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On the Cover

What is the Diagnosis?

A 58-year-old female with a history of Samter's triad (asthma, chronic sinusitis, and aspirin allergy) and chronic hepatitis C presented with 2 months of worsening cervical lymphadenopathy. Blood work noted elevated creatinine from 0.7 to 1.3 mg/dl. Computed tomography of the abdomen revealed diffuse lymphadenopathy and bilateral kidney masses. A basic infectious and autoimmune workup including hepatitis, tuberculosis, Epstein-Barr virus, syphilis, and antinuclear antibody serologies were negative. A superficial cervical lymph node biopsy showed acute inflammatory histiocytosis, positive for S-100 staining and plasma cells with emperipolesis—a term describing the presence of intact lymphocytes engulfed in the cytoplasm of the histiocytes—which is a hallmark of Rosai-Dorfman disease (RDD). The patient was diagnosed with RDD, and no further kidney workup was pursued. The patient was started on 70 mg/d prednisone with taper and experienced resolution of imaging and laboratory abnormalities over the next 3 months.

Image Description:

Left image and center image show contrast-enhanced computed tomography scan with bilateral kidney involvement with low attenuation, 1–2 cm lesions, which efface normal kidney parenchyma and cortical ribbons, suggesting infiltrative masses. There is also diffuse retroperitoneal lymphadenopathy. And the right image shows various histiocytes with pale cytoplasm and engulfed lymphocytes (emperipolesis) suggestive of RDD.

Teaching Points:

Rosai-Dorfman disease is a benign disease with histiocytosis, massive lymphadenopathy, and multivisceral involvement that has excellent response to steroid therapy. Characteristic histological findings include S100 positive, large pale histiocytes with or without emperipolesis. Sixteen percent of all kidney masses are nonmalignant, yet their diagnosis leads to avoidable invasive procedures. RDD is often diagnosed after kidney biopsies or nephrectomies, but this can be prevented by maintaining a high index of suspicion for this benign pathology, particularly in patients with atypical presentation or chronic inflammatory conditions. This can help avoid unnecessary invasive procedures leading to precious tissue loss.

(Text and images provided by Moarij Qazi, Cleveland Clinic, Nephrology, Cleveland, Ohio; Huma Qazi, Army Medical College, Rawalpindi, Punjab, Pakistan; Sudipto Mukherjee, Cleveland Clinic, Cleveland, Ohio; Priyesh Patel, Cleveland Clinic, Radiology, Cleveland, Ohio; Ali Mehdi, Cleveland Clinic Foundation, Hypertension and Nephrology, Cleveland, Ohio; Jonathan Taliercio, Cleveland Clinic, Nephrology, Cleveland, Ohio; and Georges Nakhoul, Cleveland Clinic, Nephrology, Cleveland, Ohio)