Renal Scintigraphy in Acute Scleroderma: Report of Three Cases

Robin G. Woolfson, Hugh S. Cairns, David J. Williams, Andrew J.W. Hilson and Guy H. Neild

Departments of Nephrology and Nuclear Medicine, Institute of Urology and Nephrology, University College London Medical School, London, United Kingdom

Scintigraphic findings in acute renal failure secondary to scleroderma are reported. In three patients, we have demonstrated severe reduction of renal perfusion with little or no parenchymal uptake of tracer and absent excretion. These findings are compatible with the known histological process of occlusive vasculopathy, and such scintigraphic findings at presentation may reflect a poor prognosis for renal recovery.

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Scleroderma is a progressive systemic disease of unknown etiology characterized by vascular and connective tissue lesions which may occur in the absence of accelerated hypertension (*1*). Acute renal failure ("acute transformation") may occur as a dramatic complication of apparently mild disease. We describe the scintigraphic findings of three such patients who presented to our renal unit with acute renal failure.

CASE REPORTS

Case 1

A 55-yr-old woman had presented 3 mo earlier with skin tethering over her knees, feet and upper arms with a history of xerostomia. Over the 4 wk prior to her admission, she had developed worsening exertional dyspnea and latterly paroxysmal nocturnal dyspnea. On admission, she was in gross pulmonary edema with a systemic blood pressure of 230/100. Fundoscopy revealed Grade IV hypertensive retinopathy. Plasma creatinine was 129 μmole/liter and Hb was 9.2 g/dl. Anti-hypertensive treatment with captopril was commenced on admission. Blood pressure was controlled, but urine output fell and plasma creatinine rose rapidly (643 μmole/liter after 5 days). Renal ultrasound demonstrated normal and equal sized unobstructed kidneys. Left renal biopsy was performed. Dynamic renal scintigraphy with 200 MBq ^{99m}Tc-DTPA was performed 8 days after initial admission. She remained anuric and following a period of hemodialysis was treated with continuous ambulatory peritoneal dialysis for 18 mo before dying from progressive cardio-respiratory involvement.

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For correspondence or reprints contact: R.G. Woolfson, MD, MRCP, Department of Nephrology, Institute of Urology and Nephrology, 48 Riding House St., London W1N 8AA, U.K.

Case 2

A 45-yr-old woman had presented 6 mo previously with a symmetrical distal polyarthropathy with digital edema. Skin tethering on the dorsum of fingers and central face developed over the following 2 mo and a diagnosis of scleroderma was made. She had a 7-yr history of ulcerative colitis treated with salazopyrine. The last flare-up had been 6 mo earlier. She presented to us with a central scotoma in her right eye, neck stiffness and headache. On examination, her blood pressure was 140/80 mmHg. Bilateral macular exudates were noted without evidence of hypertensive retinopathy. Plasma creatinine was 540 μmole/liter. Three days later, she became anuric and developed congestive cardiac failure with pulmonary edema and hypertension (200/100 mmHg). Her Hb was 8.0 g/dl with evidence of microangiopathic hemolysis; plasma creatinine was 790 μmole/liter. Renal ultrasound demonstrated normal sized, unobstructed kidneys. She required urgent hemodialysis but remained anuric thereafter. Dynamic renal scintigraphy with 100 MBq ^{99m}Tc-MAG3 was performed after 2 days. She died 8 days later from progressive cardiac failure.

Case 3

A 53-yr-old man had presented 3 mo previously with dry itchy skin on hands and abdomen. After 1 mo, he developed arthralgia with joint swelling together with skin tethering over the dorsum of his fingers and nose. Over the 2 wk prior to admission, he became lethargic with progressive exertional dyspnea and, latterly, paroxysmal nocturnal dyspnea. There was no previous or recent history of hypertension. On the day of admission, he had noted reduced visual acuity and had not passed urine. On examination, he was in left ventricular failure with severe pulmonary edema, blood pressure 220/120 mmHg with Grade IV hypertensive retinopathy. Hemoglobin 6.5 g/dl with evidence of microangiopathic hemolytic anemia; creatinine level 795 μmole/liter. Hypertension was controlled acutely and thereafter with captopril. Ultrasound demonstrated normal sized, unobstructed kidneys. Dynamic renal scintigraphy with 200 MBq ^{99m}Tc-DTPA was performed after 4 days. Following initial hemodialysis, he was treated with continuous ambulatory peritoneal dialysis. After 6 mo, he began to pass urine with daily volumes increasing gradually to a maximum of 1 liter at 2 yr but has remained dialysis-dependent.

Scintigraphic Findings

In each patient, renal scintigraphy demonstrated poor renal perfusion with little or no uptake of tracer and absent excretion (Figs. 1 and 2). The scan for Patient 3 demonstrated a moderate arterio-venous fistula, an occasional sequel to renal biopsy. The control images are from a 28-yr-old woman who was assessed as a live-related transplant donor.

