Systemic Sclerosis with Focus on Scleroderma Renal Crisis

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Known as systemic sclerosis (SSc), this autoimmune rheumatic disease has vast pathogenesis on many organs, including kidneys. It can lead to the point where the patient's survival relies entirely on dialysis. This report has basically focused on scleroderma renal crisis (SRC), which is the most serious renal manifestation of SSc, characterized by renal failure and sudden onset of hypertension. A 44-year-old man was hospitalized with hypertension, headache, vertigo, nausea, rhinorrhea, reflux, dysphagia, dyspnea (Fc II), visual impairment, mechanical arthralgia, and edema (+3) accompanied by a rare skin lesion. Raynaud's phenomenon was also remarkable in fingers and toes. According to signs and symptoms, SSc diagnosed and the proper treatment was applied. It is of great importance that in the case of malignant hypertension in patients with scleroderma, renal crisis always be kept in mind.

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Case Report

INTRODUCTION

Systemic sclerosis (SSc) is an inflammatory autoimmune rheumatic disease that leads to organ fibrosis. More prevalent among women, Scleroderma typically presents between ages 30-60 and is classified into two subsets: limited cutaneous SSc(lcSSc), and diffuse cutaneous SSc(dcSSc).[1]

Scleroderma renal crisis (SRC) is a complication of SSc.[2-4] SRC is present in 5% of SSc patients and is mostly diagnosed within the early 4 years of dcSSc.[5,6] Patients with SRC mainly complain about hypertension and renal failure.

Data from D-Penicillamine trial on SRD revealed a mortality and morbidity rate of 50% during the first 0.9 years.[7] Before the routine use of ACEIs, SRC was reported in 12–18% of SSc patients and was known as the major cause of their death;[8] however with the application of ACEIs, which resulted in the reduction of 12-month mortality rate from 76% to 15%,[5] the blame had been shifted to pulmonary complications.[9] Yet, the prognosis of SRD remains reserved.[4,7,10]

CASE REPORT

A 44-year-old man was hospitalized in 2012 with hypertension (145/80 mmHg), headache, vertigo, nausea, rhinorrhea, reflux, dysphagia, dyspnea (Fc II), and edema (+3) accompanied by a rare skin lesion. He claimed that he had been having mechanical arthralgia, which commenced, from his ankles a year ago. After 3 months of taking non-prescribed NSAIDs, he was diagnosed with rheumatoid arthritis and received corticosteroids. Six months later, red patches started to emerge; Initiating from his palms, they spread all over his body (Figure 1). Rashes were worsened by sun exposure and cold weather. Raynaud's phenomenon became positive.

Serology results were: WBC = 9.8*1000 /mm3, RBC = 3.02 million /mm3, Hb = 10 g/dL, MCV = 92 fl, Urea = 154 mg/dL, BUN = 202 mg/dL, Cr = 9.2 mg/dL, Uric acid = 9 mg/dL, Na = 132 mEq/L, Ca = 9.6 mg/dL, K = 5.4 mEq/L, Alb = 4g/dL, TIBC = 456 mcg/dL, Ferritin = 604 ng/mL, CRP = neg, PTH = 532 pg/mL, Viral marker = neg, U/A = pro(+2), SG = 1.005, pH = 6, U/C = neg, and ESR = 36 mm/1st hour.

Ultrasonography showed renal cortex thinning, renal shrinkage (R-7.8 / L-8.4) and increased histological echo. No kidney stone or hydronephrosis was reported. Bladder, liver, and

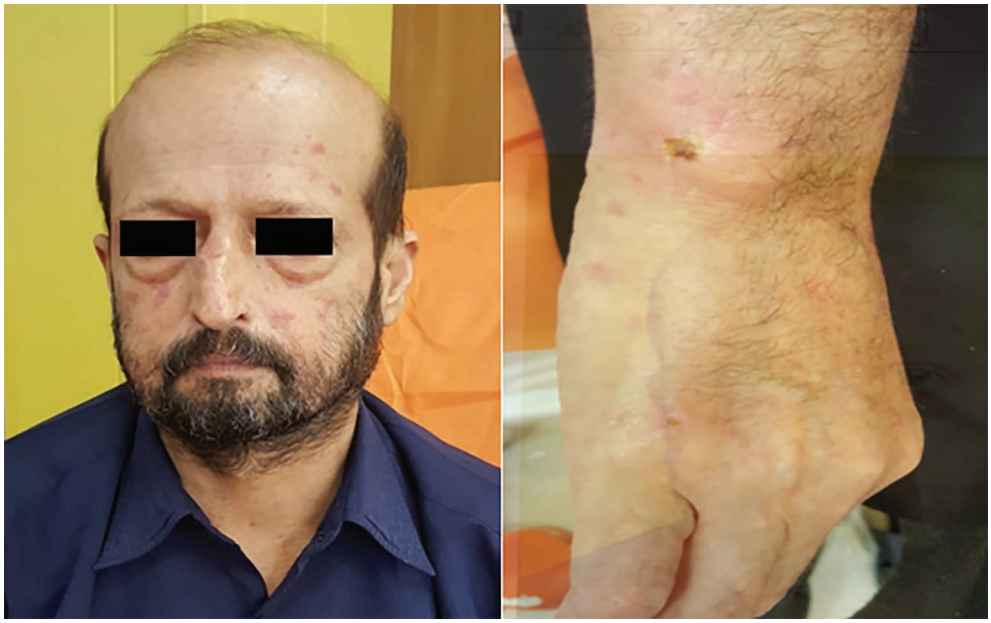

Figure 1. (A & B). Red Rashes were spread all over the body. Samples were taken from abdominal skin and right shin.

gallbladder were normal.

