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Integrated Approach in A Case of Sjogren's Syndrome with Distal Renal Tubular Acidosis presenting as Hypokalaemic Paralysis and Arthritis with Dry Eyes and Dry Mouth –A Case Study

Dr.Prashant Joshi [1]

1. MD Kaya-Chikitsa Scholar , Annasaheb Dange Ayurvedic Medical College , Ashta , Sangli

Dr.Sharat Kolke[2]

2. MD(Medicine) , DNB (Medicine), MNAMS. Consultant Physician , Fauziya Hospital, Kurla West.Mumbai 400070

Dr.Deepak Deshpande[3]

3. Professor and HOD of Kaya-Chikitsa , Annasaheb Dange Ayurvedic Medical College , Ashta , Sangli

ABSTRACT:

Sjogren's syndrome is a Chronic , Slowly Progressive Autoimmune Disease characterized by Lymphocytic Infiltration of the Exocrine Glands and B Lymphocyte Hyperactivity. The Disease may present alone (Primary Sjogren's Syndrome) or in Association with other Autoimmune Rheumatic Diseases(Secondary Sjogren's Syndrome). The Purpose of this case is to Highlight one of the under-reported presentations of Sjogren's Syndrome that could be easily missed without the Sicca symptoms. Distal Renal Tubular Acidosis (RTA) (Type 1 RTA) Secondary to Sjogren's Syndrome as a cause of Hypokalaemic Paralysis has not been emphasised enough to be widely known in Clinical practise .Also,Approach with drugs with Nil side effects is the need for Today's Era.So, in this case study, apart from Injection KCl(For Potassium Replacement) and Syrup Potassium Citrate(For Potassium and Bicarbonate replacement), I had Used Combination of Ayurvedic Drugs(For Managing Distal RTA- Arthrits-Dry Eyes- Dry Mouth) and Potassium Rich Diet(Aahara)(for managing Hypokalaemia).The above Integrated approach showed a

significant reduction in Symptoms Clinically and Improvement in Lab Reports. Further studies are to be Conducted on this as the present Research Poster is a Single Case Study . Trial in a Larger Sample is Required to Generalise the outcome.

Keywords - RTA-Renal Tubular Acidosis

INTRODUCTION

The Exact cause is unclear, it is believed to involve a Combination of Genetics and an Environmental triggers such as Exposure to Virus(EB virus,HCV , Human T-Cell Leukemia virus-1) or Bacteria. The Disease was described in 1933 by Henrik Sjogren after whom it is named.Middle Aged women (female-to-male ratio 9:1) are primarily affected , although Sjogren's syndrome may occur at any age, including Childhood.

The Prevelence of Primary Sjogren's Syndrome is ~0.5-1%, while 30% of patient's with Autoimmune Rheumatic Diseases suffer from Secondary Sjogren's Syndrome.It has been clinically observed that Ayurveda helps in Treating a case of Sjogren's Syndrome. It seems to help by correcting the Altered Immune System. Ayurvedic Medicines are very Effective in Treating Symptoms of Sjogren's Syndrome by Anti-Aam, Deepan-Pachana, Anti-Vata , Anti-Inflammatory , Nephro-Protective , Anti- Oxidants, Rasayana, Immuno-modulatory action .In this Case Study, Ayurvedic Drugs and Potassium Rich Diet are used As An Add on Therapy to Injection Potassium Chloride and Syrup Potassium Citrate For Managing Distal Renal Tubular Acidosis(Type1), Arthritis , Dry Eyes and Dry Mouth Secondary to Sjogren's Syndrome Presenting with Hypokalaemic Paralysis

Aims and Objectives

1) To Study About Sjogren's Syndrome , it's Clinical Manifestation's And Management in Detail

2) To Access the Effect of Combination of Ayurvedic Drugs and Potassium Rich Diet as an Add on Therapy to Intravenous Potassium Chloride(diluted) and Oral Potassium Citrate in Distal Renal Tubular Acidosis(Type1), Arthritis, Dry Eyes and Dry Mouth Secondary to Sjogren's Syndrome with Severe Hypokalaemia (causing Hypokalaemic Paralysis)

CASE PRESENTATION

58 years old Female Hindu patient Presented to Fauziya Hospital ICCU with

c/o Acute Onset of Bilateral Lower Limb Tingling and Numbness Followed by Bilateral Lower Limb Weakness (since 12 hrs before Admission) followed by

Bilateral Upper Limb Weakness (since 2 hrs before Admission).

