Patient Voice

1777 Home-Based Care for CKD for High-Risk Populations
Wendy Rodgers
See related editorial and article on pages 1779 and 1801, respectively.

Editorials

1779 Novel Models for Health Care Delivery for CKD for Disadvantaged Populations
Joseph Lunyera and Clarissa Jonas Diamantidis
See related Patient Voice and article on pages 1777 and 1801, respectively.

1781 Monoclonal Gammopathies and Kidney Disease: Searching for Significance
Christina Hao Wang and Jonathan J. Hogan
See related article on page 1810.

1783 Still Asking “Which Rate Is Right?” Years Later
Tyler B. Woodell and Dena E. Rifkin
See related article on page 1842.

1785 Treatment of Drug-Induced Acute Tubulointerstitial Nephritis: The Search for Better Evidence
Dennis G. Moledina and Mark A. Perazella
See related article on page 1851.

1788 Does What Goes Around Always Come Around?
Howard Trachtman
See related article on page 1859.

Original Articles

Acute Kidney Injury and ICU Nephrology

1791 Acute Kidney Injury among Hospitalized Children in China
Xin Xu, Sheng Nie, Aihua Zhang, Jianhua Mao, Hai-Peng Liu, Huimin Xia, Hong Xu, Zhangsuo Liu, Shipin Feng, Wei Zhou, Xuemei Liu, Yonghong Yang, Yuhong Tao, Yunlin Feng, Chunbo Chen, Mo Wang, Yan Zha, Jian-Hua Feng, Qingchu Li, Shuwang Ge, Jianghua Chen, Yongcheng He, Siyuan Teng, Chuanming Hao, Bi-Cheng Liu, Ying Tang, Wenjuan He, Pinghong He, and Fan Fan Hou

Chronic Kidney Disease

1801 Home-Based Kidney Care, Patient Activation, and Risk Factors for CKD Progression in Zuni Indians: A Randomized, Controlled Clinical Trial
Robert G. Nelson, V. Shane Pankratz, Donica M. Ghahate, Jeanette Bobelu, Thomas Faber, and Vallabh O. Shah
See related Patient Voice and editorial on pages 1777 and 1779, respectively.

1810 Association of Monoclonal Gammopathy with Progression to ESKD among US Veterans
Nicholas Burwick, Scott V. Adams, Jeffrey A. Todd-Stenberg, Nilka Rios Burrows, Meda E. Pavkov, and Ann M. O’Hare
See related editorial on page 1781.
Chronic Kidney Disease (Continued)

1816 PTH, FGF23, and Intensive Blood Pressure Lowering in Chronic Kidney Disease Participants in SPRINT

1825 Sleep Quality and Sleep Duration with CKD are Associated with Progression to ESKD
Ryohei Yamamoto, Maki Shinzawa, Yoshitaka Isaka, Etsuko Yamakoshi, Enyu Imai, Yasuo Ohashi, Akira Hishida and for the CKD-JAC Investigators

Clinical Nephrology

1833 Medicare’s New Prospective Payment System on Facility Provision of Peritoneal Dialysis

Diabetes and the Kidney

1842 Facility-Level Variations in Kidney Disease Care among Veterans with Diabetes and CKD
See related editorial on page 1783.

Glomerular and Tubulointerstitial Diseases

1851 Duration of Treatment with Corticosteroids and Recovery of Kidney Function in Acute Interstitial Nephritis
Gema Fernandez-Juárez, Javier Villacorta Perez, Fernando Caravaca-Fontán, Luis Quintana, Amir Shabaka, Eva Rodriguez, Liliana Gadola, Alberto de Lorenzo, Maria Angeles Cobo, Añiana Oliet, Milagros Sierra, Carmen Cobelo, Elena Iglesias, Miguel Blasco, Cristina Galeano, Alfredo Cordon, Jesus Oliva, and Manuel Praga on behalf of the Spanish Group for the Study of Glomerular Diseases (GLOSEN)
See related editorial on page 1785.

