistent Raynaud's despite therapy requiring a calcium channel blocker, aspiration risk due to esophageal dysmotility, and COPD progression requiring Trelegy Ellipta (Fluticasone, umeclidnium, vilanterol). Earlier in the year, he was switched from amlodipine to nifedipine XL due to refractory Raynaud's ulcers. On admission, his home medications included nifedipine XL 30 mg daily, pantoprazole 40 mg twice daily, fluticasone furoate, umeclidinium, vilanterol inhaler 100/62.5/25 mcg inhaled daily and when needed salbutamol.

Differential Diagnosis, Investigations, and Treatment

The patient presented with acute respiratory distress, chest discomfort, and severe hypertension in the context of several days of progressive dyspnea, cough, and purulent sputum. His background of systemic sclerosis raised immediate concern for scleroderma renal crisis, while other considerations included COPD exacerbation, flash pulmonary edema, hypertensive emergency with cardiac involvement, pre-renal acute kidney injury from hypovolemia, pulmonary infection, and occult cardiac ischemia.

Initial bloodwork revealed severe metabolic acidosis (pH 7.04), elevated pCO_2 (62.7 mmHg), low bicarbonate (16 mmol/L), high anion gap (24), and a rising creatinine of 486 µmol/L (baseline 103 µmol/L two months earlier). Troponin was elevated at 214 ng/L without ischemic ECG changes. WBC was elevated at 18.6×10^9/L. CXR showed hyperinflation, peribronchial cuffing, and mild vascular redistribution but no consolidation.

Management in the emergency department included BiPAP for respiratory failure, bronchodilator therapy, ceftriaxone and azithromycin, and cautious IV fluid administration. Hypertension was initially treated with intravenous nitroglycerin infusion and intravenous hydralazine. Although early clinical impressions considered hypovolemia, the combination of severe hypertension, serologic risk factors (RNA polymerase III positivity), rising creatinine, and emerging hemolysis shifted the working diagnosis toward evolving scleroderma renal crisis. Additionally, intravenous enalaprilat was initiated with the intent to transition to high-dose oral captopril once adequate blood pressure control was achieved and the patient was able to tolerate oral therapy.

During the first 24 hours of admission (Day 0-1), the patient required BiPAP for acute respiratory distress and was treated for a presumed COPD exacerbation and possible pneumonia with ceftriaxone, azithromycin, and bronchodilators. Severe hypertension (initially >200 mmHg systolic) was managed with continued intraveous nitroglycerin infusion and hydralazine. Cautious crystalloid boluses were given due to initial concern for hypovolemia, though fluid administration remained limited because of respiratory compromise. Initial creatinine exceeded 450 µmol/L and continued to rise.

By Day 2, microangiopathic hemolysis became evident, shifting the working diagnosis toward evolving SRC. Urine output remained low (300–400 mL overnight). IV enalaprilat 0.625 mg every 6 hours was continued, while the nitroglycerin infusion was gradually weaned as blood pressure improved. Rheumatology concurred that his serologic profile (RNA polymerase III positivity) placed him at high risk for SRC.

On Day 3, with blood pressure still intermittently exceeding 180–200 mmHg, enalaprilat was transitioned to oral captopril 25 mg three times daily, with a plan for rapid titration. Amlodipine 5 mg daily was added for additional afterload reduction and hydralazine was switched from IV to oral as needed.

By Day 4, hypertension remained labile, with systolic surges triggered by anxiety episodes. Captopril was increased toward 50 mg three times daily, and amlodipine was increased to 10 mg daily. A nitrate patch was added as nitroglycerin infusion was discontinued. Diuresis was emphasized as pulmonary edema contributed significantly to respiratory failure requiring high doses of IV furosemide and metolazone.

Day 5–6 marked worsening renal failure despite escalating antihypertensive therapy. Creatinine trended upward (see Table 1 and Figure 2), reaching values greater than 500 µmol/L. The patient briefly received continuous renal replacement therapy (CRRT), but treatment had to be stopped when the dialysis line was dislodged. Given his hemodynamic stability and improving volume status with high-dose diuretics, CRRT was not reinitiated immediately. Diuresis was highly effective during this period, and metolazone was discontinued by Day 6, while furosemide was tapered to a lower dose.

