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SCLERODERMA RENAL CRISIS: A RENIN-DRIVEN HYPERTENSIVE EMERGENCY WITH DISTINCT RENAL MICROVASCULAR PATHOLOGY

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Scleroderma renal crisis (SRC) is the most serious kidney problem caused by systemic sclerosis. This is a type of acute kidney vascular injury that is characterized by malignant hypertension, quickly worsening kidney failure, and signs of thrombotic microangiopathy. From a pathological anatomy perspective, SRC primarily affects the renal microvasculature, marked by structural vascular alterations that precede and govern parenchymal damage.

In the early stages of SRC, the kidneys are usually normal or only slightly smaller. In the later stages, the capsule is tense and the surface of the cortex is finely granular. In cut sections, cortical pallor indicative of ischemia may be evident, whereas chronic instances exhibit cortical thinning resulting from ongoing vascular compromise. Nevertheless, macroscopic observations often appear subtle in contrast to the significant microscopic changes.

Microscopically, the hallmark lesions are confined to small arteries and arterioles, displaying significant concentric intimal thickening characterized by a distinctive “onion-skin” appearance resulting from myxoid and fibrous proliferation. Severe renal ischemia is caused by narrowing and blockage of the lumen. In later stages, fibrinoid necrosis of vessel walls and intraluminal thrombosis are clear signs of acute vascular injury. The changes in the glomeruli are mostly secondary and ischemic. They include the wrinkling and collapse of capillary loops and the occasional presence of thrombi in glomerular capillaries. Tubular structures exhibit characteristics of acute tubular injury, such as epithelial cell necrosis and the absence of the brush border, indicative of downstream hypoperfusion. The interstitium exhibits mild edema and restricted inflammatory infiltration, with fibrosis emerging in subsequent stages.

These morphological observations are associated with the underlying pathogenesis, wherein endothelial injury induces excessive activation of the renin–angiotensin–aldosterone system, resulting in severe vasoconstriction and the continuation of vascular damage. In clinical terms, this presents as hypertensive emergency, acute kidney injury, and microangiopathic hemolytic anemia, whereas proteinuria remains comparatively mild due to the lesion's primarily vascular characteristics. ACE inhibitors have greatly improved outcomes by blocking the renin-driven mechanism and breaking the cycle of ischemia and vascular injury. However, temporary dialysis is often needed.

Recent research delineates SRC as a quintessential renal thrombotic microangiopathy, influenced by endothelin-mediated vasoconstriction and potential complement activation. Consequently, SRC constitutes a distinct pathological entity characterized by primary vascular lesions that delineate both the histological presentation and clinical trajectory, underscoring the paramount significance of early diagnosis and targeted therapeutic intervention.