H/O Nausea (since 3 to 4 days), Generalised Weakness , Fatigue (since 1 week)

H/O Dryness of Eyes (since 1 month)(on treatment – Carboxymethylcellulose eye drops)

H/O Dryness of Tongue and Increased Thirst (since 1 month)

H/O Back pain , Lower Abdominal Pain , Cloudy urine , Painful Urination (Since 1 month)

Sx/H/O CA Breast (Right Radical Mastectomy done 24 years back)(Received Chemotherapy during that peroid) (PET scan done 1 year back –s/o No active Benign or Malignant lesion)

H/O Allergy to Sulpa drugs

NO H/O DM / HTN/ IHD / RHD/ B.A./ COPD / CVA

General Examination : (Table No. 1)

Built	Thin	SPO2	100 %
Nourishment	Malnourished	HGT	88 mg / dl
Temperature	98.0 F	Height	5'7'' inches
Pulse	54 / min	Weight	48 kgs
Respiratory Rate	18 / min	Tongue	Uncoated, Dry
BP	90/60 mmHg		

No Pallor / Icterus / Cyanosis / Clubbing / Koilonychia / Edema / Lymphadenopathy

Table No. 2: Systemic Examination

RS	AEBE
CVS	S1, S2 (+) , Bradycardia
CNS	conscious , oriented ,Obeying to Verbal Commands , Responding to D.P.S
Plantars	Bilateral Flexors
DTR's	All ABSENT (ie Absent B/L knee jerk ,Ankle jerk , Achilles Tendon Jerk, Biceps jerk , Triceps jerk , Brachioradialis , Finger jerk)
Pupils	Bilaterally Normally Reacting to Light
Powers	Right Left UL 3/5 3/5 LL 2/5 2/5
P/A	Soft , Non Tender , L0S0K0

Investigations

1) Serum Potassium – 1.42 mEq/l (Normal Range-3.5-5.5)
2) ESR- 120 (Normal Range-0-30)
3) Serum Calcium- 7.5 mg/dl(Normal Range-8.5-10.4)
4) Serum Albumin- 2.3 gm/dl(Normal Range-3.5-5.0)
5) Serum Creatinine- 1.4 mg/dl(Normal Range-0.6-1.4)
6) TFT – WNL
7) Urine R and M- Albumin/Protein +++, Rest WNL.
8) CRP- 8.2(Normal Range- 0-6)
9) ABG- Metabolic Acidosis(pH- 7.310, pCO2-26,pO2-96.4, HCO3(A)-12.8 , HCO3(S)-15.6, O2SAT- 96.9. Serum Anion Gap Calculated – 10 (Normal Range- 3-11)
10) ANA- Positive (Pattern- Speckled)(++) (Titre 1:320)
11) Serum for ANA BLOT 17 Antigens – Positive for SSA, SSB and Ro52kD s/o Sjogren's Syndrome

12) ENA(Extractable Nuclear Antigen) Profile :

SSA-RO(Soluble Substance) IgG --Positive (166.95 Ru/ml) (Normal Range- Positive>20)

SSB-La (Soluble Substance) IgG—Positive(>200 Ru/ml)(Normal Range- Positive>20)

13) Beta2- Microglobulin- 5115 ng/ml(Normal Range- 670- 2143)
14) Serum Protein Electrophoresis s/o Hypergammaglobulinemia (Gamma Globulin- 2.34g/dl)(Normal Range 0.71-1.54)
15) Urine for 24 hours Proteins – 764 mg/24 hrs (Normal Range - <149.1)
16) Urine Protein-Creatinine Ratio- 1.03 (Normal Range-0-0.2)
17) ECG s/o Sinus Bradycardia with Evidence of U waves and Slight Prolongation of QRS complexes.
18) USG s/o Normal Sized Bright Kidneys. Rest WNL
19) 2D ECHO – WNL . LVEF-60%
20) CXR – WNL
21) Renal Biopsy – Not done in this case
22) SchirmerTest – showed Positive results (<10 mm of wetting in 5 min)

