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On the Cover

What’s the diagnosis? Positron emission tomography (PET) coronal scan (upper right) and the fused PET/CT coronal scan (upper left) demonstrate significant uptake in multiple lymph nodes and in the kidneys. The patient in this case developed a systemic illness characterized by fever, skin rash, diffuse lymphadenopathy, profound peripheral eosinophilia, mild transaminitis, altered mental status, and acute kidney injury following re-exposure to intravenous vancomycin. As seen here, renal biopsy revealed a granulomatous acute interstitial nephritis (AIN) (lower left) with a cellular infiltrate consisting of numerous eosinophils, lymphocytes, neutrophils, plasma cells, and macrophages (lower right). Lymph node biopsy was consistent with a reactive lymphadenitis. Vancomycin was discontinued, and the patient received intravenous methylprednisolone for 3 days followed by oral prednisone tapered over 4 weeks, with complete resolution of the systemic illness and recovery of kidney function back to baseline. This presentation is consistent with DRESS (Drug rash with Eosinophilia and Systemic Symptoms) syndrome, which is characterized by the presence of at least three of the following findings: fever, skin rash, peripheral eosinophilia, atypical circulating lymphocytes, lymphadenopathy, and hepatitis. DRESS syndrome often defies diagnosis initially as it has clinical features that mimic those found with other systemic disorders. This idiosyncratic reaction occurs most commonly after exposure to drugs such as allopurinol, sulfonamides, and aromatic anticonvulsants such as phenytoin, phenobarbital, and carbamazepine. Diagnosis essentially consists of recognizing the clinical syndrome and identifying the causative agent. Fused PET/CT scan in this case demonstrated a diffuse inflammatory process, including widespread lymph node enlargement and an exuberant granulomatous AIN. Treatment for DRESS syndrome includes discontinuing the suspected medication, and in some patients, treating with systemic steroids for several weeks. Surveillance for a relapse after cessation of symptoms and completion of steroids is required. (Image and text provided by Deepak Kadiyala, Gilbert Moeckel, and Mark A. Perazella, Yale University School of Medicine)