TUBULOINTERSTITIAL NEPHRITIS AND PRIMARY BILIARY CHOLANGITIS (TIN AND PBC)

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36-year-old female patient was admitted to the hospital due to laboratory signs of **tubulopathy with reduced renal function** (eGFR CKD EPI 80 ml/min/1.73m²). She had a history of Hashimoto's thyroiditis and primary biliary cholangitis. Her bloodwork showed metabolic acidosis (due to dRTA) with hypokalemia. Urine was positive for glucose with A2 albuminuria and increased levels of alfa 1 macroglobulin.

The immunology was negative except for circulating anti- mitochondrial antibodies (AMA-M2). A kidney biopsy was performed. On light microscopy in 90% of the parenchyma, a medium dense, predominantly mononuclear infiltrate was found with varying degrees of tubulitis.

Infiltrate was composed of a mixed population of CD3 and CD 20 lymphocytes with some plasma cells (CD138+). There was mild interstitial fibrosis and tubular atrophy (7%). Plasma cells were positive for IgG, but negative on IgG4. Immunofluorescence and electron microscopy was unremarkable.

The diagnosis of tubulointerstitial nephritis was made, and the patient started taking glucocorticoid therapy (initially 30 mg of prednisone with gradual tapering throughout 4 months to the maintenance dose of 5 mg). During that period a slight improvement of renal function was noted (eGFR CKD EPI 87.2 ml/min/1.73m^2), with normalization of urine sediment and lowering of albuminuria

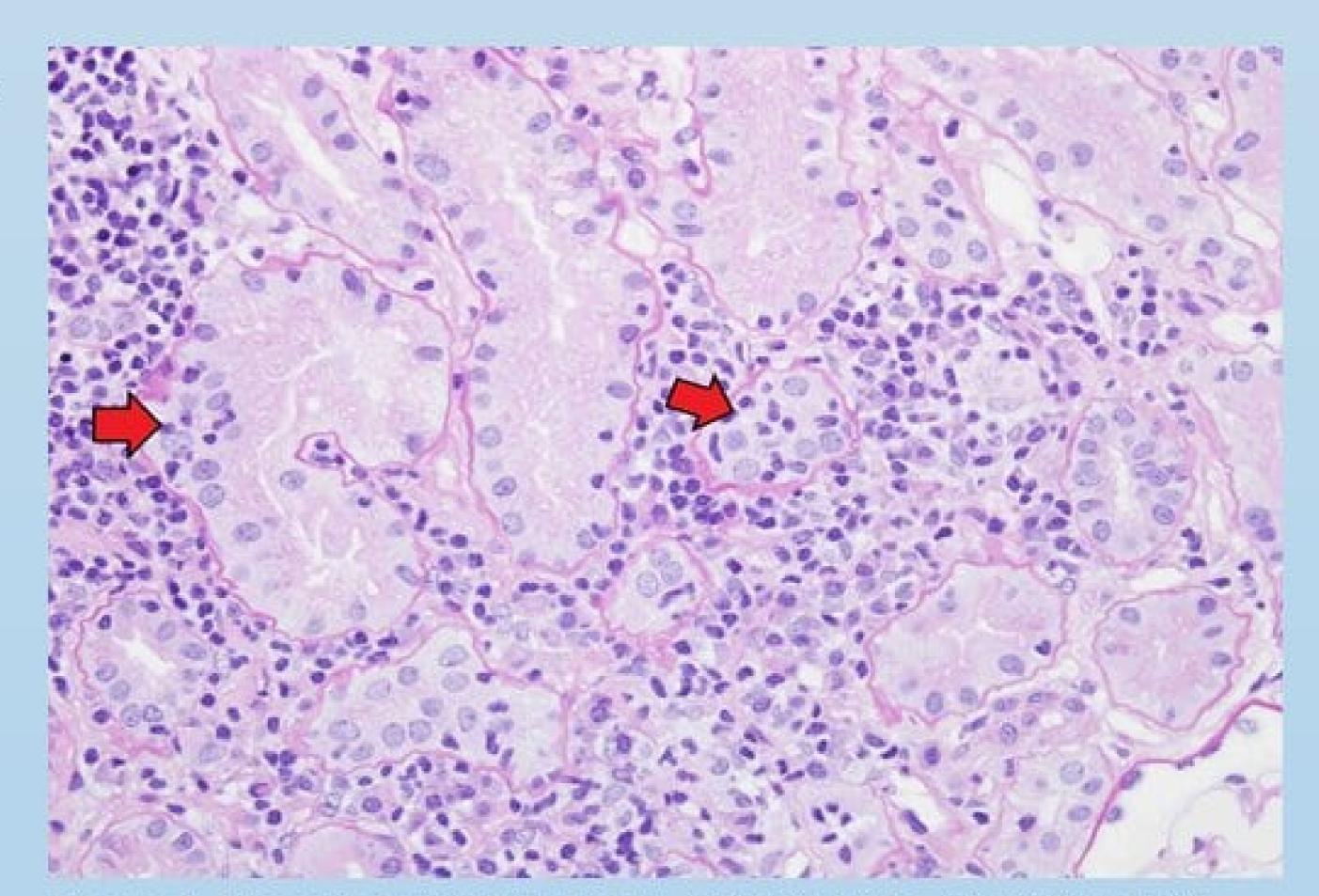


Image 1. Mononuclear inflammatory infiltrate in the interstitium with tubulitis (arrows).