1859 Adrenocorticotropic Hormone for Childhood Nephrotic Syndrome: The ATLANTIS Randomized Trial
See related editorial on page 1788.

Maintenance Dialysis

1866 Health Insurance in the First 3 Months of Hemodialysis and Early Vascular Access
Eugene Lin, Matthew W. Mell, Wolfgang C. Winkelmayer, and Kevin F. Erickson

Transplantation

1876 Procurement Biopsies in the Evaluation of Deceased Donor Kidneys

Research Letter

1886 Prevalence of Opioid, Gabapentinoid, and NSAID Use in Patients with CKD
Tessa K. Novick, Aditya Surapaneni, Jung-Im Shin, Shoshana H. Ballew, G. Caleb Alexander, Lesley A. Inker, Alex R. Chang, and Morgan E. Grams

Erratum

1889 Correction
Kidney Case Conference: Attending Rounds

1890 Lung Ultrasonography in the Acutely Dyspneic Hemodialysis Patient
Daniel W. Ross and Kenar D. Jhaveri

Kidney Case Conference: How I Treat

1893 BK Virus Nephropathy
Deirdre Sawinski and Jennifer Trofe-Clark

Nephropharmacology for the Clinician

1897 Pharmacology behind Common Drug Nephrotoxicities
Mark A. Perazella

Perspectives

1909 Addressing Disparities in Living Donor Kidney Transplantation: A Call to Action
Krista L. Lentine and Didier Mandelbrot

1912 Intravenous Fluids: Finding the Right Balance
Paul M. Palevsky

1915 Renal Fellow Network: Past and Future
Samira Farouk and Matthew A. Sparks

Features

1918 Helping More Patients Receive a Living Donor Kidney Transplant
Amit X. Garg

1924 Clinical and Regulatory Considerations for Central Venous Catheters for Hemodialysis
Douglas M. Silverstein, Scott O. Trerotola, Timothy Clark, Garth James, Wing Ng, Amy Dwyer, Marius C. Florescu, Roman Shingarev, and Stephen R. Ash on behalf of the Kidney Health Initiative HDF Workgroup

On the Cover
What's the diagnosis?
A 73-year-old woman with controlled hypertension and CKD (3B) insidiously developed abdominal and loin discomfort, weight loss, resistant hypertension and rapid decline of GFR. The physical examination was remarkable for hypertension and low-grade fever. A PET-CT scan unmasked a hypermetabolic lesion of the abdominal aorta at the level of D12-L3, further characterized on MRI (Figure 1) as an aortic wall irregularity causing luminal occlusion; the lesion extended to the renal ostia (mainly the left) and the kidneys were asymmetrical and poorly differentiated.

Differential diagnosis was vasculitis, atherosclerosis and rarer aortic neoplasm. The patient was considered unsuitable for endovascular procedure or surgery, given the extension of the disease. An empirical course of corticosteroids for vasculitis was tried. The patient deteriorated with hemodialysis dependent kidney failure, liver and intestinal ischemia and succumbed within days.

The autopsy study (Figure 2) revealed a vegetative and necrotic aortic neoplasm arising from the intima, with involvement by the contiguity of left renal artery; on hematoxylin and eosin stain (Figure 3), the luminal surface had loosely cohesive cells forming aggregates, with a high nuclei/cytoplasm ratio, scarce cytoplasm and marked pleomorphism. There was hepatic, intestinal and pulmonary microembolization.

Malignant renal artery stenosis is rare and has been described in patients with myeloproliferative neoplasms, retroperitoneal sarcoma and aortic intimal sarcoma. Clinical presentation is variable, often indistinguishable from infectious and noninfectious aortitis or atheroembolic disease. The radiomorphological pattern is nonspecific and diagnosis is commonly achieved only after resection. The prognosis is poor. When dealing with abdominal pain, uncontrolled hypertension and kidney failure, vascular disease should be suspected and ruled out.

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