By Day 7–8, antihypertensive regimens were refined: in addition to diuretics, captopril was maintained (initially 50 mg TID, later decreased to 25 mg TID as renal replacement considerations evolved), amlodipine remained at 10 mg daily, and hydralazine 25 mg four times daily and isosorbide dinitrate 20 mg three times daily was introduced with eventual dose reductions. Clonidine was briefly added during a period of refractory hypertension and subsequently weaned off. All nitrates and hydralazine were discontinued once blood pressure stabilized.

Respiratory improvement paralleled effective volume removal. The patient transitioned from BiPAP to high-

flow nasal cannula and eventually to low-flow nasal cannula as CT imaging showed significant improvement in pulmonary edema.

Throughout hospitalization, creatinine plateaued but remained severely elevated, and renal recovery remained uncertain. Table 1 and Figure 2 illustrate the trend in renal function, blood pressure and mean arterial pressure while highlighting the initial rapid rise, subsequent stabilization, and the ongoing risk of requiring intermittent hemodialysis. Nephrology noted that many patients with SRC may eventually require long-term dialysis and recommended possible transfer for outpatient hemodialysis evaluation once medically stable.

The multidisciplinary team—including rheumatology, nephrology, cardiology, and critical care—coordinated ongoing management, blood-pressure titration, discussions of long-term renal support, and avoidance of triggers that could exacerbate SRC (notably corticosteroids and beta-blockers).

DISCUSSION

This case highlights the significant diagnostic and therapeutic challenges inherent to systemic sclerosis, especially when multiple organ systems deteriorate simultaneously. Systemic sclerosis is driven by a complex interplay of immune activation, progressive fibroblast dysregulation leading to collagen deposition, and a characteristic obliterative vasculopathy.[1] Over time, these processes narrow small arteries and arterioles, impair endothelial function, reduce nitric oxide bioavailability, and amplify the activity of vasoconstrictors such as endothelin-1 and angiotensin II.[2] Clinically, this vasculopathy manifests as Raynaud's phenomenon, digital ulceration, gastrointestinal dysmotility, pulmonary involvement, and potentially SRC.[3] Although SRC is traditionally linked to diffuse cutaneous disease, serologic markers—particularly RNA polymerase III antibodies—are now recognized as strong predictors of renal crisis regardless of skin phenotype. In this patient, the presence of RP11 and RP155 antibodies indicated a high-risk vasculopathic subtype despite his classification as having limited systemic sclerosis.[6]

The pathophysiology of SRC reflects severe renal vascular injury. Progressive narrowing of the renal arterioles leads to abrupt ischemia and triggers marked activation of the renin–angiotensin–aldosterone system (Figure 1).[13] This results in malignant hypertension, further vascular damage, and rapidly worsening renal function. Microangiopathic hemolytic anemia often develops due to high shear stress across stenosed vessels. The clinical picture of sudden severe hypertension, acute kidney injury, and laboratory evidence of hemolysis in this patient was therefore consistent with evolving SRC, even though he appeared clinically volume depleted.[11,14] The coexistence of dehydration and SRC complicates early diagnostic interpretation, as both can produce acute kidney injury. Excessive fluid administration risks worsening pulmonary edema, whereas withholding ACE inhibitors risks irreversible renal ischemia. This case illustrates the need for careful, ongoing reassessment and a nuanced approach to hemodynamic management when these competing processes overlap.

