ORIGINAL RESEARCH PAPER

Volume-8 | Issue-5 | May-2019 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

RENAL TUBULAR ACIDOSIS LEADING TO QUADRIPARESIS: A RARE PRESENTATION OF PRIMARY SJOGREN'S SYNDROME.

Nephrology

Prabhakar Yadav Consultant, Department of Nephrology, Tata Main Hospital, Jamshedpur

ABSTRACT

Primary Sjogren's syndrome (pSS) is characterized by lymphocytic infiltration of exocrine glands leading to reduced saliva and tear production. Spectrum of renal involvement in pSS is broad like tubulointerstitial nephritis(TIN), renal tubular acidosis(RTA),Fanconi syndrome and glomerulonephritis. Clinical presentation includes muscle weakness, cramps, periodic paralysis, bone pain due to electrolytes disturbances and metabolic acidosis. We describe here two cases of pSS presented as quadriparesis. Both cases had severe hypokalemia with normal anion gap acidosis due to distal renal tubular acidosis and positive antiSS-A and anti SS-B autoantibodies .Early diagnosis of pSS, prevents life threatining complications.

KEYWORDS

Sjogren's syndrome, Renal tubular acidosis, Hypokalemia, Quadriparesis

Introduction:

pSS is a chronic inflammatory , slowly progressive auto immune disease characterized by dryness of mouth and dry eyes.[1] It predominently affects women and involved all age groups. About one third of patients with pSS had extraglandular involvement and can manifest as Raynaud's phenomenon, vasculitis, lymphoma and various others organs involvements, including kidneys.[1]The prevalence of pSS is apporoximately 0.5 to 1.0%.[1]

Case-1

A29 year young female presented in our emergency department with complain of generalised weakness, tingling sensation all over body for three days. She had past history of similar illness six months back. There was no history of trauma, seizure, and ingestion of carbohydrate rich diet, dryness of mouth, grity sensation over eyes and joint pain. On physical examination patient was afebrile, pallor, icterus absent and no oedema. Her pulse was 90/ min, regular, blood pressure was 130/80 mmHg and respiratory rate was 14/min. Systemic examination revealed flacid quadriparesis with power of grade 1/5 in all four limbs.Deep tendon reflexes were absent. There was no bladder and bowel involvement , other systems did not reveal any abnormality. Laboratory reports are shown in Table-1. She had severe hypokalemia with normal anion gap hyperchloremic metabolic acidosis, suggestive of distal RTA. She was managed with injectable potassium chloride and sodabicarb followed by oral medications. She was doing well with oral sodium bicarbonte and Potassium chloride for one year, on susequent follow up she presented with dryness of mouth and joint pain. On further evaluation she was found to have anti-SS-A and anti-SS-B positive suggesting pSS. She was stared with tablet Prednislone (1mg/kg) tappered and stopped in six months. She was also started with tablet hydroxychloroquine for her joint pain as immunomodulating agent .She became asymptomatic after six months of treatment and she is in remission at present .

Case-2

A 39 year female admitted with complain of vomiting for four days associated with weakness all four limbs for 3 days. She had similar symptoms 2 months back . There was no history of trauma, seizure, and ingestion of carbohydrate rich diet, dryness of mouth,grity sensation in eyes and joint pain. On physical examination patient was afebrile, pallor present, icterus absent and no oedema. Her pulse was 80/ min, regular, blood pressure was 140/80 mmHg and respiratory rate was 16/min. Systemic examination revealed flacid quadriparesis with power of grade 1/5 in all four limbs.Deep tendon reflexes were absent. There was no bladder and bowel involvement, other systems did not reveal any abnormality. Laboratory reports are shown in Table-1. She had severe hypokalemia with normal anion gap hyperchloremic metabolic acidosis, suggestive of distal RTA. On further evaluation she was found to have anti-SS-A and anti-SS-B positive suggesting pSS. She was managed with injectable potassium chloride and sodiumbicarbonate followed by oral medications.Since she had no joint pain and sicca symptoms, we did not gave her steroids and any immunomodulatng agent.She was doing well with oral poatssium chloride and sodium bicarbonate.

Table-1

Laboratory parameters	Case-1	Case-2
Haemoglobin	9.6	10.0
Total leucocyte count	10000	14200
Platelets	1.18	3.1
ESR	64	54
Creatinine	0.9	0.81
Phosphorus	2.0	1.5
Calcium	9.9	9.1
Electrolytes(mEq/L)	Sodium-150, potassium-1.4	Sodium-148, potassium-1.5
Liver enzymes	AST-44.9,ALT-28.1.ALP-107.9	AST-34.2,ALT-21.2,ALP-104
ABG	pH-7.18,chloride-129, bicarbonate-12.8,pO2-108, pCO2-16.2	pH-7.27, chloride-127, bicarbonate-13.0,pO2-118, pCO2-19.2
Urine pH	6.5	6.8
Urine routine	Normal	Normal
USG-abdomen	Normal study	Normal study
ANA	Positive	Negative
Anti DsDNA	Negative	Negative
Anti-SS-A and Anti –SS/B	Positive	Positive
Anti-HbsAg,anti-HCV,HIV 1&2	Negative	Negative
Schimer's test	Negative	Negative

ESR:

Erythrocyte sedimentation rate,ABG: Arterial blood gas,USG: Ultrasound sonography,ANA: Antinuclear antibody.AST: Aspartate aminotransferase,ALT: Alanine aminotransferase,ALP:Alkaline phosphatase. Anti DsDNA: Anti double stranded DNA.