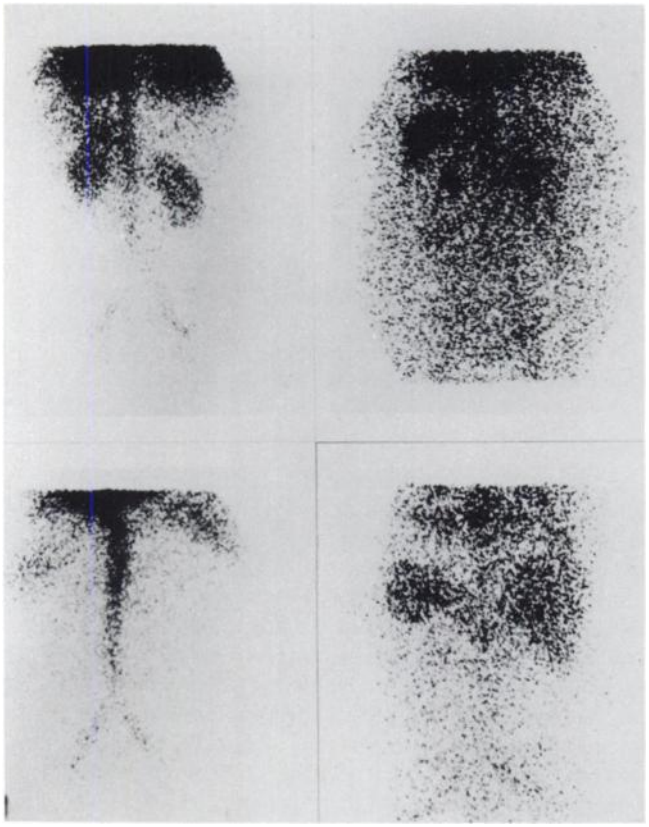

FIGURE 1. Scintigraphic appearance at 15 sec. Top left frame: Control and, then clockwise, Patients 1 to 3. Poor renal perfusion with little or no uptake of tracer is clearly demonstrated in the scleroderma patients.

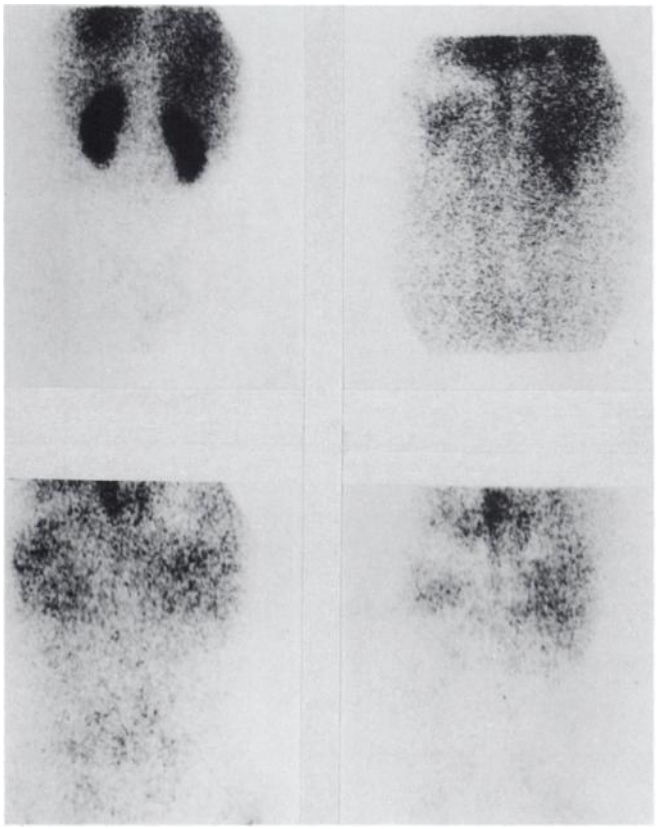

FIGURE 2. Scintigraphic appearance at 2 min. Top left frame: Control and, then clockwise, Patients 1 to 3. Uptake remains poor in the scleroderma patients, whereas the kidneys are now well defined in the control.

DISCUSSION

The scintigraphic findings in these three patients with acute scleroderma demonstrate almost complete loss of renal perfusion and parenchymal uptake. In each patient, this was associated with rapid onset of acute renal insufficiency and subsequent failure to achieve dialysis independence.

Sudden transformation to acute renal failure may affect 2%–8% of all patients with scleroderma, which often complicates disease of recent onset. Although usually heralded by symptoms and signs of severe uncontrolled hypertension, transformation may develop in patients with stable hypertension or even normal blood pressure (*1*). Identification of those at risk of transformation has not been successful although hyperreninemia may be a late sign (*2*). Established adverse prognostic indicators include age, male sex, hypertension, congestive heart failure, Grade III or IV hypertensive retinopathy and microangiopathic hemolytic anemia at presentation (*3*). The pathological processes are similar to those found in the malignant phase of essential hypertension and involve intimal proliferation and thrombotic occlusion of arterioles, small- and medium-sized arteries (*4*). Renal failure results from ischemia which may be so profound as to lead to renal cortical necrosis. The impairment of renal perfusion on scintigraphy in these patients is considerably more severe than that observed in most other forms of renal failure. The lack of parenchymal uptake with DTPA or MAG3 in our patients suggests a gross loss of renal function consistent with the underlying histology. There are no comprehensive series describing DTPA or MAG3 uptake in patients with acute renal failure. However, it appears that parenchymal uptake favors recovery (*5*), with the absence of uptake making recovery less likely but not impossible (*6*). This is consistent with the scintigraphic findings in our study.

The observation of high levels of renin occurring in scleroderma renal crisis has suggested a role for early treatment with angiotensin converting enzyme (ACE) inhibitors. Prompt introduction of an ACE inhibitor may significantly increase both the proportion of patients who become dialysis-independent together with the overall 1-yr survival (*3*). Treatment with an ACE inhibitor should probably be continued throughout the period of dialysis dependence and maintained thereafter.

In conclusion, poor perfusion and parenchymal uptake on renal scintigraphy at presentation of acute scleroderma may reflect the lack of potential for renal recovery in these patients.

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