Management in SRC must therefore be dynamic, iterative, and grounded in real-time hemodynamic reassessment. ACE inhibition is the cornerstone of therapy, and its timely initiation is the intervention most strongly associated with improved survival and renal recovery.[3] In patients with SRC, gastrointestinal involvement—including esophageal and gastric dysmotility—is common and may be exacerbated during acute illness, frequently manifesting as nausea, vomiting, or impaired oral absorption. In this context, the use of intravenous enalaprilat provides a reliable means of initiating renin–angiotensin–aldosterone system (RAAS) blockade while ensuring predictable bioavailability during periods of gastrointestinal dysfunction. Short-acting ACE inhibitors such as enalaprilat and captopril are preferred in SRC because they allow rapid titration and fine control of blood pressure, which is critical in a disease characterized by extreme hemodynamic lability driven by pain, anxiety, hypoxia, or procedural stress. Importantly, ACE inhibitors should be continued despite worsening creatinine, as early rises in serum creatinine reflect ongoing renal vascular injury rather than medication toxicity. As gastrointestinal symptoms improved and oral intake became reliable, intravenous enalaprilat was transitioned to high-dose oral captopril, allowing continued RAAS suppression with flexible dose adjustment. Escalation of captopril dosing was essential in mitigating hypertensive surges in this patient, particularly during episodes of heightened sympathetic activation, and formed a central component of his stabilization.

Importantly, our patient had been appropriately managed prior to admission with nifedipine XL for severe Raynaud's phenomenon and digital ischemia, in keeping with guideline-based first-line therapy for peripheral vasculopathy in systemic sclerosis. Despite optimal vasodilator treatment, he subsequently developed scleroderma renal crisis, highlighting that established therapies targeting peripheral vascular disease do not reliably prevent renal vascular involvement in high-risk patients. Positivity for RNA polymerase III antibodies has been associated with a substantially increased lifetime risk of SRC, with cohort data suggesting that up to 50% of these patients may ultimately develop renal crisis.[6] This case therefore underscores a critical gap in current preventive strategies for SRC and raises the question of whether earlier identification of high-risk serologic phenotypes could enable targeted interventions to mitigate renal vascular injury before irreversible crisis occurs. Notably, prior studies have shown that prophylactic initiation of ACE inhibitors before the onset of SRC is associated with worse outcomes, limiting

current pharmacologic prevention options and further emphasizing the need for novel preventive approaches.[4]

A central feature of this case—and an under-emphasized yet clinically critical aspect of SRC—is the extreme blood pressure lability that develops as renal ischemia accelerates RAAS activation. This patient demonstrated dramatic fluctuations, with blood pressure remaining relatively controlled while resting quietly, only to surge into malignant ranges with minimal stimuli such as anxiety, coughing, positional changes, or transient hypoxia. These episodes reflect the impaired autoregulatory capacity of stenosed renal arterioles, heightened sympathetic tone, and stiff vascular beds characteristic of systemic sclerosis. Blood pressure lability complicates management because it requires clinicians to modulate therapy in real time, anticipating surges while avoiding precipitous drops that could compromise renal perfusion. This dynamic instability explains why short-acting antihypertensive agents are essential early in SRC, enabling clinicians to adjust doses minute by minute as physiologic triggers fluctuate.

Sequential and multimodal antihypertensive therapy formed the cornerstone of this patient's stabilization. Enalaprilat, followed by captopril, provided the necessary suppression of RAAS activity, and its continued use despite rising creatinine reflects a fundamental principle in SRC: worsening renal function during early ACE inhibitor therapy is expected and should not prompt discontinuation. Instead, ACE inhibitors mitigate the underlying vasculopathy by reducing efferent arteriolar constriction, improving cortical perfusion, and disrupting the feed-forward loop of renin-driven hypertension. Adjunctive therapies—including nitroglycerin infusion, hydralazine, amlodipine, clonidine, and isosorbide dinitrate—were required to blunt hypertensive spikes that occurred unpredictably throughout the hospital course. Such cumulative therapy mirrors the reality of SRC, in which multiple agents are often necessary not to normalize blood pressure rapidly, but to maintain steady reductions while preserving renal perfusion pressure. Avoidance of beta-blockers was crucial, given their potential to worsen digital ischemia and reduce cardiac output in a vasculopathic state.

