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Scleroderma Renal Crisis: Identifying Risk Factors and Preventing Adverse Outcomes

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Introduction

Scleroderma renal crisis (SRC) is characterized by an abrupt onset of severe hypertension that is typically associated with increased plasma renin activity and acute renal failure.

- Risk factors include a previous diagnosis of diffuse scleroderma, positivity in RNA polymerase III IgG autoantibodies and recent use of high dose glucocorticoids.
- This case presents a patient who was diagnosed with scleroderma renal crisis after a two week course of steroids.

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Case Presentation

- A 57-year-old female with a past medical history of non-small cell lung cancer (NSCLC) with metastasis to the brain and diffuse scleroderma presented to the emergency department with a three day history of fatigue and weakness.
- She had been receiving radiation therapy for her brain metastases and was concurrently taking dexamethasone 4 mg daily.
- One month prior to admission, the patient was seen by rheumatology for scleroderma follow up at which point positivity of RNA polymerase III autoantibodies was seen on lab work.
- On admission she had profoundly elevated blood pressures without a longstanding history of hypertension. Lab work revealed a hemolytic anemia and acute renal failure complicated by hyperkalemia and a creatinine 7x her baseline. Urinalysis showed proteinuria and hematuria.
- Suspicion for SRC was high, but due to hyperkalemia, ACE-Inhibitors were held at admission. Emergent hemodialysis was initiated and captopril was subsequently administered.
- Unfortunately, the patient's hospital course was complicated by septic shock and respiratory failure secondary to pneumonia. Given her underlying NSCLC and poor prognosis her family ultimately decided to proceed with comfort measures only.

Discussion

- This patient had multiple risk factors for scleroderma renal crisis, including a diagnosis of diffuse sclerosis, the presence of autoantibodies directed against RNA polymerase III, and recent steroid use.
- Given her risk factors, closer monitoring of her renal function after starting the steroid regimen may have led to an earlier diagnosis.
- Literature suggests that prompt diagnosis and treatment is critical as this can slow irreversible renal injury. For high risk patients, literature states that daily home blood pressure measurements are recommended.
- Early recognition in this case was especially important given her co-morbidities. By the time she was admitted, her development of hyperkalemia and sepsis made administration of ACE-Inhibitors more challenging.
- ACE Inhibitors are the initial treatment of choice for SRC, with a goal of returning the patient back to his or her baseline blood pressure within 72 hours.
- If there is central nervous system involvement, intravenous nitroprusside can be added for a short duration of therapy.
- Had this patient been diagnosed earlier, literature suggests that ACE-Inhibitor therapy affects the survival rate at one year significantly, from 15% to 76%.