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T. Dwight McKinney

On the Cover

What's the diagnosis? Urine sediment examination under Bright field microscopy (400×) reveals ovoid and biconcave crystals within a cast, as seen in the upper left panel. The crystals within the cast are strongly birefringent under polarized light microscopy (upper right panel). This finding is consistent with calcium oxalate monohydrate crystal casts in the urine of a patient with primary hyperoxaluria type 1. Kidney biopsy reveals intratubular calcium oxalate crystals within the lumens of the tubules on Bright field microscopy (400×) in the left lower panel and strongly birefringent crystals on polarized light microscopy (400×) in the right lower panel. The urinary sediment findings nicely reflected the kidney biopsy findings. Primary hyperoxaluria type 1 is a genetic disorder that has deficiency of the hepatic peroxisomal enzyme alanine:glyoxylate-aminotransferase. This disorder is characterized by excessive production of oxalate results in increased plasma levels and hyperoxaluria. In the setting of hyperoxaluria, calcium-oxalate crystals precipitate in the renal tubular lumens and interstitium. Both acute and chronic oxalate nephropathy and calcium-oxalate stones develop from the associated hyperoxaluria. Patients frequently develop ESRD, requiring renal replacement therapy. A combined liver-kidney transplant is required to correct hyperoxaluria and prevent loss of the renal allograft. (Images and text provided by José Antonio Tesser Poloni, Irmandade da Santa Casa de Misericórdia de Porto Alegre, Porto Alegre, Brazil, and Mark A. Perazella, Yale University, New Haven, Connecticut)