Another uniquely instructive aspect of this case was the interruption of continuous renal replacement therapy (CRRT), a complication that has significant implications for SRC but is seldom addressed in published case series. The sudden removal of the dialysis catheter forced a transition to aggressive diuretic therapy at a time when hemodynamics were fragile and antihypertensive titration was ongoing. Managing SRC under these conditions requires balancing RAAS blockade, vasodilator therapy, and diuresis without the stabilizing effect of controlled ultrafiltration. Despite these challenges, sequential nephron blockade with high-dose loop diuretics and metolazone successfully reduced volume overload, improved respiratory function, and prevented further cardiac strain. This scenario highlights an underappreciated reality: behavioral and psychiatric factors can directly impede optimal SRC management, and clinicians must adapt therapeutically even when ideal modalities such as CRRT cannot be sustained. Cases like this underscore the importance of individualized care pathways, contingency planning, and multidisciplinary coordination when unexpected interruptions arise in the treatment of severe renal failure.

Respiratory failure in this patient evolved from multiple physiologic insults, including COPD exacerbation, hypertensive pulmonary edema, and likely aspiration related to esophageal dysmotility—a complication of systemic sclerosis. The use of BiPAP provided both ventilatory support and hemodynamic benefit by reducing preload and sympathetic drive, thereby preventing further hypertensive surges. Treatment of potential infectious triggers mitigated inflammatory effects on vascular tone and renal perfusion. This interplay between pulmonary, cardiac, and renal systems illustrates the systemic nature of SRC and reinforces the need for integrated management strategies.

Finally, social determinants of health played a meaningful role in this patient's disease trajectory. Housing insecurity, inconsistent follow-up, ongoing tobacco use, and limited access to specialty care likely delayed recognition of early warning signs such as progressive Raynaud's phenomenon, worsening dyspnea, or rising blood pressure. Systemic sclerosis requires vigilant long-term monitoring and prompt response to new symptoms; thus, barriers to care profoundly influence outcomes. This case demonstrates how clinical deterioration in systemic sclerosis often reflects the intersection of biological vulnerability and social instability.

There are limitations to this report that reflect the practical challenges of retrospective case reconstruction. The clinical timeline was pieced together from emergency medical services documentation, emergency department assessments, inpatient notes, and outpatient rheumatology records, each of which varied in completeness and detail. Missing or inconsistent documentation introduces uncertainty into the exact chronology of symptom evolution, blood pressure fluctuations, medication adherence, and timing of organ dysfunction. As an isolated case, the experience described here cannot be generalized to all patients with systemic sclerosis, nor can it establish the relative effectiveness of individual therapeutic interventions. Finally, the patient's social instability and limited follow-up complicate the understanding of how earlier medical engagement—or lack thereof—may have influenced the severity of his presentation.

Despite the limitations of retrospective reconstruction, this case underscores several core principles of

scleroderma renal crisis management: the critical importance of early and uninterrupted ACE inhibition, the need for short-acting agents to manage profound blood pressure lability, and the clinical challenges posed by interruptions in renal replacement therapy. More broadly, it illustrates how high-risk serologic profiles, progressive vasculopathy, autonomic instability, and social determinants intersect to drive severe multisystem deterioration in systemic sclerosis, necessitating coordinated, pathophysiology-guided multidisciplinary care.

CONCLUSION

This case highlights the rapidly evolving and multifactorial nature of scleroderma renal crisis in a patient with high-risk RNA polymerase III serology despite a limited cutaneous phenotype. Management required early and uninterrupted ACE inhibition using short-acting agents to control extreme blood pressure lability, alongside aggressive diuresis and adaptive renal support when continuous renal replacement therapy was interrupted. The patient's course underscores the challenges posed by overlapping cardiopulmonary disease, hemodynamic instability, and the need for individualized antihypertensive and volume strategies. Finally, social determinants of health likely contributed to delayed recognition and severity of presentation, reinforcing the importance of early risk stratification, multidisciplinary care, and close longitudinal follow-up in this high-risk population.