Differential Diagnosis

The Finding of Severe Hypokalaemia with Episode of weakness followed by Paralysis and the Favourable Response to Potassium Replacement by Intravenous Potassium Chloride ,Oral route Potassium Citrate and Potassium Rich Diet(Fruits , Juices , coconut water) , Ruled out Any Psychiatric causes like Conversion disorder, in our patient with no Psychiatric history . The Finding of Excessive Loss of Potassium in the urine (Renal loss) in the setting of Normal Anion Gap Metabolic Acidosis pointed towards a diagnosis of Distal RTA (Renal Tubular Acidosis) . The Presentation of Sicca Symptoms helped us Establish a Cause-Effect Relationship between Sjogren's Syndrome and Hypokalaemia

Table No.3 :Treatment

Treatment A	Treatment B	Treatment C
1) Intravenous Potassium Chloride (diluted) (Infusion) + 2) Oral Potassium Citrate10 ml TDS diluted in Full glass of Water + 3) Coconut Water QDS (Treatment A given for first 3 days)	1) Oral Potassium Citrate 10 ml TDS (diluted in full glass of water) + 2) Coconut water TDS+ 3) 1Banana 3 times a day (Treatment B given from 4th day to 6th day)	1) Oral Potassium Citrate 10 ml BD (diluted in full glass of water) (From Day 7th to Day 90th) + 2) Coconut water OD(From Day 7th to Day 90th)+ 3) 1 Banana 3 times a day + (From Day 4th to Day 90th)+ 4) Aam Pachana Vati 500 mg 1 Vati 3 times a day for 1 week (For Deepana-Pachana-AamPachana) (ie From Day 7 to Day 13th) followed by 5) Shunthi Churna 3 gms 3 times a day with warm water for 1 week(From Day 14th to Day 20th) + 6) Maha-Vata Gajankush Rasa 250 mg 1 BD (From Day 21st to Day 35th) + 4) Giloy Ghanvati 500 mg 1 BD(From Day 21st to Day 90th) + 5) Chandraprabhavati 500 mg 1 BD (with Milk)+ (From Day 21st to Day 90th)+ 6) Haridra churna(3gms) With Ashwagandha Churna(3gms) (with milk) BD (Treatment C

		given From 7th day to Day 90th)+ 7) Triphala Ghrita Instillation into eyes (HS)(From Day 7th to Day 90th) 8) Adequate Hydration with Water (10 glasses of Water/ Day)

Table No 4 :Lab Reports- Observations

Investigations	Observations After Treatment A	Observations After Treatment B	Observations After Treatment C
1) ESR	120	118	90
2) Serum Potassium Level	3.30 mEq/l	3.6 mEq/l	4.6 mEq/l
3) Serum Creatinine	1.4 mg/dl	1.4 mg/dl	1.2 mg/dl
4) ABG	Metabolic Acidosis (Moderate)	Metabolic Acidosis (Mild)	WNL
5) Urine Albumin	++++	++++	+++
6) RA factor	Positive (Stongly)	Positive (Strongly)	Positive(Moderately)
7) eGFR	41 ml/min/1.73 m2	41 ml/min/1.73 m2	49 ml/min/1.73m2

Table No 5 :Clinical Observations

Clinical Features	Observations After Treatment A	Observations After Treatment B	Observations After Treatment C
1) Dryness of Eyes	Present(Severe)	Present(Severe)	Present (Minimal)
2) Dryness of Mouth	Present(Severe)	Present(Severe)	Present(Minimal)
3) Early Morning Stiffness of Joints	Present(Severe)	Present (Severe)	Absent
4) Joint Redness	Present (Minimal)	Present (Minimal)	Absent
5) Joimt Pain-Tenderness	Present(Severe)	Present(Severe)	Present(Minimal)
6) Joint Swelling	Present(Severe)	Present(Severe)	Present(Minimal)
7) Joint Mobility-Motility	Decreased(Moderately)	Decreased(Moderately)	Increased
8) Lower Abdominal Pain	Present	Present	Absent
9) Froth in Urine	Present	Present	Occasionally Present
10) Burning Micturation	Present	Present	Absent

(Source: Primary Data)

Results:

