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Medullary Sponge Kidney Presenting as Hypokalemic Paralysis

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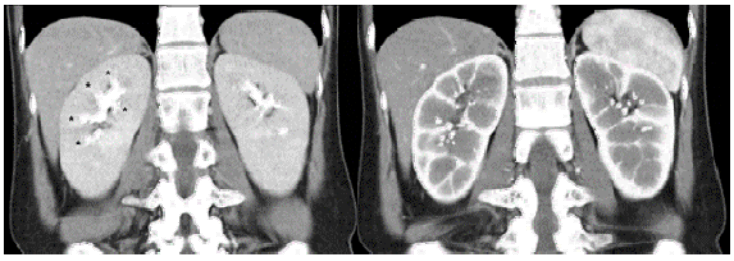
Hypokalemic paralysis features muscle paralysis with ictal hypokalemia. Hypokalemia is often associated with dRTA. While dRTA shares some autoimmune background, medullary sponge kidney (MSK) represents a anatomical abnormality associated with dRTA. We report a case of MSK as a rare cause of hypokalemia with RTA. A 33-year-old woman presented with cramps that developed into generalized weakness. She had no comorbidity and denied of any substance abuse. Despite a strong family history of SLE, she had no manifestation of SLE apart from mild episodes of weakness. Initial test revealed; K 2.2mEq/L, Na 136 mEq/L, Cl 112 mEq/L, Cr 1.0 mg/dl, and BST 109 mg/dl. The ABGA showed; pH 7.31, HCO3 9.6 mmol/L. Spot urine showed; 296mOsm/kg, Na 56 mmol/l, K 30mmol/l, Cl 62mmol/l, Cr 44.6mg/dl, and Ca 10.7mg/dl. TTKG was 9.2. Urine pH remained at 7, SG 1.008, and 5-9/HPF RBCs were observed. She had normotensive hypokalemia with a normal AG hyperchloremic metabolic acidosis, persistent alkaline and positive anion gap urine which were compatible with dRTA. CT showed MSK with nephrocalcinosis. Upon recovery of serum K+ level by supplement, she fully regained muscle power. MSK is seldom associated with hypokalemia and RTA. It is essentially a radiological diagnosis demonstrating a pathognomic 'brush' radiating outward from the calyces which signifies ectasia of the papillary collecting ducts. Because most of the patients are asymptomatic and thus a diagnosis is made rather incidentally, we suggest that kidney imaging be mandatory in the course of hypokalemia work up.



Contrast-enhanced CT scan of the abdomen showing 'bouquet of flowers' appearance (arrow) in the excretory phase and nephrocalcinosis.

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The Natural Course of IgA Nephropathy Presenting with Isolated Hematuria at the Time of Diagnosis

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Background: The outcome of patients with IgA nephropathy who present with isolated microscopic hematuria at the time of diagnosis is not well studied. **Methods:** We investigated the long-term prognosis of 162 patients with biopsy-proven IgA nephropathy, who presented with isolated microscopic hematuria, preserved renal function (estimated glomerular filtration rate (eGFR) ≥60 ml/min/1.73 m^2), and minimal or no proteinuria (spot urine protein/creatinine ratio ≤0.2). **Results:** Baseline characteristics were as follows. Mean age (SD) at the time of diagnosis was 37.9 (12.5) years, median eGFR (interquartile range) at the time of diagnosis was 97.3 (88.1-110.8) ml/min/1.73 m^2, and proteinuria level 0.08 (0.06-0.12). After a median follow-up period of 107 months, eGFR was 87.7 (77.9-101.9) ml/min/1.73 m^2 and proteinuria 0.07 (0.05-0.11). No patients progressed to advanced chronic kidney disease (eGFR below 30 ml/min/1.73 m^2). Significant proteinuria (spot urine P/C ratio ＞0.2) developed in 20 (12.3%) patients, and median proteinuria level in this group was 0.82 (0.33-1.36). When the group of patients who developed significant proteinuria was compared to the group of patients who did not, we found that follow-up median eGFR was significantly lower in the group with proteinuria compared to the group without proteinuria (79.1[69.4-99.1] vs. 89.3[78.9-102.1], $p=0.021$). There was no difference of baseline characteristics (underlying diabetes, hypertension, sex, age, baseline eGFR, and use of anti-hypertensive medication) between the 2 groups. On histopathological findings using the semiquantative classification, there was no significant difference of the median activity index score (total 10) and chronicity index score (total 12) between the 2 groups. **Conclusion:** Our study is one of the largest single-center studies to date on the prognosis of this cohort. As reported before, the long-term prognosis of IgA nephropathy patients with isolated hematuria at the time of diagnosis is excellent.