Discussion:-

pSS is a rarest cause of hypokalemic periodic paralysis. One third of patients with pSS have systemic manifestation at the time of presentation.[2]Spectrum of renal involvement in pSS includes RTA(distal and proximal), tubular proteinuria,diabtese insipidus,Gitelman's and Barter's syndrome, glomerular disease and renal failure.[3,4] pSS can presents with systemic symptoms before sicca symptoms evolved. Prevlance of renal involvement in pSS are 5-14% in most european studies.[5,6] Distal RTA is most frequent tubular abnormalties in pSS.[7] Tubular involvement is mainly due to TIN and antibodies against tubular transporters.[8]The mechanism of distal RTA in pSS is incompletely understood. Several case reports with immunocytochemical analysis on renal biopsy showed a complete absence of the H+ATPase pump in the intercalated cells of the collecting tubules, that is largely responsible for distal proton secretion.[9,10] The exact mechanism of immune injury leads to loss of H+ATPase activity is not known. Autoantibody directed against carbonic anhydrase II has been proposed as another mechanism of distal RTA in pSS.[11]Hypokalemia is the most common electrolytes abnormality in distal RTA.[2] Severe hypokalemia leading to quadriparesis has been reported in few cases only. In India, Rao et al

reported 31 cases of hypokalemic periodic paralysis, pSS was the cause in three of them only.[12] Sporadic cases are also reported from other centres.[13,14] Hypokalemia induced quadriparesis as the first manifestation of pSS is a rare entity.

Conclusion:

RTA leading to quadriparesis is a rare manifestation of pSS, sometime can be in the absence of sicca symptoms, so in young females presented with isolated qudriparesis without exocrine manifestations ,we should always rule out pSS to prevent fatal complications.

Conflict of Interest: None

REFERENCES:

1. Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson JL, Loscalzo J Harrison's principles of internal medicine, editors. Sjögren's Syndrome. 18th ed. New York: McGraw-Hill; 2012. pp. 2770–2773.
2. Shiboski SC, Shiboski CH, Criswell L, Baer A, Challacombe S, Lanfranchi H, et al. American College of Rheumatology classification criteria for Sjögren's syndrome: A data-driven, expert consensus approach in the Sjögren's International Collaborative Clinical Alliance cohort. Arthritis Care Res (Hoboken) 2012;64:475–87. [PMC free article] [PubMed]
3. Chan JC, Alon U. Tubular disorders of acid-base and phosphate metabolism. Nephron. 1985;40:257–79.[PubMed]
4. Francois H, Mariette X. Renal involvement in primary Sjogren syndrome. Nat Rev Nephrol. 2016;12:82–93. [PubMed].
5. Goules AV, Tatouli IP, Moutsopoulos HM, Tzioufas AG. Clinically significant renal involvement in primary Sjogren's syndrome: clinical presentation and outcome. Arthritis Rheum. 2013;65:2945–2953.[PubMed]
6. Seror R, Ravaud P, Bowman SJ, et al. EULAR Sjogren's syndrome disease activity index: development of a consensus systemic disease activity index for primary Sjogren's syndrome. Ann Rheum Dis. 2010;69:1103–1109. [PMC free article] [PubMed]
7. Ram R, Swarnalatha G, Dakshinamurty KV. Renal tubular acidosis in Sjögren's syndrome: A case series. Am J Nephrol. 2014;40:123–30. [PubMed]
8. Gerhardt RE, Loebl DH, Rao RN. Interstitial immunofluorescence in nephritis of Sjögren's syndrome. Clin Nephrol. 1978;10:201–7. [PubMed
9. DeFranco PE, Haragsim L, Schmitz PG, Bastani B. Absence of vacuolar H(+)-ATPase pump in the collecting duct of a patient with hypokalemic distal renal tubular acidosis and Sjögren's syndrome. J Am Soc Nephrol. 1995;6:295–301. [PubMed]
10. 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;10:908–9.[PubMed]
11. Pertovaara M, Bootorabi F, Kuuslahti M, Pasternack A, Parkkila S. Novel carbonic anhydrase autoantibodies and renal manifestations in patients with primary Sjogren's syndrome. Rheumatology (Oxford) 2011;50:1453–7. [PubMed]
12. Rao N, John M, Thomas N, Rajaratnam S, Seshadri MS. Aetiological, clinical and metabolic profile of hypokalaemic periodic paralysis in adults: A single-centre experience. Natl Med J India. 2006;19:246–9.[PubMed]
13. Palkar AV, Pillai S, Rajadhyaksha GC. Hypokalemic quadriparesis in Sjogren syndrome. Indian J Nephrol. 2011;21:191–3. [PMC free article] [PubMed]
14. Naik M, Bhat T, Naqash M, Qadri M, Yusuf I, Ali I, et al. Hypokalemic quadriparesis in an elderly female. Indian J Nephrol. 2012;22:402–3. [PMC free article] [PubMed]