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Appendix

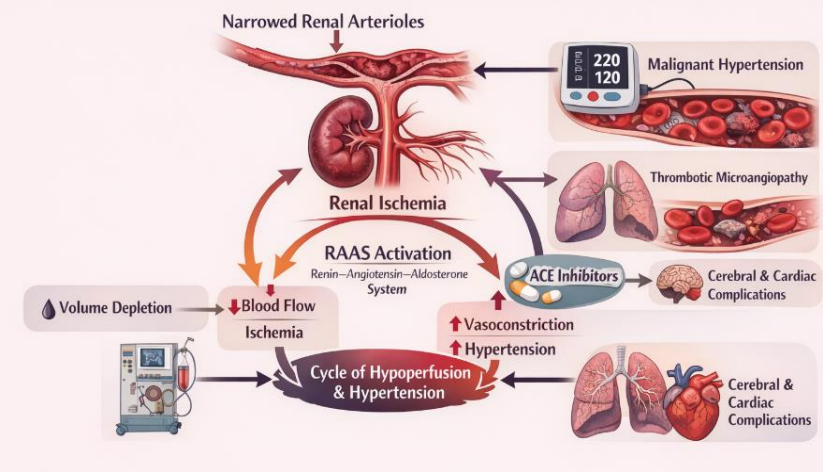

Figure 1: Scleroderma Renal Crisis: Pathophysiology of SRC.

Table 1: Serum Creatinine Trajectory across Hospital Course.

Hospital Date	Creatinine (umol/L)	Mean Blood Pressure (mm HG)	Mean Arterial Pressure
Day 0 (admission date)	486	220/140	166
Day 1	463	165/95	124
Day 2	469	155/100	119
Day 3	471	160/75	103
Day 4	484	165/75	105
Day 5	514	170/75	103
Day 6	534	155/78	101
Day 7	521	140/74	96
Day 8	561	136/85	88
Day 9	435	109/68	82
Day 10	476	113/67	82
Day 11	516	113/71	85
Day 12	527	105/65	78
Day 13	598	114/65	81

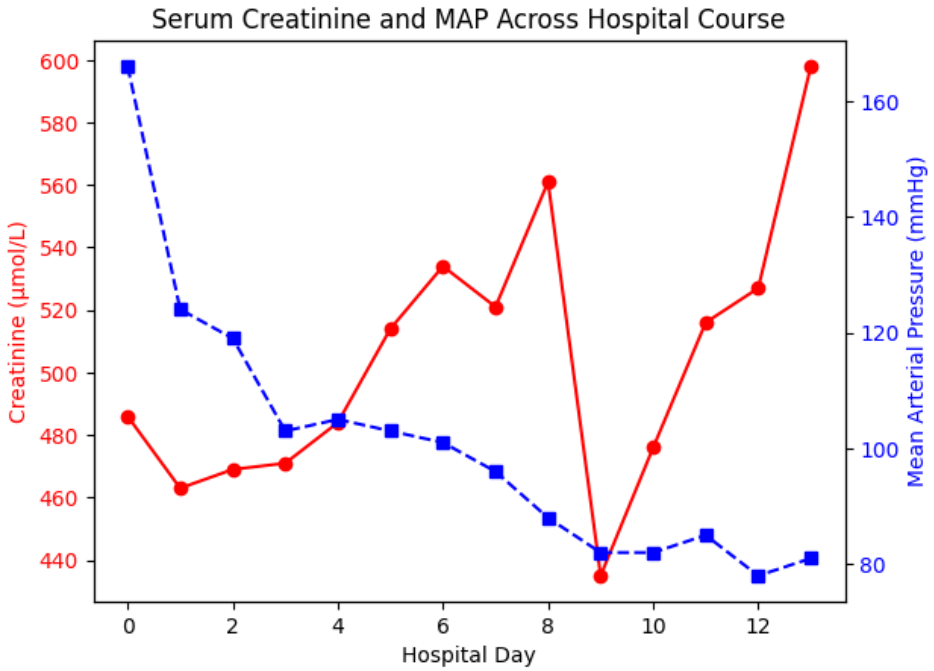

Figure 2: Serum Creatinine and Mean Arterial Pressure Trajectory across Hospital Course.