Splenorenal Fusion Mimicking Renal Neoplasm in a Patient with von Hippel-Lindau Disease

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Picture 1. A heterogeneously contrast-enhanced mass in hilum of left kidney on computed tomography (arrow).

Picture 2. Surgical specimen showed a well-circumscribed dark red and soft nodule in hilum of left kidney.

Picture 3. (A) Histopathologic section showing splenic tissue on left and renal tissue on right. Note the nodule has a fibrous capsule (Hematoxylin and Eosin staining, ×25). (B) The sinusoid-like vascular lining cells are CD8 positive, which is characteristic of splenic sinusoidal structures. Positive cells are indicated by brown staining (arrow) (Immunohistochemistry, ×1,000).
We report a 49-year-old asymptomatic woman with von Hippel-Lindau disease presenting with hemangioblastoma in the central nervous system, pancreatic cysts and a heterogeneously contrast-enhanced mass in the hilum of the left kidney on computed tomography (Picture 1). She was underwent a total nephrectomy. Gross examination of the surgical specimen showed a well-circumscribed dark red and soft nodule measuring 3 cm × 3 cm × 2.5 cm (Picture 2). Microscopically, the nodule was partially encapsulated and was composed of fibrous cords separated by vascular channels with lymphocytes, hemorrhage, focal hemosiderin and scattered aggregates of lymphoid cells resembling splenic red and white pulp (Picture 3A). The immunophenotype of vascular channels was identical to splenic sinusoids. The vascular lining cells were CD8 and CD31 positive, which is characteristic of splenic sinusoidal structures (Picture 3B). The present patient had no history of splenectomy or splenic trauma. Heterotopic spleen is a rare developmental anomaly that results in the fusion of splenic and renal tissues and may present as a mass simulating primary renal neoplasm (1-3). Magnetic resonance imaging with ferumoxide and technetium sulfur colloid scan may characterize this mass as ectopic splenic tissue and an unnecessary nephrectomy may be avoided (4, 5). Immunohistochemistry confirmed the diagnosis (6).

References