Polycystic horseshoe kidney: dealing with double trouble

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A 45-year-old male presented with a clinical history of vague upper abdominal pain. There was no significant medical illness or any history of renal disease in the past. Clinical examination and laboratory parameters were unremarkable. A serum creatinine value of 0.83 mg/dL, serum sodium of 139 mEq/L and serum potassium of 4.2 mEq/L were reported. Contrast-enhanced CT study of the abdomen revealed a ‘horseshoe’ configuration of the kidneys with a thick pre-aortic parenchymal isthmus (Figure 1). The kidneys also showed multiple parenchymal cysts of varying sizes (Panels A and B). These cysts were seen to involve the cortical as well as medullary regions. The renal parenchymal enhancement and contrast excretion were preserved. No definite hydronephrosis, calculus or mass lesion was identified. Multiple small parenchymal cysts were also seen in the liver. No definite cysts were seen in pancreas or seminal vesicles. The diagnosis of polycystic horseshoe kidney was made, which was in fact an incidental detection, as the abdominal pain was ascribed to cholelithiasis and the renal functions were preserved.

Polycystic horseshoe kidney is a combination of two distinct renal disorders. Horseshoe kidney is a renal fusion anomaly during embryogenesis while autosomal-dominant polycystic kidney disease (ADPKD) is a hereditary disorder due to gene mutations. Horseshoe kidney is the most common of all renal fusion anomalies and is often asymptomatic. ADPKD is the third most common cause of end-stage renal disease. Polycystic horseshoe kidney is, however, extremely rare with only 20 cases reported in the literature. Concomitant presence of renal polycystic disease and horseshoe anomaly is likely to lower the age of renal failure and therefore warrants closer surveillance of these patients, despite the incidental diagnosis.

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