

Sclerodermic Renal Crisis (SRC)

Alvaro Pedroza Pallares

Internist – Nephrologist Arches Sanatorium. Buenos Aires, Argentina.

Corresponding Author: Alvaro Pedroza Pallares, Internist – Nephrologist Arches Sanatorium. Buenos Aires, Argentina.

Received Date: September 09, 2022; Accepted Date: September 19, 2022; Published Date: September 23, 2022

Citation: Alvaro Pedroza Pallares (2022). Sclerodermic Renal Crisis (SRC). *International Journal of Clinical Nephrology*. 1(2); DOI:10.37579/2834-5142/006

Copyright: © 2022 Ajibade AJ. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Systemic sclerosis (SS), is characterized by the presence of thickening and hardening of the skin (from the Greek "sclero"), it has forms of limited and diffuse manifestation. It is chronic in pattern and manifests with vascular dysfunction and severe systemic connective fibrosis.

Keywords: sclerodermic renal crisis; systemic sclerosis

Systemic sclerosis (SS), is characterized by the presence of thickening and hardening of the skin (from the Greek "sclero"), it has forms of limited and diffuse manifestation. It is chronic in pattern and manifests with vascular dysfunction and severe systemic connective fibrosis. Sclerodermic renal crisis (SRC) is the severe manifestation in the kidney, and is characterized by:

- Malignant hypertension, of sudden onset associated with increased plasma renin activity. There is a clinical variant, which presents with normotension, which has a worse prognosis. (UTAH)
- Acute Renal Failure (ARF).
- Proteinuria in the non-nephrotic range. Microscopic hematuria and casts are uncommon.

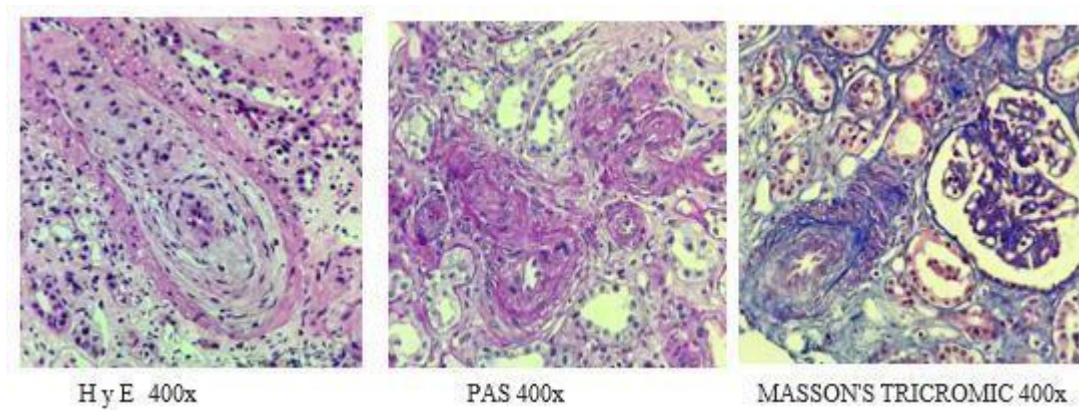
In our experience, we found the case of a 60-year-old woman with a history of systemic scleroderma with severe skin involvement and diagnosed Raynaud's phenomenon, due to skin ulcers, sclerodactyly, microstomia and marked sclerosis of the face and trunk; pulmonary hypertension and arterial hypertension. At the time of onset, he presented with severe arterial hypertension, which required systemic vasodilator drugs for its control, associated acute renal failure with signs of fluid overload. In the laboratory, increased nitrogen products, negative immunological markers of glomerular disease. After ruling out renal artery stenosis with Doppler ultrasound, treatment was started with enalapril 10mg every 12 hours, achieving rapid and effective control of blood pressure figures. Transthoracic echocardiogram shows increased pressure in the right heart chambers, and pulmonary hypertension, starting treatment with tadalafil. Due to a tendency to oliguria, a renal biopsy was performed, which reported "thrombotic

microangiopathy and subintimal arteriolar sclerosis linked to scleroderma renal crisis".

Given the findings in the pathology report, it was decided to continue treatment with enalapril 2.5mg every 12 hours, tolerating doses of up to 10mg per day. Renal failure persists, associated with a decrease in urine volume, and he begins treatment with three-weekly hemodialysis, which continues until discharge from the clinic. He currently retains urinary volume.

SRC is characterized by arterial hypertension, progressive renal failure, and thrombotic microangiopathy. It occurs in 5% of patients with SS. Diagnosis is based on clinical symptoms, but confirmation is through renal biopsy. The presence of anti-RNA polymerase III antibodies is associated with a higher risk of SRC, however, the presence of anti-topoisomerase and anti-centromere antibodies are associated with a better prognosis. ACE inhibitors are the main treatment, even in the presence of renal failure. By being able to competitively inhibit the conversion of angiotensin I to angiotensin II, vasoconstriction and hypertension are reduced, and although both angiotensin I and renin continue to accumulate, they are biologically inactive.

Dialysis is indicated for fluid overload and hypertension refractory to pharmacological management, however, 55% of patients requiring renal replacement therapy (RRT) manage to gradually come out of it and present a decrease in nitrogen values. If after 12 to 18 months, the patient continues with an indication for dialysis, transplantation is a therapeutic option. There is the possibility of relapse with the transplanted kidney, although this situation must be differentiated with histological studies from toxicity due to immunosuppressants.

**Ready to submit your research? Choose ClinicSearch and benefit from:**

- fast, convenient online submission
- rigorous peer review by experienced research in your field
- rapid publication on acceptance
- authors retain copyrights
- unique DOI for all articles
- immediate, unrestricted online access

At Auctores, research is always in progress.

Learn more <https://clinicsearchonline.org/journals/international-journal-of-clinical-nephrology>



© The Author(s) 2022. **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.