SCLERODERMA RENAL CRISIS

Scleroderma renal crisis (SRC) used to be the most feared and deadly scleroderma complication. With the use of angiotensin converting enzyme (ACE) inhibitors it has become a manageable disease. Early recognition and aggressive treatment results in a very good outcome in 61% of patients.[1] More than half of patients who initially require dialysis are permanently able to discontinue it.

The definition of SRC is an acute episode of malignant hypertension and progressive renal failure over days to weeks. Microangiopathic hemolytic anemia, abnormal urinalysis and an increased serum creatinine are other findings associated with SRC. Ten percent of patients are normotensive but usually the blood pressure is increased from baseline and other manifestations of SRC are present.[2] Decreased creatinine clearance is not uncommon in scleroderma, but there are many other potential etiologies and it should not be assumed that it is from scleroderma kidney disease. Renal failure independent of SRC is a rare occurrence and likely to be another etiology.[3] Risk factors for renal crisis include early disease (80% of SRC occurs in the first 4 years) with rapidly progressive diffuse scleroderma. Prior increases in blood pressure, increased creatinine or urinary abnormalities do not predict renal crisis.[4] However, high doses of corticosteroids may precipitate renal crisis.

The frequency of SRC varies from country to country because of genetic differences as has been shown in this study by Gupta et al.[5] Ethnicity and genetics are strongly associated with scleroderma specific autoantibodies. SRC occurs in 25% of patients with RNA polymerase III (RNA pol III) antibody.[6] It also occurs in patients with anti-topoisomerase antibody, but is rarely seen in patients with anti-centromere positive limited scleroderma patients. Thus, frequency of renal crisis is dependent on the genetic frequency of these antibodies within the population.

Some have questioned whether the frequency of renal crisis has decreased over the last 20 years. This may be because physicians are not using high doses of corticosteroids which can precipitate renal crisis.[7] Or it may be the more aggressive use of ACE-inhibitors even before the diagnosis of renal crisis has been made. There is no evidence that ACE-inhibitors prevent renal crisis and in fact, there are suggestions that the use of ACE-inhibitors prior to the onset of renal crisis may be associated with a worse prognosis of renal crisis.[8]

Patients in the first four years of rapidly progressive diffuse scleroderma should monitor their own blood pressure and notify their physician if it should become abnormal. When this happens, an ACE-inhibitor should be started immediately. Other causes of kidney problems need to be excluded, but the goal should be to maintain the patient's blood pressure as close to normal as soon as possible. Improved renal function may be delayed for a time after the blood pressure is controlled, but other non-ACE-inhibitor treatment at that point does not improve the overall outcome. Early, aggressive treatment with ACE-inhibitors will give the best outcome.

REFERENCES


VIRGINIA STEEN
Division of Rheumatology, Georgetown University, Washington, DC 20007, USA.
E-mail: steenv@georgetown.edu

This PDF is available for free download from a site hosted by Medknow Publications (www.medknow.com)