



A CASE OF HYPOKALEMIC PERIODIC PARALYSIS SECONDARY TO SJOGREN'S SYNDROME

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KEYWORDS :

INTRODUCTION:

Periodic paralysis (PP) is characterized by episodes of painless muscle weakness, which occurs episodically. PP is classified as hypokalemic (most common) or hyperkalemic (rare) with autosomal dominant inheritance.

Secondary causes of PP is often due to distal renal tubular acidosis (RTA); major causes being Sjögren's syndrome and familial hypercalciuria. Thus, adults with seemingly idiopathic distal RTA should be evaluated for Sjögren's syndrome.

PRESENTATION:

53 years old lady presented with vomiting for 1 month, muscle cramps and myalgia, weakness of limbs for 1 week. On examination, she was hemodynamically stable. Systemic examination revealed decreased muscle tone and grade 1 power in all 4 limbs. On further enquiry she gave history of dry eyes, dry mouth and dry skin of 2 years duration and episodic parotid enlargement of 1 year duration. ABG and serum electrolytes revealed hypokalemia(2.49), hyperchloremia, metabolic acidosis(pH-7.34, pCO2-20) with compensatory respiratory alkalosis, normal anion gap. Urine biochemistry showed increased spot K+(33.2 mEq/L) and Cl-(79.1 mmol/L). Her ESR was 37, CRP 2.9, RF 21 and CCP was negative. ENA test showed positive ANA, anti SSA ab, anti SSB Ab, anti Ro ab. Schirmer test was positive. TFT was normal.

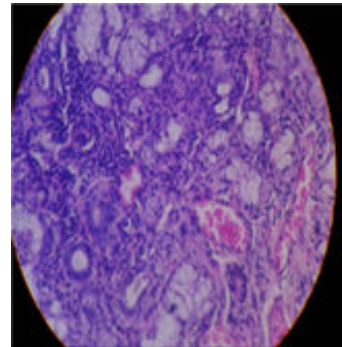


Figure 2: Salivary gland infiltrates with congested vessels, compatible with lymphoepithelial cyst.



Figure 3: Ecg Hypokalemia/ T wave inversion.

Baro	765.8 mmHg
Temp	37.0 °C
A/F	adult
pH	7.349 (-)(7.350-7.450)
PO2	120.4 mmHg(+)(60.0-100.0)
PCO2	20.0 mmHg(-)(35.0-45.0)
Na	141.9 mmol/L(135.0-148.0)
K	2.49 mmol/L(-)(3.50-4.50)
Cl	Slope nOk 1074
iCa	Not activated 1070
SO2	1P Error 1076
Hct	34.1 %(-)(35.0-50.0)
tHb	1P Error 1076
chCO3	10.8 mmol/L
ctCO2(P)	11.4 mmol/L
SO2(c)	98.2 %
BE	-12.4 mmol/L
BEeef	-14.9 mmol/L
BB	35.7 mmol/L
ctO2	20.9 Vol%
ctCO2(B)	9.5 mmol/L
pHst	7.184
chCO3st	14.5 mmol/L
H+	44.8 nmol/L
PAO2	127.9 mmHg
AaDO2	7.5 mmHg
a/AO2	94.1 %
RI	6 %
niCa	Missing data 1007
AG	Missing data 1007
pHT	7.349
H+t	44.8 nmol/L
PCO2t	20.0 mmHg
PO2t	120.4 mmHg
PAO2t	127.9 mmHg
AaDO2t	7.5 mmHg
a/AO2t	94.1 %

Figure 1: ABG Profile

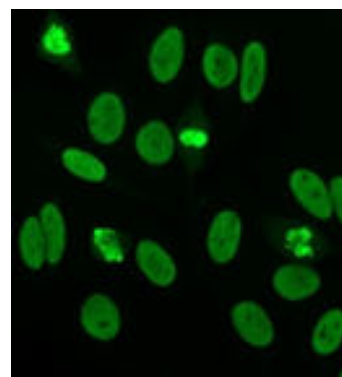


Figure 4: ANA cytoplasmic pattern.

MANAGEMENT:

Patient was managed with HCQs, Shohl's solution, artificial tears, moisturizers for dry lips, calcium and vitamin D supplements. Sjögren's syndrome (SS) is a chronic, multisystem autoimmune disease characterized by lacrimal and salivary gland inflammation, with resultant dryness of the eyes and mouth and occasional glandular enlargement. In addition, a variety of systemic manifestations may occur, including fatigue, musculoskeletal symptoms, rashes, and internal organ (eg, pulmonary, renal, hepatic, and neurologic) disease. Some patients present with a normal anion gap

metabolic acidosis due to distal RTA without a prior diagnosis of Sjögren's syndrome. So in any adult with unexplained distal RTA autoimmune disorders need to be ruled out.

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2. Bastani B, Haragsim L, Gluck S, Siamopoulos KC. Lack of H-ATPase in distal nephron causing hypokalaemic distal RTA in a patient with Sjögren's syndrome. *Nephrol Dial Transplant* 1995; 10:908.