



Onion Skin Lesion of the Renal Small Artery in a Patient With Polymyositis and Hemolytic Uremic Syndrome

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A woman in her 50s presented with muscle weakness and pain in the proximal muscles of the upper and lower limb, absence of skin lesions, and a typical muscle biopsy finding and was diagnosed with polymyositis. Prednisolone was started at a dose of 45 mg, and tacrolimus was started at a dose of 4 mg. Nine months later, the patient suddenly developed headaches and decreased vision in the left eye. The development of hemolytic uremic syndrome was noted with hemolytic anemia (hemoglobin, 9.9 g/dL) with schistocytes (5.9%), thrombocytopenia (41,000/ μ L), and reduced kidney function (serum creatinine, 4.21 mg/dL). Severe hypertension (210/120 mm Hg) was characteristic. Kidney biopsy showed an onion skin-like lesion from the interlobular arteries to the afferent arteriole and

vascular pole (preglomerular arteriole) characterized by advanced intimal thickening (hyperplasia) due to vascular endothelial cell proliferation and fibrosis (Fig 1A-C), and consequent severe narrowing of the vessel lumen and diffuse glomerular capillary collapse. Thrombosis was not observed. Immunofluorescence testing and electron microscopy did not reveal immune deposits. Lupus nephritis, antiphospholipid syndrome, or scleroderma renal crisis that can cause hemolytic uremic syndrome were excluded¹⁻³ This indicates that polymyositis is another disease that can cause hemolytic uremic syndrome. The only kidney biopsy image of a patient with hemolytic uremic syndrome due to polymyositis has been that of thrombotic microangiopathy with thrombus formation, as reported by Fukuda et al.⁴ However, our case was

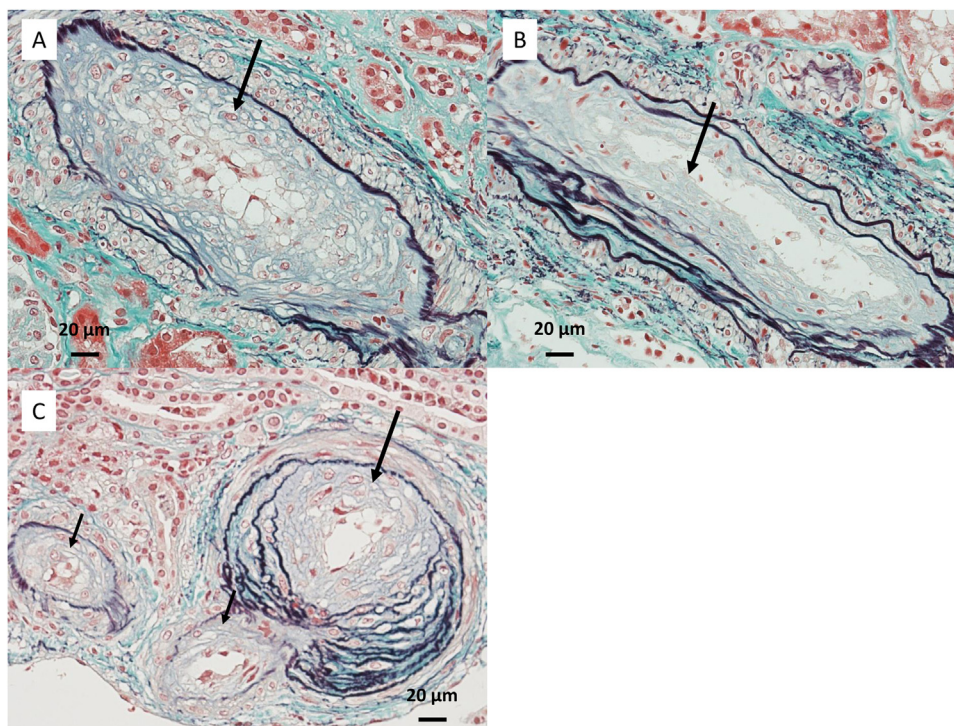


Figure 1. (A) Longitudinal image of an interlobular artery showing advanced intimal thickening (hyperplasia) due to endothelial cell proliferation (arrow) of the interlobular arteries. Masson trichrome staining; original magnification, $\times 400$. (B) Longitudinal image of an interlobular artery showing fibrosis of the interlobular arteries. Masson trichrome staining; original magnification, $\times 400$. (C) Transverse section of an interlobular artery and arteriole with onion skin-like intimal thickening of the interlobular artery (large arrow) and arteriole (small arrow). Masson trichrome staining; original magnification, $\times 400$.

different, as there was no thrombotic onion skin-like lesion of small renal arteries and consequent glomerular collapse, likely due to a hemodynamic mechanism related to accelerated hypertension.⁵

ARTICLE INFORMATION

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