

Scleroderma renal crisis sine scleroderma in pregnancy: A case report

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ABSTRACT

Scleroderma renal crisis has been documented as the presenting manifestation of systemic sclerosis sine scleroderma in pregnancy only once in the literature. Unfortunately, since scleroderma renal crisis in sine scleroderma pregnant patients is so rare, that patient expired. We present a case of a sine scleroderma pregnant patient with an initial manifestation of scleroderma renal crisis surviving due to successful diagnosis and treatment.

Introduction

Systemic sclerosis sine scleroderma was first described by Rodnan in 1961 (1). Patients affected by systemic sclerosis sine scleroderma initially present with internal organ involvement but usually develop skin thickening within several months. Scleroderma renal crisis (SRC), first described in 1863 by Auspitz, has been reported as the initial presentation of systemic sclerosis sine scleroderma previously (2-6). SRC is defined by the abrupt onset of accelerated hypertension, followed by rapidly progressive oliguric renal failure (7). Although there has been a report of a pregnant patient without scleroderma presenting with scleroderma renal crisis, the diagnosis was determined with renal biopsy and the patient expired (8). We present a case of a pregnant patient with scleroderma renal crisis as the initial manifestation of systemic sclerosis sine scleroderma.

Case presentation

A previously healthy 27-year-old Caucasian female presented at 23 weeks gestation following a seizure thought to be secondary to eclampsia. The blood pressure on admission was 200/130 mmHg. There was no history of Raynaud phenomenon, arthritis, pruritis, or skin changes. The initial physical examination revealed a gravid abdomen. There was no skin thickening or tendon friction rubs. Initial laboratory values revealed the following: AST 60 IU/L (8-34 IU/L); ALT 38 (7-35 IU/L); Platelet count 33 K/cm³; Hematocrit 32.6%; White Blood Count 16.1 K/cm³; Creatinine 1.5 mg/dl (0.6-1.1 mg/dl); Urine analysis 9 RBCs, 9

WBCs, 14 Hyaline Casts, and Protein 300 mg/dL.

At this time the working diagnosis was eclampsia and HELLP (Hemolysis Elevated Liver enzymes Low Platelets) syndrome. A Caesarean section delivered a non-viable 445g fetus. Although the fetus was delivered, the patient's condition failed to improve. The liver enzymes remained elevated; AST was 99 IU/L and ALT was 40 IU/L. Platelet count was 53 K/cm³, hematocrit was 27.5% and white blood count was 12.5 K/cm³. The serum creatinine was elevated at 3.0 mg/dl. She then developed pulmonary edema and hypoxia. Her respiratory status worsened and she briefly required mechanical ventilation. In the setting of renal insufficiency, thrombocytopenia, anemia, and fever, a diagnosis of thrombotic thrombocytopenic purpura (TTP) was entertained; plasmapheresis was performed five times without improvement in her clinical condition or laboratory abnormalities.

Upon further evaluation, it was ascertained that the patient's mother had systemic lupus erythematosus. Further laboratory evaluation revealed an ANA of 1:320 in a homogenous pattern; serum complement levels were low (C3 66 mg/dL, C4 11 mg/dL); antibodies to double stranded DNA were absent. Anti-SCL-70 (anti-DNA topoisomerase I) was positive at 1:320; the remainder of the extractable nuclear antigen profile was negative. Nail fold capillaroscopy exam was performed and vascular dilation and wide areas of avascularity were observed. At that time the diagnosis was felt to be systemic sclerosis sine scleroderma manifesting as scleroderma renal crisis. She was treated with intravenous captopril and had rapid improvement in hypertension and thrombocytopenia. A high resolution chest CT scan in the hospital showed peripheral and basilar predominant septal thickening and ground glass opacities consistent with interstitial lung disease. Pulmonary function tests indicated a restrictive pattern (FVC 2.16 L (64% predicted), TLC 3.09 L (71% predicted), Diffusion 7.7 L (35% predicted), and Diffusion adjusted for ventilation 3.26 L (69% predicted).

Renal function continued to slowly improve over several months. Six months after initial presentation, the serum creatinine was 1.2 mg/dL. Two months following hospital discharge the patient developed skin thickening of her hands, arms, legs, and chest with a modified Rodnan skin score of 15 (0-51). A repeat high resolution CT scan of the chest six months later continued to demonstrate bilateral lower lobe ground glass opacities consistent with alveolitis. A bronchoalveolar lavage revealed 4.2×10^5 inflammatory cells/cm³ with 13% neutrophils (normal less than 3%); 1% eosinophils; 2.5% lymphocytes; and 84% alveolar macrophages. Treatment with daily oral cyclophosphamide was initiated for interstitial lung disease (9) and captopril was continued.

Discussion

SRC has been described as occurring at some point in the disease course in 10% of patients with SSc (10). In a study of 94 patients with SRC, 2 presented with systemic sclerosis sine scleroderma (11). Although the incidence of morbidity and mortality of SSc in pregnancy is unknown, a prospective study did not find an increased rate of miscarriage in SSc patients (10). Early onset of diffuse skin involvement seems to be an important factor in pregnant patients developing SRC (12). To our knowledge, there has been only one other case of a pregnant patient with systemic sclerosis sine scleroderma presenting with SRC reported in the literature (8). Our patient did not have skin involvement at the time of her SRC, but she did develop diffuse skin disease soon after her initial presentation. Detection of SRC is based on physical examination and laboratory evaluation (10). The most important physical examination finding is an elevated blood pressure. However, the elevation may only be relatively elevated for the patient and so actually normotensive. Normotensive SRC occurs in about

10% of all SRC patients (13, 14). Laboratory evaluation for patients with suspected renal crisis includes a complete blood count, metabolic profile, and urinalysis. The complete blood count classically reveals a microangiopathic hemolytic anemia and thrombocytopenia. The metabolic profile may show azotemia and increased creatinine. The urinalysis is usually remarkable for an active sediment, including hematuria, pyuria, and granular casts. 20-50% of patients with systemic sclerosis will be positive for SCL 70 (15, 16) Our patient had positive SCL-70 antibody, positive antinuclear antibody, and a pattern of nailfold capillary abnormalities consistent with systemic sclerosis.

Even though angiotensin converting enzyme (ACE-I) inhibitors are teratogenic, their use is still recommended in pregnant patients. In fact, five patients with a history of SRC were treated successfully with ACE-1 throughout their entire pregnancy (12). Since the introduction of captopril, the mortality for SRC has improved from a 10% one-year survival to 85% 8-year survival (10). Captopril is usually chosen as the initial ACE-I of choice since it can be titrated quickly to achieve a blood pressure less than 120/80 (10). Once there is no longer a need for intravenous medication, any oral ACE-I may be started, maintaining the same blood pressure goal.

In conclusion, the positive serologies, nail fold capillary findings, and response to ACE-I treatment helped to distinguish SRC from eclampsia, HELLP, and TTP. The diagnosis of SRC was confirmed by her subsequent development of diffuse skin involvement in the following months. We believe this case is unique as the diagnosis was made without a renal biopsy and the patient survived the renal crisis.

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