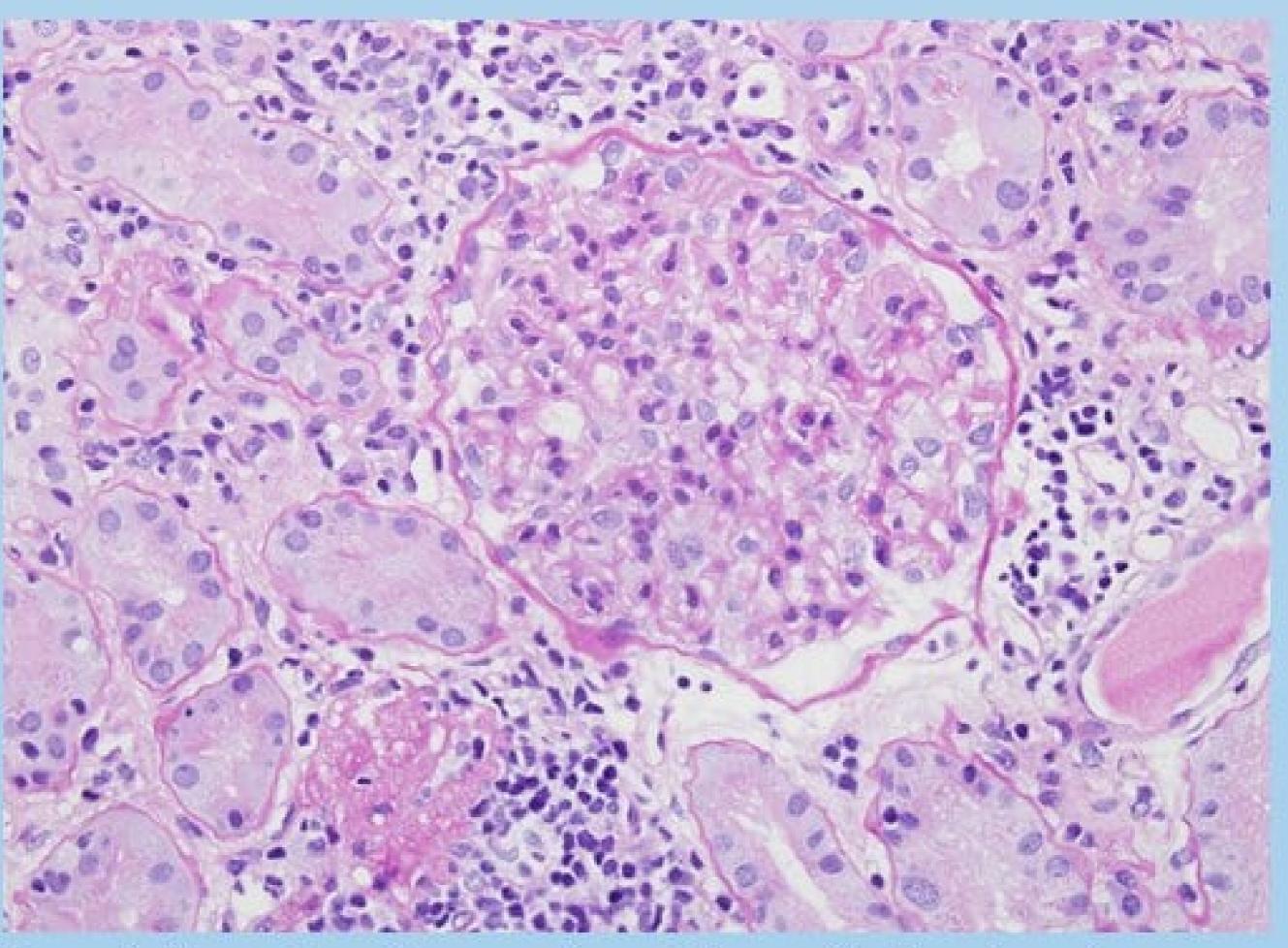


Image 2. Glomerulus with a normal morphology, globally sclerosed glomerulus and mononuclear inflammatory infiltrate in the interstitium. PAS staining, original magnification x400.

There are several cases of **primary biliary cholangitis linked with tubulointerstitial nephritis** and are all pertaining to women in the age range of 28 -77 years. In all of the cases the kidney function improved once they were started on glucocorticoid therapy, and in few cases, a deterioration of kidney function was noted when the glucocorticoid therapy was excluded. Although the relapse of tubulointerstitial nephritis is rarely seen, many patients do remain on maintenance therapy to sustain a stable kidney function.