11.28 Systemic Diseases and the Kidney

**RENEAL INVOLVEMENT IN SCLERODERMA**

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**FIGURE 11-50**

The main features of renal involvement in scleroderma are summarized. The major manifestation is the so-called renal crisis. Besides this often life-threatening manifestation, other patients may display milder forms of renal involvement, clinically characterized by mild proteinuria or slight deterioration of kidney function. Renal involvement is more common in patients with the diffuse form of scleroderma that is serologically characterized by antibodies against topoisomerase I or RNA polymerase III. Patients with progressive skin disease should be monitored carefully for hypertension and signs of renal involvement. Early institution of angiotensin-converting enzyme (ACE) inhibition in patients with micro-albuminuria can prevent further deterioration of kidney function [96,97]. ACE inhibition is also the mainstay of treatment for patients with scleroderma renal crisis, because it will significantly reduce progression to renal failure, increase the chance of recovery if renal failure has already developed, and improve the 1-year patient survival rate. Renal replacement therapy (hemodialysis or continuous ambulatory peritoneal dialysis) should be offered to patients whose renal function does not recover. The patient survival rate, however, is lower than in patients with other collagen-vascular diseases such as lupus nephritis. Limited experience with renal transplantation indicates that successful transplantation is possible, especially in patients with quiescent disease. Recurrence in the transplanted kidney has been reported [84]. References 96 to 98 provide more extensive reviews on the subject.

**FIGURE 11-51**

Scleroderma. In the acute phase, small- and medium-sized renal arteries show mucoid thickening of the intima with severe narrowing of the lumen. Sometimes these lesions are accompanied by thrombosis and fibrinoid necrosis of the arterioles and glomeruli. Morphologically, the vascular alterations resemble malignant nephrosclerosis (malignant hypertension) or hemolytic-uremic syndrome. In the chronic phase, the mucoid intimal material is replaced by fibrous tissue.

A. Severe narrowing of a small-sized renal artery owing to extensive endothelial widening with ischemia of glomeruli.

B. Accumulation of mucopolysaccharide material in the widened endothelial layer.

(Continued on next page)


