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Short Report

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Renal biopsy teaching case: A patient with scleroderma, hypertension, acute kidney injury and PR3 ANCA positivity

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Introduction

Scleroderma Renal Crisis (SRC) is the most common cause of Acute Kidney Injury (AKI) in scleroderma and occurs early during the disease course while ANCA Associated Vasculitis (AAV) is a rare cause of acute kidney injury occurring late in the disease course. We report a case of AKI with positive PR3 ANCA serology late in the disease course with a renal biopsy revealing findings of SRC.

Case report

A 65-year-old African-American male, with a history of limited scleroderma (SSc) for 16 years, complicated by severe gastrointestinal dysmotility and interstitial lung disease, presented to the clinic with elevated blood pressure of 190/100 mmHg, on a background of previously well-controlled blood pressure. He was found to have an acute kidney injury with a serum creatinine of 2.5 mg/dL, compared to his baseline of 1.3 mg/dL. Urine studies demonstrated microscopic hematuria, with 3.4 grams of proteinuria. Hemoglobin was 7.4 g/L with no evidence of hemolysis and normal platelet count. Serologies revealed a positive c-ANCA serology with PR3 positivity at 97.4 IU/mL with negative ANA, ds-DNA, Scl-70, anti-smith, anti-Ro, Anti-La, and RNP. A renal biopsy was performed which demonstrated arteriolar microangiopathy with fibrinoid necrosis and concentric lamellation with no evidence of ANCA GN (Figure 1A-G). He was treated with ACE inhibitor with improvement in blood pressure and serum creatinine (1.7 mg/dl).

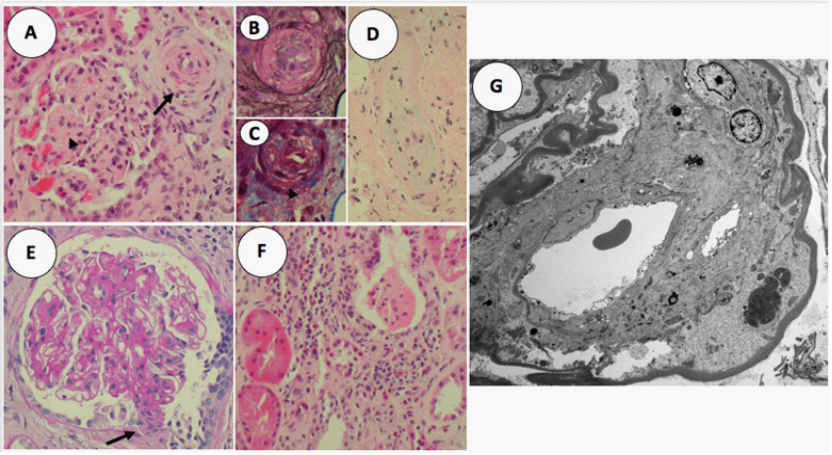

Figure 1: (A) Glomerulus (HE) and glomerular arteriole with entrapped RBCs (arrow) with vessel wall and surrounding interstitial.

edema and a consolidated ischemic glomerular appearance with potential entrapped RBCs (arrowhead).

(B and C) Glomerular arteriole (b, silver stain) shows lamination of the vessel wall and fibrinoid material entrapped within the vessel wall (Masson's trichrome.

(D) Small artery with subintimal mucoid change (HE).

(E) Glomerulus (PAS) with synechial adhesion and associated glomerular solidification.

(F) Foci of interstitial inflammation including eosinophils (HE).

(G) Ultrastructural evidence of microvascular injury including subendothelal widening with prominent cellular interposition.

Discussion

We report the first case of late onset SRC associated with PR3 ANCA positivity. Overlap syndrome of scleroderma and ANCA vasculitis is a well-recognized but rare phenotype. Majority of these overlap cases are MPO ANCA positive and vasculitic manifestation is universal in these cases [1,2]. Our case is unique in being PR3 ANCA positive without any vasculitic manifestation but with presentation as late onset SRC.

Derret-Smith and colleagues, demonstrated a possibility of shared HLA haplotypes with SSc/AAV, especially in the HLA-DQ region [2]. Further research in this area may reveal common immunogenetic factors involved in patients with Ssc/AAV overalap. Alternatively, PR3 ANCA in our patient could belong to a repertoire of natural antibodies [3].

This case of late onset scleroderma renal crisis highlights that atypical presentations of scleroderma renal crisis can exist, and concomitant ANCA positivity can be misleading in such situations. Therefore, it is imperative that if a scleroderma patient demonstrates atypical features- an expedient renal biopsy should be pursued.

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