CJASN®
Clinical Journal of the American Society of Nephrology

March 2021 • Vol. 16 • No. 3

Patient Voice

Editorials

Original Articles

Acute Kidney Injury and ICU Nephrology

Chronic Kidney Disease

Clinical Nephrology

Research Letters (Continued)

Erratum

Genomics of Kidney Disease

Kidney Case Conference: How I Treat

Perspectives

Feature

Review

On the Cover

What is the diagnosis?

A 72-year-old patient with recent history of systemic lambda light-chain AL amyloidosis under chemotherapy, malabsorptive diarrhea, and normal baseline kidney function was admitted for acute kidney failure. The laboratory tests showed that serum creatinine raised from 1.13 mg/dl to 3.97 mg/dl, and proteinuria was at 0.18 g/24h with a small passage of albumin and lambda monoclonal light chains in urine and normal serum light chains.

Image Description:

With light microscopy (left image: hematoxylin staining, middle image and right image: Congo red staining and polarized light; ×300), histopathology showed birefringent crystals in several tubules (arrowheads). Amyloid deposits were observed in medium size vessels (arrows) and at the vascular pole of a glomerulus (asterisk) and in scarcity in glomerular and tubule-interstitial compartments. Tubular atrophy was associated with interstitial fibrosis (25% of total surface).

Teaching Points:

These findings suggest the importance of considering acute oxalate nephropathy in the diagnosis of nonresolving acute kidney failure induced by diarrhea even in patients with another active kidney disease. The gastrointestinal amyloid infiltrations lead to lymphatic congestion resulting in lipid-rich lymph leakage into the intestinal lumen (1) ensuring a saponification process due to calcium, thereby oxalate free for absorption in the colon. Chronic dehydration, low calcium levels, and diarrheal loss of crystallization inhibitors such as magnesium and citrate favor crystallogenesis. The prognosis is poor, and the treatment consists in a low oxalate and fatty diet and correction of ionic disorders (2). Resumption of chemotherapy is important in order to regress amyloid infiltration in the digestive tract. Glomerular deposits exist in almost 97% of cases, leading to proteinuria in the nephrotic ranges and mainly composed of light chains (3). Vascular-limited deposits lead to low-grade proteinuria and much worse kidney prognosis because the lack of proteinuria may delay the diagnosis and the treatment; therefore, kidney biopsy should be performed.

References:

1. James DG, Zuckerman GR, Sayuk GS, Wang HL, Prakash C: Clinical recognition of AL type amyloidosis of the luminal gastrointestinal tract. *Clin Gastroenterol Hepatol*. 5: 582–588, 2007

2. Lumlertgul N, Siribamrungwong M, Jaber BL, Susantitaphong P: Secondary oxalate nephropathy: A systematic review. *Kidney Int. Rep*. 3: 1363–1372, 2018

3. Said SM, Sethi S, Valeri AM, Leung N, Cornell LD, Fidler ME, Hernandez LH, Vrana JA, Theis JD, Quint PS, Dogan A, Nasr SH: Renal amyloidosis: Origin and clinicopathologic correlations of 474 recent cases. *Clin J Am Soc Nephrol* 8: 1515–1523, 2013

(Text and images provided by Andrei-Radu Anghel, Department of Nephrology, CHU Dijon, Bourgogne, France; Georges Tarris, Department of Pathology, CHU Dijon, Bourgogne, France; Laurent Martin, Department of Pathology, CHU Dijon, Bourgogne, France and INSERM UMR 1098, Besançon, France; Denis Caillot, Department of Haematology, CHU Dijon, Bourgogne, France; Jean-Michel Rebibou, Department of Nephrology, CHU Dijon, Bourgogne, France and INSERM UMR 1098, Besançon, France; and Mathieu Legendre, Department of Nephrology, CHU Dijon, Bourgogne, France)