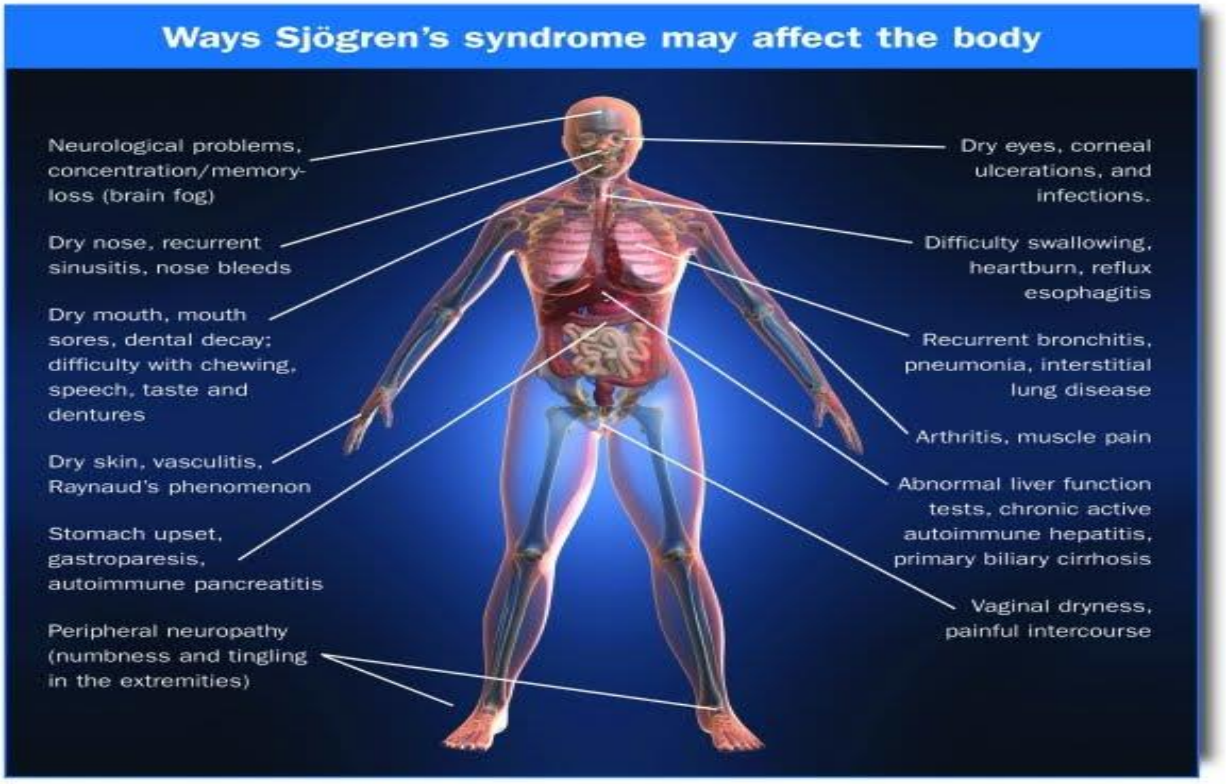
Symptoms Had Been Significantly Decreased in the past 3 months and Repeat Potassium Levels , ABG and Serum Creatinine during the Recent Followup were Within Normal Limits .

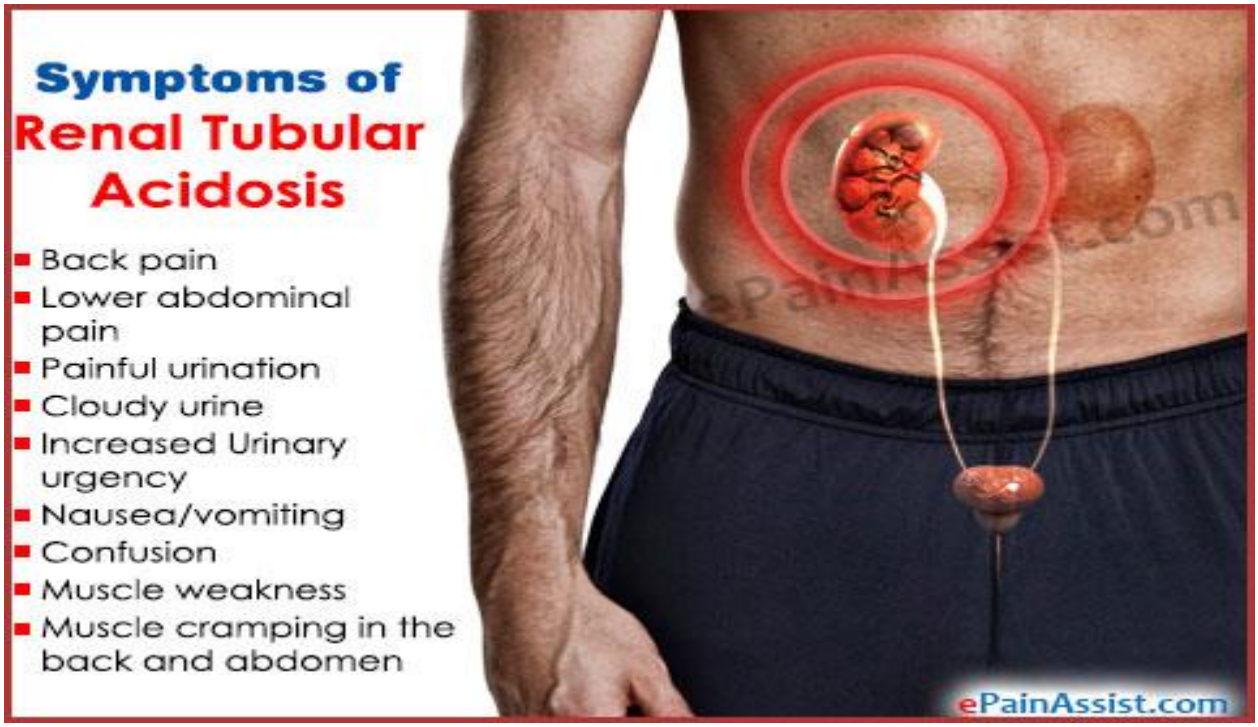
Discussion:

