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Sjogren's syndrome presenting as a hypokalemic quadriparesis-a case report

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Abstract:
a 27-year-old female presented to the emergency department with complaints of difficulty in using both upper and lower limbs for the past two days. The patient was asymptomatic for two days after which she developed difficulty in using both her lower limbs after which she developed weakness in both her upper limbs. She had no complaints related to cranial nerves and her sensory system. however, she had a history of joint pain in all small and large joints for the past two years without any swelling. she had a history of a similar illness for which she was treated and her previous records were not available. she had complaints of dryness in her mouth for the past 2 years and dryness in her eye for the past 3 months. Physical examination showed decreased power in all four limbs and decreased reflexes in both upper and lower limbs. Routine blood investigations revealed hypokalemia. Which was corrected with intravenous potassium which resulted in reversing the weakness in all four limbs. On further investigations for dry mouth and eyes, the Schirmers test was performed which showed 4mm in 15 minutes. this further warranted ana profiling which was positive for ss-a. ss-b, ro-52 thus confirming the diagnosis of Sjogren's syndrome. This paper aims to emphasize on early diagnosis and management of Sjogren's syndrome to prevent further complications.

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I. Introduction

Sjögren's syndrome (SS) is a chronic autoimmune disease of unknown etiology, characterized by lymphocyte infiltration of exocrine glands resulting in dryness of mucosal surfaces including mouth (xerostomia), eyes (xerophthalmia), nose, pharynx, larynx, and vagina.[1] there are two types of Sjogren's syndrome they are primary and secondary Sjogren's syndrome. Middle-aged women (female-to-male ratio, 9:1) are primarily affected, although it may occur in all ages, including childhood. The prevalence of primary Sjögren's syndrome is approximately 0.5–1%, while 30% of patients with autoimmune rheumatic diseases suffer from secondary Sjögren's syndrome.[2]. various presentations include xeropthalmia, xerostomia, parotid gland enlargement, arthritis, Raynaud's phenomenon, renal tubular acidosis, vasculitis, lymphoma of these, renal tubular acidosis (RTA) may occur, causing a normal anion gap metabolic acidosis, a urine pH of >5.5, and hypokalemia. RTA is typically a type 1 distal tubule predominant in SS. Long-term RTA can result in nephrocalcinosis or nephrolithiasis. Therefore, a high clinical suspicion is needed to detect Sjogren's syndrome presenting as RTA.

II. Case Report

A 27-year-old female presented to the emergency department with complaints of quadriparesis for the past two days with a history of joint pain of all 4 limbs, both small and large joints, on and off, for the past 2 years without any swelling of joints. The patient noticed dryness of the mouth for the past 3 years and dryness in the eyes for the past 2 months. The patient was hospitalized with similar complaints (weakness of all 4 limbs) in the past 3 years, weakness of limbs improved after allopathic treatment (no records were produced). the patient had no other co-morbidities. On examination: Patient comfortable at rest, conscious, oriented, afebrile No pallor, No icterus, No cyanosis, No clubbing, No lymphadenopathy, and No pedal edema. BP -130/90 mmHg in the right upper limb in supine posture. PR -60/min regular rhythm, normal volume, and character, felt same in all peripheries, no radio-radial delay, no radio femoral delay, no vessel wall thickening. Spo2 -97% in room air RR – 16/ min

Motor system examination:

parameter	findings	right	left
bulk	UL, LL	normal	normal
tone	UL, LL	hypotonia	hypotonia
power	UL	3/5	3/5

	LL	2/5	2/5
Superficial reflexes		present	present
Deep tendon reflex	UL, LL	+1	+1

The sensory system examination was intact and within normal limits. cerebellar function tests could not be done due to weakness.
Examination of other systems was normal.

