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A case of Bartter syndrome accompanied by de novo focal segmental glomerulosclerosis

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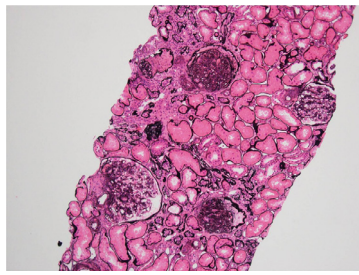
Introduction: Bartter syndrome is a rare inherited defect in thick ascending limbs of loop of Henle with mutations of genes encoding Na-K-2Cl co-transporters. Typical manifestations of Bartter syndrome are hypokalemia, metabolic alkalosis, hypercalciuria, nephrocalcinosis, and hyper-reninemic-hyperaldosteronism, but some studies report the cases with unusual clinical findings. Herein, we report an unusual case of Bartter syndrome with progressive renal dysfunction and proteinuria. **Case report:** A 42-year-old woman visited the clinic with generalized edema for a year after C-section due to preeclampsia. She was diagnosed with Bartter syndrome atypical manifestations mentioned above eighteen years ago. She has had hypothyroidism for five years, type 2 diabetes mellitus, and hypertension for three years. At admission, 24-hour urine total protein showed 2.45g, and serum creatinine level was 1.45mg/dL, compared with 1.09 mg/dL four months before admission. Renal ultrasonogram showed increased parenchymal echogenicity, and the size of left kidney was 8.7cm, and that of right kidney was 8.8cm. Kidney biopsy showed that there were 16 global sclerosis and 2 segmental sclerosis among 26 glomeruli, and there was no crescent formation, minimally increased cellularity with mesangial cell proliferation, and hyperplastic juxtaglomerular apparatus. Hyperplastic juxtaglomerular apparatus including renin-secreting juxtaglomerular cells is a typical finding in Bartter syndrome, and was accompanied by uncommon focal segmental glomerulosclerosis in our case. Taking angiotensin Ⅱ receptor blocker, proteinuria was reduced and renal function was maintained without deterioration. **Conclusion:** In the pathogenesis of Bartter syndrome, increased renin and angiotensin II levels can evoke both intra-glomerular hypertension and intrarenal activation of transforming growth factor-β, associated with interstitial proliferation and glomerulosclerosis. If a Bartter syndrome patient shows unusual clinical findings such as renal dysfunction and persistent proteinuria, kidney biopsy should be done to identify whether some glomerular diseases are accompanied like our case.



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A case of acute renal failure as a rare form of presentation of sarcoidosis

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Introduction: Sarcoidosis is a systemic disease characterized by noncaseating epitheloid granulomas in multiple organs. In korea, sarcoidosis is a very rare disease (0.125/100,000 in 1998) and renal involvement is uncommon feature of sarcoidosis. Some studies have reported that granulomatous tubulointerstitial nephritis caused by sarcoidosis is associated with acute renal failure and may lead to hemodialysis. Here we report a rare case of acute renal failure related to sarcoidosis. **Case report:** A 49-year-man with a previous history of pulmonary sarcoidosis for the last 11 years, visited the outpatient clinic with oliguria and general weakness. His laboratory blood test revealed acute renal failure with blood urea nitrogen of 53.6mg/dL and serum creatinine 8.24mg/dL. There was hypercalcemia of 13.5mg/dL. Hemodialysis was performed. A renal biopsy was performed and revealed non-caseating granulomatous interstitial nephritis compatible with renal sarcoidosis. He started corticosteroid therapy in the form of oral prednisolone 0.5mg/kg/day. In terms of follow-up, his renal function was recovered and maintained without hemodialysis. Renal involvement of sarcoidosis is still a very rare disease in Korea, but when acute renal failure and hypercalcemia were accompanied, renal sarcoidosis may be considered as one of the cause. **Keywords:** Sarcoidosis; Acute renal failure; Granulomatous interstitial nephritis