Sjogren's Syndrome is a Systemic Auto-Immune Disorder Characterised by a Unique Set of Signs and Symptoms Pre-dominantly caused by a Cell-Mediated Auto-immunity against Exocrine Glands. Extra-Glandular Manifestations of the Disease Arise from a Similar Pathogenic Mechanism affecting the Kidneys , Liver , Lungs , Pancreas , and the Nervous System. Renal Involvement in Sjogren's Syndrome has been known for a Long Time and Has Become one of the most commonly Encountered Extra-Glandular Manifestation .

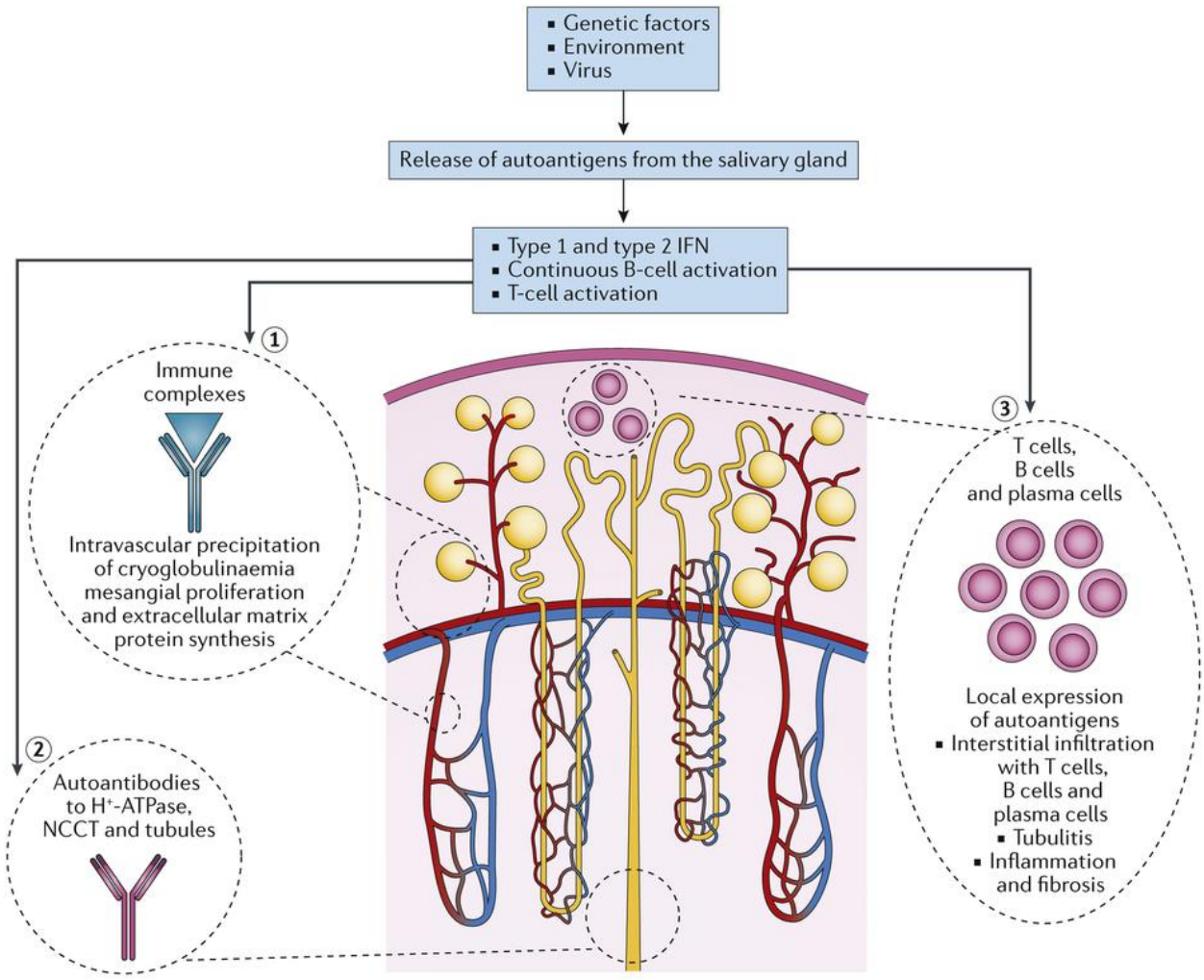
Distal RTA is Characterised by an Inability of the Kidneys to Excrete Hydogen Ions in the Urine in a Setting of Metabolic Acidosis Resulting in Inappropriately Normal or Alkaline Urine. The Condition can be Diagnosed by Normal Anion-Gap Metabolic Acidosis and Positive Urine Anion- Gap . Renal Potassium Wasting Leads to Hypokalaemia. Although Distal RTA is very commen in Sjogren's Syndrome , it is usually Asymptomatic and goes undetected in most cases.

