RENAL TUBULAR ACIDOSIS AS THE MAIN KIDNEY MANIFESTATION OF SJÖGREN'S SYNDROME: CASE REPORT

Abstract No: **WCN24-AB-1462**

Coello-Rivera, Solange[1]; Armas-Eguizábal, David[1]; Solares-Ovando, Sofia[1]; De León, Werner[2]; Palacios-Calderón, Oscar[3]; Moreno-Alvarado, Rodolfo[4],

[1]Internal Medicine Resident, Military Medical Center, Guatemala; [2]SERPAT, Private Pathology Laboratory; [3]Intensive Care Unit, Military Medical Center, Guatemala; [4]Nephrology Department, Military Medical Center, Guatemala

BACKGROUND

Sjögren's syndrome is an autoimmune disease. Kidney involvement can also be present in up to 33% of patients as renal tubular acidosis, nephrocalcinosis, and electrolyte disturbances.[1] Being tubulointerstitial nephritis the most common histopathological finding in kidney biopsy. [4]

CASE PRESENTATION

A 29-years old Guatemalan female with no past medical history, consulted to the emergency room with 2-years history of generalized weakness and weight loss. The patient disclosed polyuria, polydipsia, xerophthalmia and xerostomia.

Laboratories:

Glucose >800 mg/dl, potassium 3.8 mEq/L, sodium 131 mEq/L. calcium 8.5 mEq/L, phosphorus 1.6 mg/dl, chloride 101 mEq/L and magnesium in 2.46 mg/dl. Urine showed hematuria (10 cells/hpf), and renal blood tests had a creatinine value of 1.68mg/dl, (eGFR 42 ml/min/1.73m^2 by CKR-EPI) associated with proteinuria 290 mg/day and urinary anion gap 4.8. Venous blood gas analysis reported metabolic acidosis with normal anion gap. Due to the suspicion of autoimmune disease, we performed Schirmmer test, Anti-Ro, Anti-La antibodies and C3-C4, being all positive except for C3-C4. Bilateral renal USG reported normal size kidneys with bilateral medullary nephrocalcinosis.

Image 1

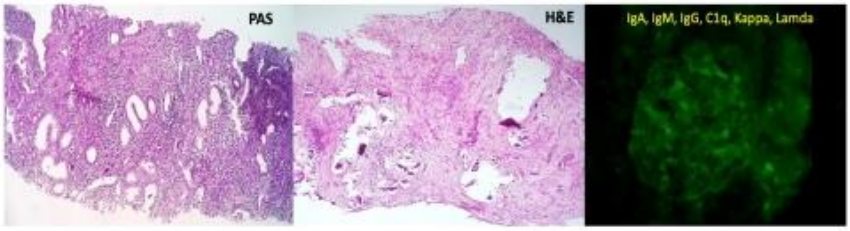


Image 2

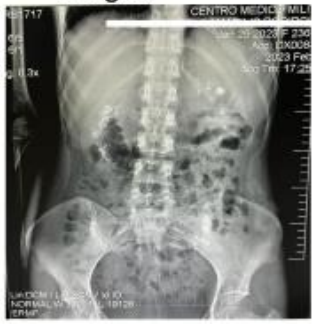
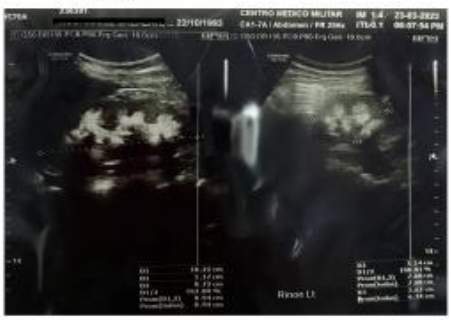


Image 3



CASE PRESENTATION cont.

Kidney biopsy:

Reported: 28 glomeruli, 9 with global sclerosis, mesangial expansion, lymphoplasmacytic infiltrate in the tubule-interstitium associated with tubular atrophy in 15-20%, with no endocapillary or extra capillary hypercellularity. Medulla with amorphous intratubular calcium deposits and negative immunofluorescence for IgG, IgA, IgM, C1q, Kappa, Lambda.

Clinical evolution:

It was determined the patient debuted with tubular acidosis secondary to Sjogren's Syndrome. The patient was medicated with prednisone (1mg/kg/day), azathioprine 50 mg day, and sodium bicarbonate 1gr TID. One week later, microproteinuria showed a 20% decrease (232 mg/day), metabolic acidosis ameliorated, and symptoms significantly improved. One month later the metabolic acidosis continued (HCO3std 14mmol/l), the patient was titrated up of oral sodium bicarbonate dose with clinical improvement.

CONCLUSIONS

Kidney involvement in Sjögren's syndrome occurs in 33% of cases as renal tubular acidosis. electrolyte abnormalities, AKI, nephrocalcinosis and tubulointerstitial nephritis as the main histopathological finding. [1-2] The finding of nephrocalcinosis and acute kidney injury is associated with poor prognosis. Treatment for kidney manifestations of SS consists in immunosuppressants for control of the underlying autoimmune disease but further investigation is needed to understand the best treatment.[3]

REFERENCES

1. Maripuri S, et al. Renal Involvement in Primary Sjögren's Syndrome: A Clinicopathologic Study. Clinical Journal of the American Society of Nephrology. 2009 Aug 13;4(9):1423–31.
2. Ramos-Casals, M., & EULAR Sjögren Syndrome Task Force. (2015). Characterization of systemic disease in primary Sjögren's syndrome: EULAR-SS Task Force recommendations for articular, cutaneous, pulmonary and renal involvements. *Rheumatology (Oxford, England)*, *54*(12), 2230–2238. doi.org/10.1093/rheumatology/kev200
3. Baenas, D. F., (2018). Nefrocalcinosis y acidosis tubular renal proximal en síndrome de Sjögren. *Revista de La Facultad de Ciencias Médicas*, *75*(2), 139. doi.org/10.31053/1853.0605.v75.n2.19161
4. Camino Benavides, (2017). Manifestaciones extraglandulares renales del síndrome de Sjögren. *Revista Cubana de Reumatología*, *19*(3), 126–131. http://scielo.sld.cu/scielo.php?script=sci_arttext&pid=S1817-59962017000300004