A Case of Systemic Sclerosis Sine Scleroderma

Adrian Lugo, M.D., Andrew Cappiello, M.D., Nemer Dabage, M.D., Guruswamy Ramamurthy, M.D.

Introduction

- Scleroderma and systemic sclerosis are autoimmune phenomena that can cause progressive and permanent damage to the organs of the human body.
- They can cause fibrosis of various internal organs, including the kidneys, lungs, GI tract, and heart
- Rarely, the only manifestation of systemic sclerosis is severe hypertension with renal failure, a subset known as systemic sclerosis sine scleroderma.

Case Presentation

- In this case report, a 39-year-old Hispanic female with a past medical history of GERD and gestational hypertension had presented for nausea and vomiting. She was a Jehovah's witness. Upon review of admission labs and studies from years prior, her chemistries had shown signs of progressive renal failure and proteinuria, but the cause of such abnormalities was never pursued. On admission, she was immediately transferred to the intensive care unit for severe metabolic acidosis requiring emergent hemodialysis, treatment for hypertensive emergency, severe anemia, and electrolyte derangements. Moreover, due to her religion, she declined blood products, so she required erythropoietin and iron products instead. She remained in the ICU for three days for severe metabolic acidosis with a chronic, non-oliguric nephrotic syndrome pattern.

Characteristics

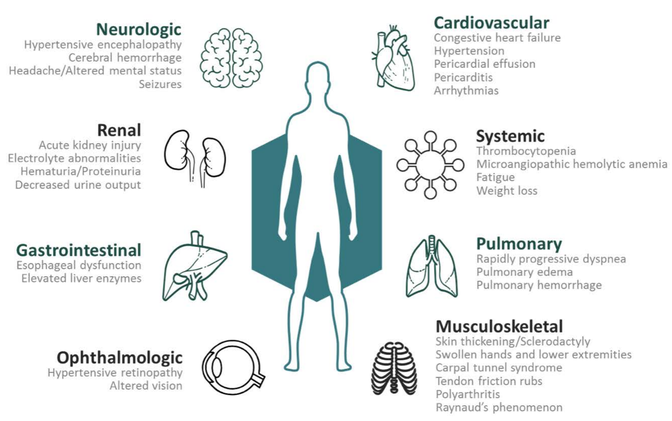

Figure 1: Clinical manifestations commonly found in systemic sclerosis (Montrief).

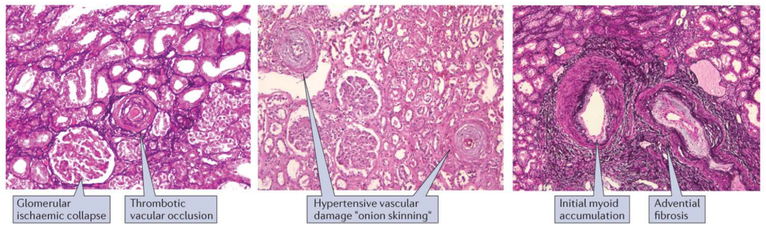

Figure 2: The hallmark of scleroderma renal crisis is a proliferative, obliterative arteriolar vasculopathy with (a) hypertensive vascular damage, thrombotic vascular occlusion, glomerular ischaemic collapse, and diffuse tubular degeneration (b), onion skinning, and (c) intimal myoid accumulation and adventitial fibrosis (Woodroth).

Results

- An extensive immune panel was positive for the antinuclear antibody, the Scl-70 antibody, and low C3. A biopsy was not attempted because the patient's hemoglobin was too unstable at the time.
- On admission, labs were: Hb 7.2, K 3.1, HCO3- 14, BUN 111, Cr 17.9, GFR 2, Ca2+ < 5, Ph 8.3, and severe proteinuria in the urine. Previous outpatient labs showed a Cr of 1.2 in 2015, Cr of 1.7 in 2017 with stage 3B CKD, and increasing proteinuria between 2014 and 2017 that was never pursued.

Conclusion

- This demonstrates a rare incidence of systemic sclerosis sine scleroderma in a patient who was positive for the Scl-70 antibody with no skin findings.
- Several limitations, including religious beliefs, reduced the scope of a full investigation and treatment options.
- It also highlights the importance of pursuing abnormalities on routine blood studies because it can ultimately prevent irreversible kidney damage and can aide in anticipating further internal organ involvement.

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