RENAL INVOLVEMENT IN SYSTEMIC SCLEROSIS: SCLERODERMA RENAL CRISIS – A CASE REPORT

Margareta Fistrek Prlic1, Stela Bulimbasic 2,3, Marko Baresic4, Ema Ivandic1, Jelena Kos1, Marijana Coric 2,3, Ivana Vukovic Brinar1,3
1 Department of Nephrology, Arterial Hypertension, Dialysis and Transplantation, University Hospital Center Zagreb, Zagreb, Croatia
2 Department of Pathology, University Hospital Center Zagreb, Zagreb, Croatia
3 Medical School, University of Zagreb, Zagreb, Croatia
4 Department of Clinical Immunology and Rheumatology, University Hospital Center Zagreb, Zagreb, Croatia

- OBJECTIVE: SCLERODERMA RENAL CRISIS (SRC) IS A RARE COMPLICATION OF SYSTEMIC SCLEROSIS (SS), OCCURRING IN 5% OF PATIENTS. IT IS CHARACTERIZED BY MALIGNANT ARTERIAL HYPERTENSION (AH), ACUTE RENAL FAILURE (ARF) AND LEFT VENTRICULAR INSUFFICIENCY (LVI). SC IS MORE COMMON IN PATIENTS RECEIVING CORTICOSTEROIDS. THROMBOTIC MICROANGIOPATHY (TMA) IS DETECTED IN 40% OF SC. RENAL BIOPSY IS NOT NECESSARY IF CLASSICAL CLINICAL FEATURES ARE PRESENT, BUT HELPS TO DETERMINE THE DIAGNOSIS IN ATYPICAL FORMS.
- AIM: WE PRESENT A CASE REPORT OF A PATIENT PRESENTING WITH SC.
- CASE REPORT: A 71-YEAR-OLD FEMALE PATIENT WAS ADMITTED DUE TO DYSPNEA, CHEST PAIN, ARF, AND UNREGULATED AH. FOUR MONTHS EARLIER SHE WAS DIAGNOSED WITH DIFFUSE CUTANEOUS SS WITH THE AFFECTION PREDOMINANTLY OF THE SKIN, BUT ALSO PERIPHERAL CIRCULATION AND ESOPHAGUS. LOW DOSE CORTICOSTEROID THERAPY WAS STARTED. AT ADMISSION, HER BLOOD PRESSURE WAS UP TO 180/100 MMHG, SHE WAS DYSPNEIC WITH AUDIBLE CREPITATIONS IN THE LOWER THIRDS OF HER LUNGS. SHE HAD MICROSTOMIA AND TIGHT SKIN ON THE FINGERS AND HANDS WITH ULCERATION ON THE FINGERS. WORKUP SHOWED ARF REQUIRING DIALYSIS, AND A MYOCARDIAL AFFECTION WITH LVI. A KIDNEY BIOPSY WAS PERFORMED, FINDINGS CORRESPONDED TO TMA WITH INVOLVEMENT OF EXTRAGLOMERULAR BLOOD VESSELS AND MOSTLY SECONDARY GLOMERULAR CHANGES (MESANGIOLYSIS, ARTERIOLES WITH SWOLLEN ENDOTHELIUM IN THE VASCULAR POLES). FIBRINOID NECROSIS OF ARTERIOLES WAS FOUND (FIGURES 1,2, AND 3). ANGIOTENSIN-CONVERTING ENZYME (ACE) INHIBITOR WAS STARTED, BUT DUE TO AN UNFAVORABLE CLINICAL COURSE, PLASMA EXCHANGE, AND VASODILATORS WERE ALSO ADDED. THE PATIENT'S CONDITION DETERIORATED WITH A FATAL OUTCOME.

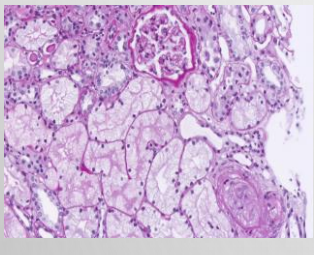

FIGURE 1. MUCOID PROLIFERATION WITH OBLITERATION OF ARTERY, ACUTE TUBULAR INJURY WITH ISOMETRIC VACUOLISATION AND BLOODLESS GLOMERULUS (PAS, 200X)

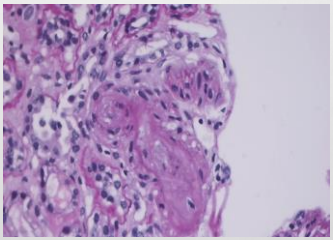

FIGURE 2. FIBRINOID NECROSIS OF ARTERIOLE (PAS, 400X),

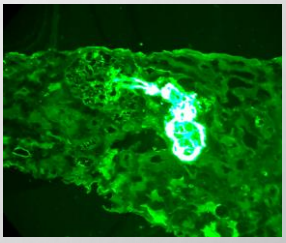

FIGURE 3. STRONG FIBRINOGEN POSITIVITY IN EXTRAGLOMERULAR ARTERIOLE AND VASCULAR POLE OF THE GLOMERULUS

- DISCUSSION: IN OUR PATIENT, THE DIAGNOSIS WAS PROMPT. HOWEVER, HER CONDITION DETERIORATED IN TERMS OF CARDIAC AND RENAL FAILURE.
- CONCLUSION: SRC IS A RARE BUT SEVERE COMPLICATION OF SS. IT IS CRUCIAL TO PROMPTLY INTRODUCE AND CONTINUE THE THERAPY WITH ACE INHIBITORS DESPITE THE END STAGE RENAL FAILURE BECAUSE IT GREATLY IMPROVES PROGNOSIS. HOWEVER, THE 5-YEAR SURVIVAL IS LOW (65%). ADDITIONAL THERAPIES ARE NEEDED TO IMPROVE THE PROGNOSIS.