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Short Communication

Scleroderma renal crisis: Importance of knowing and differentiating in emergency unit

Scleroderma Renal Crisis (SRC) is a rare and severe complication of systemic sclerosis precisely in patients with diffuse cutaneous systemic sclerosis, defined by accelerated phase hypertension and acute renal failure, headaches, hypertensive retinopathy and encephalopathy. Laboratory tests may demonstrate Micro Angiopathic Hemolytic Anemia (MAHA), Urinalysis displays non-nephrotic range proteinuria and hematuria. The presence of Anti-RNA-polymerase III antibodies and recent use of high-dose corticosteroids are linked to Scleroderma renal crisis. Use of Angiotensin-Converting Enzyme Inhibitors (ACEI) is mandatory and associated with dramatically improvement in the prognosis of SRC and by knowing its contraindication during pregnancy therefore patient should be counseled regarding morbidity and mortality.

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Introduction

Scleroderma is an autoimmune multisystem inflammatory connective tissue disease that causes fibrotic changes in the skin and vasculature affecting the major organ system [1]. Scleroderma Renal Crisis (SRC) is considered as a fatal complication of systemic sclerosis which is defined by abrupt onset of accelerated high blood pressure and renal failure. It is associated with a high mortality rate that requires specific and careful management [2,3]. SRC during pregnancy required high end management and it is a challenging task for physician's and also vital to differentiate it from preeclampsia. In this backdrop, the present review is focused on SRC emphasizing on clinical presentation, pathogenesis, risk factors, treatment and prognosis.

Clinical presentation

SRC can occur without proceeding symptoms with sudden onset of severe uncontrolled hypertension (>150/85 mmHg), however 10% are normotensive (diastolic BP ≤90 mm Hg). Decreased renal function with approximately >30% reduction in estimated glomerular filtration rate (eGFR) which preludes to acute oligo-anuric renal failure. Microangiopathic hemolytic anemia is common and it is an independent predictor of impending renal failure secondary to the fragmentation of erythrocytes passing through renal arterioles that have become narrowed by intravascular fibrin, although significant coagulopathy is rare. Urinalysis commonly demonstrates non-nephrotic range proteinuria and hematuria. Patients may report symptoms like frequent headache, fever, malaise, hypertensive retinopathy associated with visual disturbances, encephalopathy, pericardial effusion, seizures, and pulmonary edema [4-10]. The data on pregnancy and Scleroderma are limited but SRC is a fear complication and it will be difficult to differentiate if renal dysfunction with high blood pressure occurs during pregnancy due to systemic sclerosis or preeclampsia [11-13].

Pathogenesis

Although the pathogenesis of SRC is not fully understood, endothelial wall injury causes intimal proliferation that leads to narrowing of arterioles and formation of micro thrombi. With possibility of renal Raynaud's phenomenon contributes to renal hypo perfusion, increasing renin releasewhich cause vasoconstriction and worsening renal ischemia which also contributes to accelerated hypertension in hypertensive SRC [14,15].

Risk factors for SRC

SRC has been observed to be associated with patient's characteristic of new onset (within 4 years) and rapidly progressive skin disease (diffuse cutaneous systemic sclerosis), presence of Anti-RNA-polymerase III antibodies (in one third of

patients), recent use of high-dose corticosteroids and, the use of cyclosporine. While, recent use of low-dose steroids, continuous use of any steroid dose, NSAIDs, calcium channel blockers, and ACE inhibitors are not associated with an increased risk of SRC [16,17]. Few study mentioned the benefit of prophylactic use of D-penicillamine if corticosteroid use cannot be avoided. However, one study elicited that there is no advantage of using D- penicillamine in doses higher than 125mg. In addition, D-penicillamine is ineffective in treating early, diffuse, systemic sclerosis [18-21].

Treatment of SRC

Patient with SRC requires intensive care and ACE inhibitor like captopril should be used to control blood pressure aggressively along with other antihypertensive agents, with the goal of decreasing systolic blood pressure by 20 mmHg within the first 24 hours. When the blood pressure decreases below the optimal level, it leads to decrease in renal perfusion and elevates the risk of acute tubular necrosis. ACE inhibitors significantly improves the blood pressure for many patients and, in some cases, may also lead to regression of skin manifestations [22]. SRC during pregnancy will be challengeable as ACE inhibitor is contraindicated and patient should be counseled regarding morbidity and mortality [23]. Dialysis is frequently indicated but can be stopped in about half of patients, mainly those with good blood pressure control. Plasma exchange is considered if there is substantial thrombotic microangiopathy and endothelin receptor antagonists have been recommended as future therapeutic approach [24]. Renal transplantation may be considered in patients who required dialysis for more than 2 years [25,26].

Prognosis

Poor prognostic factors are predicted by normotensive at presentation, cardiac involvement with myocarditis or arrhythmias, presence of acute vascular changes on renal biopsy, and older age if dialysis is required [27-29]. Mortality rates are high and highest in male and those requiring permanent dialysis, short-term prognosis of SRC has improved, but long-term prognosis remains disappointing [29].

Conclusion

The presence of Anti-RNA-polymerase III antibodies and recent use of high-dose corticosteroids are linked to Scleroderma renal crisis. Use of Angiotensin-Converting Enzyme Inhibitors (ACEI) is mandatory and associated with dramatically improvement in the prognosis of SRC and by knowing its contraindication during pregnancy therefore patient should be counseled regarding morbidity and mortality.

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