HYPOKALEMIC PERIODIC PARALYSIS - A RARE PRESENTATION OF DISTAL RENAL TUBULAR ACIDOSIS IN SJOGREN'S SYNDROME

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Introduction

- Sjogren's syndrome (SS) is a chronic autoimmune inflammatory disorder characterized by involvement of exocrine glands (lacrimal and salivary glands)
- Renal tubular acidosis (RTA) is one of the renal manifestations of SS
- We report unusual case series of SS with RTA presenting as hypokalemic periodic paralysis [1,2] without the components of Sicca syndrome.

Case Series

- Over a span of two years,3 middle aged females presented to our emergency department with acute onset of flaccid quadriparesis.
- The clinical, biochemical and serological features are shown in table 1.

All of them had features suggestive of Distal RTA

- profound hypokalemia with High urine spot K+, K+ /creatinine ratio and high TTKG
- Normal anion gap metabolic acidosis with positive urine anion gap and a high urine pH
- There was no glycosuria, phosphoturia or aminoaciduria

On Immune workup-

- ANA was positive and autoantibody profile showed positive SS-A and SS-B suggestive of Sjogren's syndrome.
- None of the patients had any glandular manifestation suggestive of Sicca syndrome and Schirmer test was negative.

- They underwent kidney biopsy in view of mild proteinuria which showed interstitial nephritis on light microscopy.
- serum electrophoresis was normal . Urine was negative for Bence Jones protein.
- A normal ultrasonographic and radiological examination of the kidneys excluded medullary sponge kidney, nephrocalcinosis, and obstructive uropathy as potential causes.

- They were treated with steroids and potassium supplements
- All of them recovered completely and are currently asymptomatic while on regular follow-up.

Table 1. Clinical, biochemical and serological features

Feature	Case 1	Case 2	Case 3
Age(Years)	27	40	41
Sex	Female	Female	Female
Duration of flaccid Quadriparesis(days)	3	5	7
Serum K+ (mmol/L)	2.6	1.5	2.2
Urine K+/Creatinine ratio (meq/g)	38	45	48
Spot Urine K+(meq/L)	26	61	25
TTKG	6	7	5
Anion Gap(meq/L)	8	10	11
Serum pH	7.35	7.17	7.3
Bicarbonate (meq/L)	15.2	9	10.5
Urine PH	6.5	7	7
Urine Anion gap	Positive	positive	positive
Aminoaciduria	Nil	Nil	Nil
Phosphoturia	Nil	Nil	Nil
Glycosuria	Nil	Nil	Nil
Dry mouth	No	No	No
Dry eye	No	No	No
Parotid gland enlargement	No	No	No
SCHIRMER test	negative	negative	negative
ANA	++	++	++
SS-A	++	++	++
SS-B	++	++	++
Ultrasound KUB	Normal	Normal	Normal
RA FACTOR	Negative	Negative	Negative
Serum creatinine(mg/dl)	0.7	1.1	0.7
Urine Proteinuria(g/day)	0.5	0.4	0.4
Kidney biopsy	Acute Interstitial nephritis	Acute interstitial nephritis	Acute on chronic Interstitial nephritis

Conclusion

- Primary SS should be considered in patients presenting with hypokalemic paralysis with RTA, even in the absence of apparent Sicca syndrome.
- We report our case series to increase awareness of this dramatic unusual presentation of SS which can be successfully treated with steroids and potassium supplements.

References

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