Table no 1: Salient Laboratory Investigations:

investigations	
hemoglobin	11.8g%
urea	32mg/dl
creatinine	0.9mg/dl
Total bilirubin	0.5mg/dl
Serum sodium, urine sodium	136mEq/l,88.9mEq/l
Serum potassium, urine potassium	2.36mEq/l,26.5mEq/l
Serum chloride	119mEq/l
Urine spot pcr	0.5
Urine ph	7.0

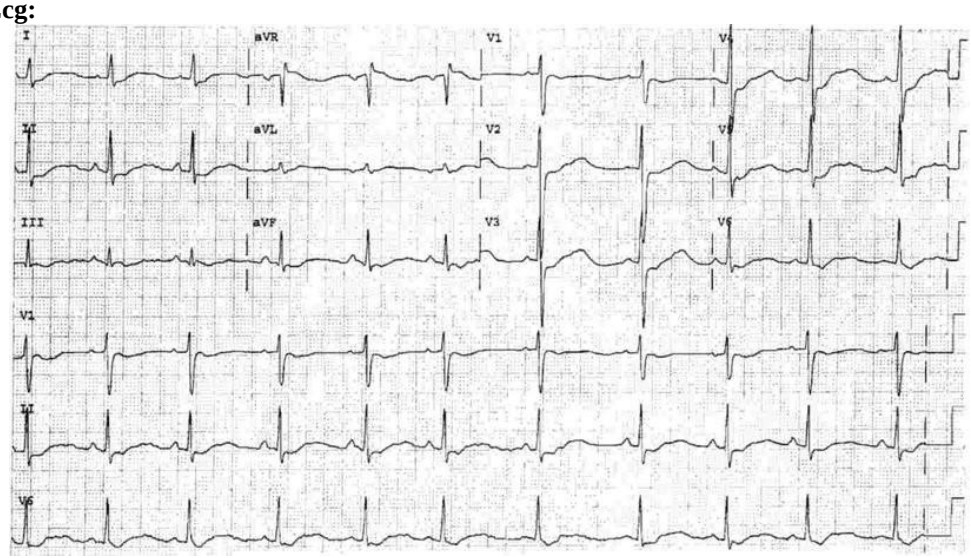
Table no 2: Abg:

pH	7.2
HCO3	7.8
pCO2	20
pO2	126
sO2	99
cSODIUM	140
potassium	2.1
calcium	103
Lactate	1.52

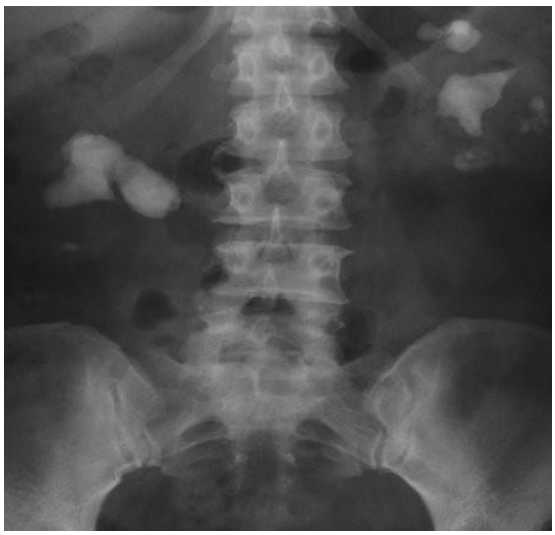
Table no 3: ana profile

ANA profile	
nRNP/8m	Positive (39.0)
sm	Negative (1.0)
SS-A	Positive (94.0)
Ro-52	Positive (106.0)
SS-B	Positive (42.0)
Scl-70	Negative (1.0)
PM-scl	Negative (0.0)
Jo-1	Negative (1.0)
CEMP-B	Negative (1.0)
PCNA	Negative (1.0)
ds DNA	Negative (0.0)
Nucleosome (NUC)	Negative (0.0)
Histone	Negative (0.0)
Rib P-protein	Negative (1.0)
AMA M2	Negative (1.0)

Ecg:



Ecg findings:
Prominent U waves, ST-segment depression, T wave flattening
Schirmers test: positive 4mm after 15 minutes
Radiological investigations: Xray –chest – normal lung fields
X-ray KUB-probably B/L nephrocalcinosis
USG abdomen-B/L intrarenal calculi, Staghorn calculi right kidney
CT –KUB --- multiple calculi seen in calyces of both kidneys largest measuring 1.8cm Right kidney,2.2 cm in left kidney Bilateral multiple renal calculi.