According to Modern Medicine,Combination of Corticosteroids, and other Anti-Inflammatory drugs and Immunosuppresants(Mycophenolate mofetil) have Been Reported to Slow Progression of Renal Damage in Sjogren's Syndrome , But there Are lots of Adverse Effects of Steroids Therapy and Immunosuppresants Therapy. So, as per this Case Study it is concluded that above used Ayurvedic drugs are Anti- Aam, Deepana-Pachana, Nephro-Protective , Anti-Inflammatory Drugs , Immuno-Modulators , Rasayana, Anti- oxidants which Acts on Pathophysiology of Sjogren's Syndrome and on Kidneys and are free from Adverse Effects and are Effective as an Add on Therapy To Potassium Rich Diet And Potassium Citrate Syrup in managing Sjogren's Syndrome with Distal Renal Tubular Acidosis with Arthritis and Dry Eyes-Dry Mouth


Ways Sjögren's syndrome may affect the body
Neurological problems, concentration/memory-loss (brain fog)
Dry nose, recurrent sinusitis, nose bleeds
Dry mouth, mouth sores, dental decay; difficulty with chewing, speech, taste and dentures
Dry skin, vasculitis, Raynaud's phenomenon
Stomach upset, gastroparesis, autoimmune pancreatitis
Peripheral neuropathy (numbness and tingling in the extremities)
Dry eyes, corneal ulcerations, and infections.
Difficulty swallowing, heartburn, reflux esophagitis
Recurrent bronchitis, pneumonia, interstitial lung disease
Arthritis, muscle pain
Abnormal liver function tests, chronic active autoimmune hepatitis, primary biliary cirrhosis
Vaginal dryness, painful intercourse


Symptoms of Renal Tubular Acidosis
Back pain
Lower abdominal pain
Painful urination
Cloudy urine
Increased Urinary urgency
Nausea/vomiting
Confusion
Muscle weakness
Muscle cramping in the back and abdomen
ePainAssist.com

Environmental factors, genetic factors & viruses can contribute to epithelial lesions & the release of autoantigens from the salivary glands.These events triggers release of type 1 & type 2 interferon (IFN) in genetically susceptible individuals, as well as T-cell activation leading to continuous B-cell activation and autoantibody synthesis. Immune complexes formed by autoantigens and autoantibodies (1), autoantibodies alone (2), & activated T cells, B cells, & plasma-cells (3) can be found in the circulation of the patient. Immune complexes are trapped within the glomeruli where they can form deposits & precipitates & trigger mesangial proliferation & extracellular matrix protein synthesis (corresponding to membranoproliferative glomerulonephritis secondary to cryoglobulinaemia). Autoantibodies to various specific renal transporters in the distal collecting ducts are trapped within the kidney where they might trigger electrolyte disturbances & distal renal tubular acidosis. In addition, activated T cells, B cells & plasma cells reach the kidney interstitium leading to the development of tubulointerstitial glomerulonephritis.These last 2events may be enhanced by local expression of autoantigens within the tubules.Local renal inflammation contributes to tubular lesions, including tubulitis, & triggers renal interstitial fibrosis & tubular atrophy contributing to Chronic Kidney Disease