Abdominal and right shin skin lesions biopsies revealed vascular interface reaction with superficial and deep dermal vasculopathy and sclerodermoid pattern, in favor of mixed-connective tissue disease.

Irreversible bilateral macular edema was diagnosed and the visual acuity was decreased to R-4/10 and L-2/10. Surgery was recommended (Figure 2).

CXR showed pericardial effusion. Myocardial perfusion scan by Tc-MIBI showed mild stress-induced ischemia in the apicoseptal segment and a prominent diaphragmatic attenuation in the interoapical segment (Figure 3).

BMD illuminated spinal osteoporosis and mild osteopenia in femur and wrist (Table 1).

Accordingly, SSc was diagnosed and the proper treatment was applied. After 5 years, the symptoms were worsened due to drug attainment difficulties (Prostin VR 500mg (2cc/1 qw for 2 weeks-every 6 months)).

Thanks to regular hemodialysis, patient's general condition is currently stable; however, pulmonary complications such as mild rales have recently emerged.

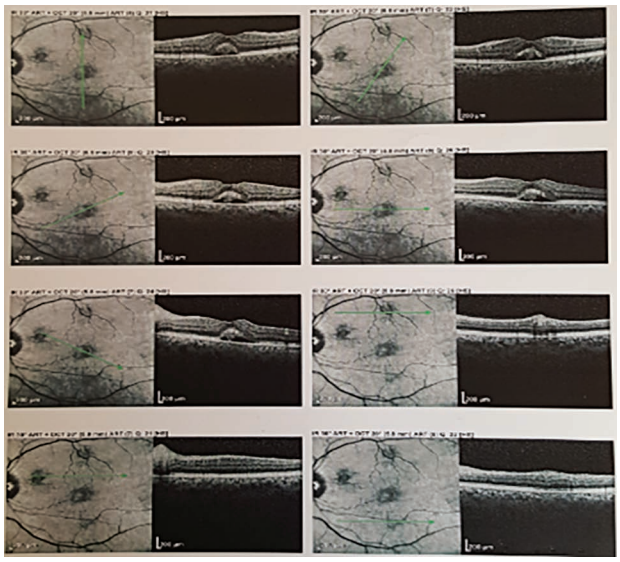

Figure 2. Tracking Laser Tomography. Irreversible bilateral macula edema was diagnosed.

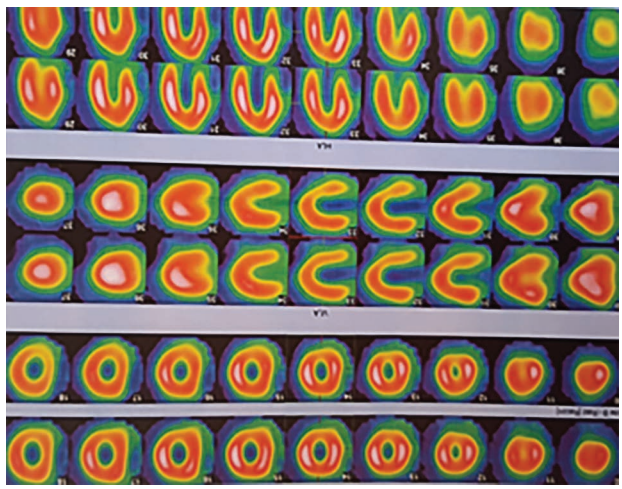

Figure 3. Myocardial Perfusion

Tabel 1. The BMD Measurement was Performed by Dual Energy X-ray Absorptiometry Method (DXA)

R.O.I	BMD, g/cm^2	Standardized BMD, mg/cm^2	T Score	Young Ref, %	Z Score	Relative Risk of Fracture	Diagnosis WHO Criteria
Spine (L1-L4)	0.811	872	-2.5	74	-2.4	7.2	Osteoporosis
Femur (Neck)	0.774	786	-1.1	83	-0.6	2.9	Osteopenia-Mild
Wrist (1/3 Radius)	0.732	-	-1.6	90	-1.3	4.6	Osteopenia-Mild

DISCUSSION

About 60-80% of SSc patients suffer from SRD.[11] Reported in 86% of patients with dcSSc and 10% with lcSSc, SRC is the most serious renal manifestation of SSc.[12,13] Due to systemic hypertension: PAH, hypertensive retinopathy, encephalopathy, pericarditis, and myocarditis might be present.[8,14,15]

Conspicuous changes in patients' blood test are: microangiopathic hemolytic anemia (MAHA) and an elevated level of soluble adhesion molecules such as VCAM-1, ICAM-1, E-selectin, and renin.[16] Immunohistochemical staining technics can validate endothelin axis up-regulation; including endothelin-1 (ET-1) and endothelin B.[17]

Urinalysis reveals hematuria and non-nephrotic range proteinuria. Granular casts may be detected.[18] Kidney biopsies show mucin accumulation in interlobular arteries and fibrinoid necrosis of arterioles.[19] Propounded risk factors of SRD that can partially effective in severity of the disease are: diffuse and progressive skin disease,[19,24] anemia,[20] cardiac events,[7] large joint contractures,[7] steroid usage (415 mg, prednisolone/d),[21] cyclosporine therapy,[22] presence of anti-RNA-polymerase antibody,[23] early stages (< 4 years) of SSc.[25]

ACEIs seem to be crucial for controlling SRC and can lead to a higher rate of recovery and survival.[5,21] Yet, 20-50% of patients with SRC require renal replacement.[15]

CONCLUSION

We presented a case of scleroderma with an exclusive focus on the renal crisis. It is important that in cases of Scleroderma with malignant hypertension, renal crisis always be kept in mind.

CONFLICT OF INTEREST

None declared.

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