A Case Report of Systemic Sclerosis Sine Scleroderma Presenting as Renal Crisis

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INTRODUCTION

Systemic sclerosis sine scleroderma (ssSSc) is a rare subset of scleroderma seen only in 2-9% of patients, defined by isolated organ involvement in the absence of skin fibrosis. Scleroderma renal crisis (SRC) is a severe manifestation of systemic sclerosis characterized by malignant hypertension, oligo or anuric renal failure and thrombotic microangiopathy. Corticosteroids are a well-recognized trigger of scleroderma renal crisis.

CASE DESCRIPTION

A 55-year-old male with uncontrolled hypertension presented to the hospital with hematospermia and dark urine.

Vitals: Temp 36.8 ° C, HR 100 bpm, BP 173/97 mmHg, RR 21 bpm, SpO2 99% on room air.

Physical Exam:
General: Alert and oriented
HEENT: **Scleral icterus**, mucous membranes moist
Heart: Tachycardic, regular rhythm, no murmurs
Lungs: Clear to auscultation
Abdomen: Soft, mild tenderness to palpation diffusely
Skin: No rashes or lesions

Labs:

137	100	**84**		**9.4**	
4.6	23	**8.39**	147	8.6	**82**
				28.3	

LDH	752 IU/L
D Dimer	0.80 mcg/mL
Total Bilirubin	1.1 mg/dL
Reticulocyte Count	4%
Haptoglobin	<30 mg/dL
Fibrinogen	423 mg/dL
Direct Coombs Test	Negative
Hepatitis Panel	Negative

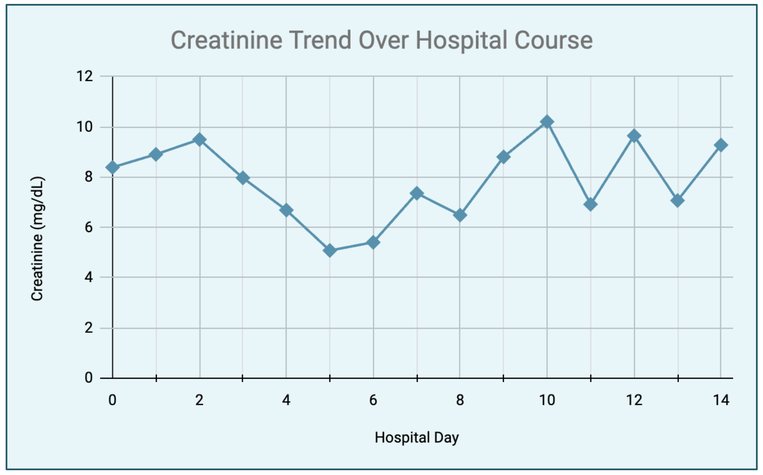
Peripheral Blood Smear: Decreased platelet count without clumping and **numerous schistocytes**

Urinalysis: moderate blood, 30 mg/dL protein

CLINICAL COURSE

Given concern for thrombotic thrombocytopenic purpura (TTP) and hemolytic uremic syndrome (HUS), the patient was empirically started on plasma exchange and received 3 doses of intravenous methylprednisolone followed by a prednisone taper.

The patient had improvement in hemolytic parameters after treatment with blood pressure control, corticosteroids and plasma exchange **but had no improvement in renal function, ultimately requiring hemodialysis.**



Stool studies were negative. An ADAMTS13 level later resulted with a level of 0.6 IU/mL (normal 0.68 - 1.63 IU/mL). The patient was treated with eculizumab therapy for a suspected diagnosis of complement mediated thrombotic microangiopathy.

He underwent a renal biopsy which revealed thrombotic microangiopathy.

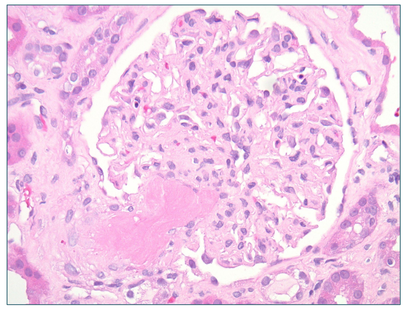


Figure 1. Biopsy of left kidney showing fibrin thrombus at vascular pole

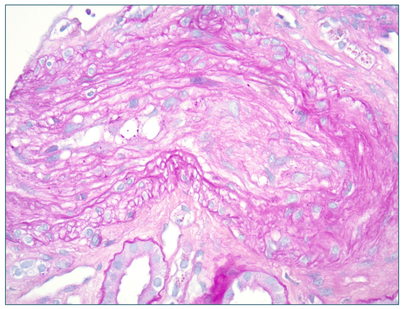


Figure 2. Biopsy of left kidney showing arterial onion skinning

Rheumatologic studies revealed an **ANA titer of 1:320** (normal < 1:40) with a nuclear speckled pattern, Anti-Ro antibody > 8 AI (normal <1 AI), and **RNA polymerase III antibody positive** at 63 U (normal <20U).

CLINICAL COURSE CONTINUED

The patient was diagnosed with systemic sclerosis sine scleroderma presenting as scleroderma renal crisis. Prednisone was discontinued and ACE-inhibitor was maximized.

Day	KEY EVENTS
0	Admitted to hospital
1	PLEX 1
2	HD started/ PLEX 2
3	HD Day 2. PLEX 3. Solumedrol 1000 mg
4	HD Day 3. Solumedrol 1000 mg
5	HD Day 4. PLEX 4. Solumedrol 1000 mg
6	No HD. Kidney Biopsy. Prednisone 60mg daily
7	HD Day 5. PLEX. ADAMS TS13 negative. ANA +
9	Additional rheumatologic investigations ordered
11	Final kidney biopsy shows TMA
12	Eculizumab
14	Discharged from hospital
16	AHUS panel: equivocal
18	RNA Polymerase III results positive
20	Follow up in our office

DISCUSSION

- SRC should be on the differential diagnosis for thrombotic microangiopathy despite lack of cutaneous manifestations of systemic sclerosis as renal involvement can occur in 22% of ssSSc.
- Morbidity and mortality in SRC is high with 40-50% of patients requiring hemodialysis and a 5-year mortality rate of 50-70%.
- The empiric management of TTP can include the use of corticosteroids which can exacerbate SRC, an early clue in the diagnosis of this disease.
- Kidney biopsy was crucial in guiding diagnosis as the pathologist noted that overt fibrin thrombosis seen in our patient's biopsy is not a frequent finding in TMA due to accelerated hypertension alone and the arterial involvement favors SRC over HUS or TTP. The biopsy prompted further rheumatologic investigations.
- This case illustrates the importance of keeping a broad differential when clinicians are presented with thrombotic microangiopathies, and the challenges associated with empiric management of this rare but often fatal disease.