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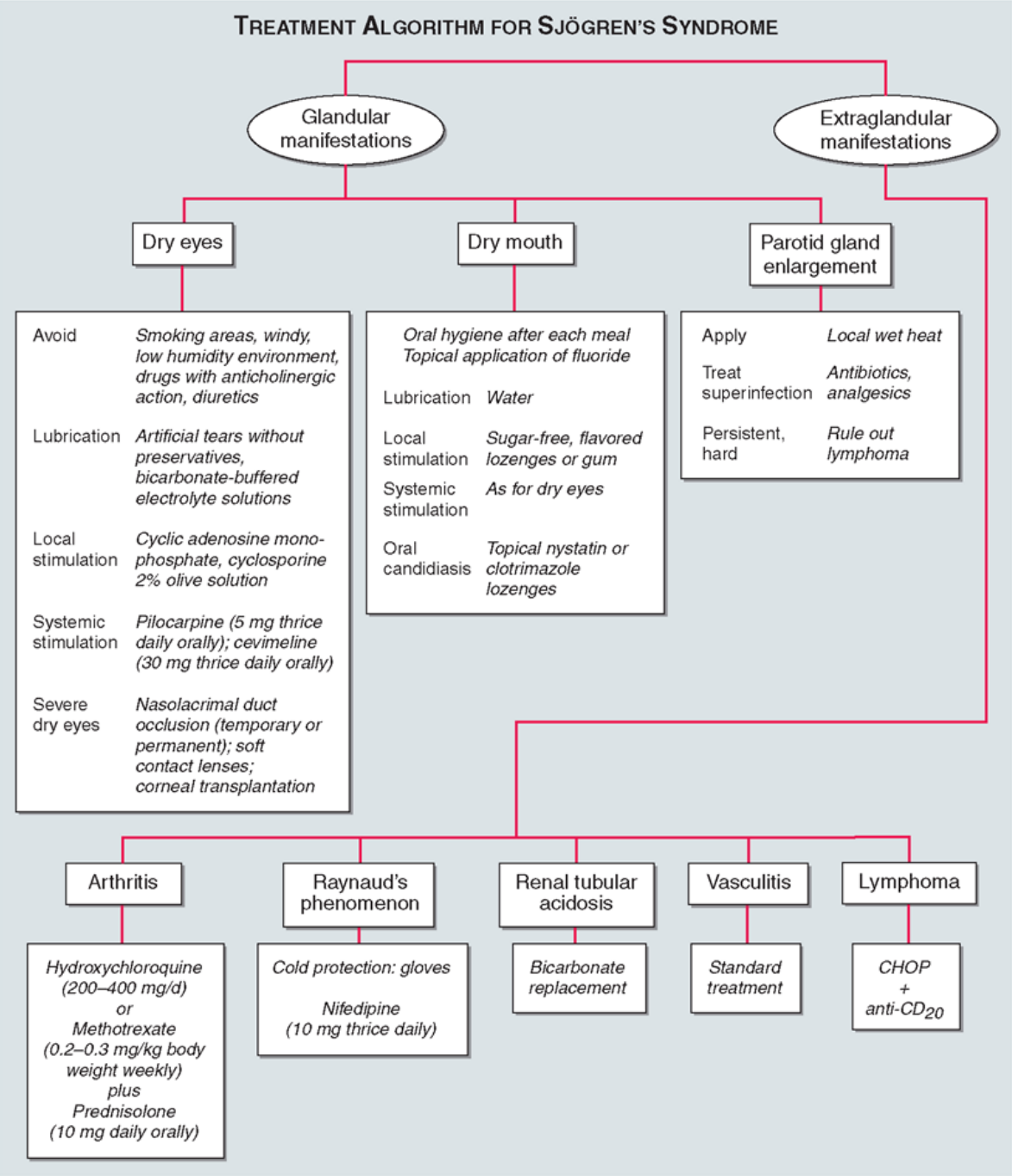

The Above Algorithm is Modern Management of Sjogren's Syndrome

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