The above investigations pointed towards Hypokalemia with normal anion gap metabolic acidosis. Which was corrected with intravenous potassium after which the patients' general condition improved workup for xerostomia and xeropthalmia the patient was diagnosed with Sjogren's syndrome. The patient was discharged after advising appropriate medications and lifestyle modifications.

III. Discussion

Sjögren's syndrome is an autoimmune condition that typically involves lymphocytic infiltration of the salivary, parotid, and lacrimal glands, resulting in the characteristic symptoms of xerosis (dry eyes) and xerostomia (dry mouth). This immune process can also affect non-exocrine organs, such as the skin, lungs, gastrointestinal tract, and kidneys.[3] Although Sjogren syndrome commonly presents as dry eyes and dry mouth there are many extra glandular manifestations of this disease including arthritis, Raynaud's phenomenon, renal tubular acidosis, vasculitis, lymphadenopathy, and neuropathy. The renal involvement in primary Sjogren's syndrome is a well-known extra glandular manifestation with a prevalence of 5-14% in most European studies

[4] renal tubular acidosis can cause normal anion gap metabolic acidosis involving the distal tubules it can manifest as hypokalemic periodic paralysis, nephrocalcinosis. Glomerulonephritis is a rare finding that occurs in patients with mixed cryoglobulinemia, or systemic lupus erythematosus overlapping with Sjögren's syndrome [5]. the objective tests for sicca symptoms include Schirmer's test and Saxon's test are easy to perform, but their results do not correlate well with patient complaints and should be evaluated in the overall context. Here, cooperation with the ophthalmology department is crucial. The topical application of vital stains (lissamine green or fluorescein) is used to visualize and grade corneal and conjunctival lesions associated with keratoconjunctivitis sicca. Direct measurement of salivation is the diagnostic gold standard, but it is time-consuming in daily clinical practice. Parotid sialography and salivary gland scintigraphy lack sufficient specificity. As a non-invasive method, ultrasonography of the major salivary glands is an integral part of daily clinical practice [6]. Immunofluorescence testing for the anti-nuclear antibody is done for the diagnosis of Sjogren's syndrome. In patients with positive ANA titers, a fine speckled fluorescence pattern is strongly indicative of anti-Ro/SSA and/or anti-La/SSB antibodies, which is revealed in approximately 40% to 75% and 23% to 52% of primary Sjogren syndrome patients, respectively [7] Other serological abnormalities include the presence of rheumatoid factors (60–75%) as well as polyclonal hypergammaglobulinemia as a sign of increased B cell activity.[8] Despite continued advances in our understanding of the mechanisms involved in the pathogenesis of the disease, targeted treatment of Sjögren's syndrome is not available at present. Treatment is decided on an individual basis according to disease activity and the presence and extent of extra glandular manifestations. In patients with secondary Sjogren's syndrome, the indication for treatment is based on the underlying disease. In general, treatment should be provided by an interdisciplinary team, including family physicians, rheumatologists, ophthalmologists, ETN specialists, as well as dentists. Subject to the organ(s) involved and the presenting symptoms, consultation with other specialists (gynecologists, pulmonologists, neurologists, etc.) may be required. Disease-modifying therapy is reserved for patients with extra glandular involvement. To measure systemic disease activity, the EULAR Sjögren's Syndrome Disease Activity Index (ESSDAI) was developed and validated [9] Due to the presence of variable clinical presentation of this disease a high amount of clinical suspicion is required for the correct diagnosis of Sjogren's syndrome.

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