Medullary Spongy Kidney in a Middle-aged Woman with Primary Biliary Cirrhosis, Sjogren's Syndrome and Type 1 Renal Tubular Acidosis

Sir,

About 0.5% of the general population and 8.5% of individuals with nephrolithiasis have medullary spongy kidney (MSK), which is a rare congenital disorder defined by cystic dilatation of the terminal collecting ducts of the pyramids, giving a sponge-like look to the medulla.[1] One or both kidneys may be affected. The condition is typically asymptomatic, discovered incidentally when abdominal imaging is conducted for another reason. Nevertheless, it is also well-recognised that MSK poses a risk for hematuria, urinary tract infections, renal acidification, nephrocalcinosis, and nephrolithiasis.[2] Herein, we report a middle-aged woman who suffered from Sjogren's syndrome (SS), type I renal tubular acidosis (RTA), and primary biliary cirrhosis (PBC) and presented with signs and symptoms related to MSK.

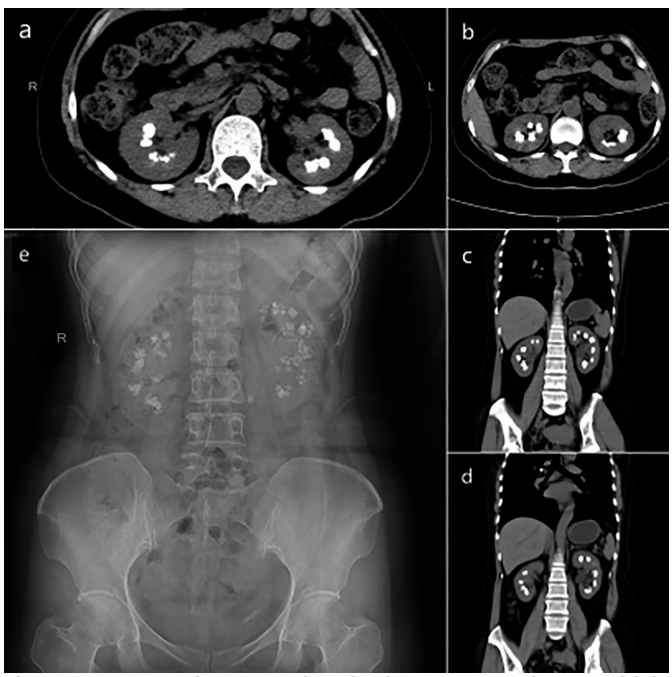

Figure 1: Computed tomography of urinary system shows multiple patchy high-density shadows in the bilateral renal sinusa, (a, b, c and d), abdominal X-ray shows multiple speckled high-density shadows in both renal areas (e).

A 47-year old woman was admitted to the hospital with complaints of dryness of mouth and eyes for 2 years and fatigue for 1 month. She had a past medical history of SS, type I RTA, and PBC. She had orally taken hydroxychloroquine, ursodeoxycholic acid, methotrexate, and a potassium citrate mixture. She arbitrarily discontinued hydroxychloroquine for half a year. Her physical examination on admission showed no abnormality. The main laboratory results are displayed in Table I. Computed tomography (CT) of the urinary system (Figure 1(a-d)) and abdominal X-ray (Figure 1e) indicated MSK. She was treated with methylprednisolone (30 mg, once a day), hydroxychloroquine (0.1 g, twice a day), ursodeoxycholic acid (250 mg, three times a day), and potassium citrate mixture (adjusted for blood potassium). The patient's discomfort of dryness and fatigue resolved significantly after 2 weeks. A follow-up examination showed a significant reduction in 24-h urine protein level, an increase in the plasma albumin level, and a remarkable improvement in general condition. The patient is currently undergoing regular long-term follow-up, and her condition is stable.

Table I: Results of laboratory tests.

	Normal range	Day 1	Day 7	Day 14
WBC (×10^9/L)	4-10	6.5	7.8	9.7
Hb (g/L)	110-150	138.5	142.1	146.1
Urine PH	5.0-6.5	7.5	7.0	6.5
Alb (g/L)	40-55	36.1	39.2	44.5
24hUpro (g)	0-0.15	1.25	1.05	0.72
UREA (mmol/L)	2.60-7.50	6.54	6.88	7.21
CREA (umol/L)	41.0-73.0	72.5	71.3	68.9
Potassium (mmol/L)	3.50-5.30	3.36	3.58	4.14
ANA	<1:100	1:1000	1:1000	<1:100
Anti-SS-A	-	+++	++	+
Anti-SS-B	-	+++	++	+
AM2A	-	++	++	+
ALT (U/L)	7-40	58.5	55.3	28.2
AST (U/L)	13-35	43.8	39.8	22.7
GLO (g/L)	20-35	37.85	35.47	32.41
γ-GGT (U/L)	40-150	167.8	158.2	148.2
IGG (g/L)	7.51-15.60	20.90	18.34	16.21
IGM (g/L)	0.46-3.04	4.15	3.84	3.18

*WBC (white blood cell), Hb (haemoglobin), Alb (albumin), 24hUpro (24 hours urine proteins), CREA(creatinine), antinuclear antibody (ANA), anti- Sjögren syndrome antigen A (SS-A), anti- Sjögren syndrome antigen B (SS-B), antibody Anti-mitochondrial M2 antibody (AM2A), alanine aminotransferase (ALT), glutamic-oxaloacetic-transaminase(AST), globulin(GLO), γ-Glutenoacetate aminotransferase(**γ**-GGT), immune globulin G(IGG), immune globulin M(IGM).*

PBC and SS are both autoimmune diseases and their specific aetiology is still not fully elucidated.[3] Moreover, PBC combined with SS is not uncommon in clinical practice and is more common in middle-aged and elderly female patients.[4] However, it is rare for PBC and SS to be complicated with RTA. SS is the most common cause of type I RTA.[5] Therefore, the type I RTA of this patient was considered to be secondary to her SS. The patient was diagnosed with MSK by CT. MSK is a congenital disease but manifests in adulthood. The pathogenesis of MSK is complicated and not fully understood. Usually, MSK is linked to higher levels of urine calcium in addition to normal urinary potassium and bicarbonate levels in the majority of cases.[2] The level of urine pH, ammonia, and titratable acidity in MSK patients is elevated.[1] The treatment mainly consists of glucocorticoids, immunosuppressive agents, and symptomatic supportive therapy. After several days of comprehensive treatment, the patient's condition improved significantly, and her condition was stable in outpatient follow-up.

In conclusion, to the best of our knowledge, this is the first case that reported a middle-aged woman with SS, type I RTA, and

PBC presenting as MSK, which showed significant improvement on appropriate treatment.

ETHICAL APPROVAL:
The patient provided her written informed consent to participate in this study. Written informed consent was obtained from the individual for the publication of any potentially identifiable images or data included in this article.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
XL: Manuscript writing, study design, and manuscript revision.
LS: Data and image collection.
SZ: Formulating patient's treatment.
All authors contributed to the article and approved the submitted version.

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