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Dr. Pranay Penmetsa  
*JSS AHER*

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A Case of Sjogren’s Syndrome with Leucocytoclastic Vasculitis with Hypokalemic Periodic Paralysis

Dr. Pranay Penmetsa, Dr. Subramanian R, Associate Professor And HOD, Dept Of Rheumatology & Immunology

CLINICAL HISTORY:

37-year-old female presented with Complaints of dryness of eyes, mouth for 6 months.

Tingling sensation in both upper limbs for 3 days, insidious in onset, no postural variation, no aggravating and relieving factors

Complaints of upper and lower limb weakness, insidious onset, progressive, initially able to mix food and grip onto slippers now unable to lift hands above the level of head for 2 days, patient was able to get up from sitting position initially, now unable to do so.

Breathlessness since 1-day, insidious onset, progressive, initially on strenuous exercise progressed to at rest (MMRC 0 – MMRC 4), no postural variation, no diurnal variation. History of multiple joint pains for 4 days.

No h/o fever, No mouth ulcers.

No h/o change in voice, blurring of vision.

No h/o loss of smell, photophobia, double vision.

No h/o facial deviation, loss of sensation over face
No h/o difficulty in chewing, swallowing

No h/o auditory disturbances, vertigo, dizziness.

No h/o deviation of tongue, drooping of shoulder.

No h/o palpitations, sweating, loss of consciousness

No h/o increased thirst, increased appetite, increased frequency of passing urine.

No h/o vomiting, diarrhea, abdominal pain

She had past h/o Henoch Schoenlein Purpura in 2016 on alternate medication. 10 days post discharge, during follow-up for above complaints, she presented with a rash on the lower limbs, insidious onset, progressive, erythematous, itchy rash. No other new symptoms.

**EXAMINATION AND INVESTIGATIONS:**

A middle-aged female patient, moderately built and nourished, well oriented to time, place and person, alert, conscious and cooperative.

No Pallor, No Icterus, No Cyanosis, No Clubbing, No Lymphadenopathy, No Edema

**Vitals:**

Pulse rate: 88 bpm

BP: 100/60 mmHg

RR: 20 cpm
Temperature: Afebrile

**Systemic Examination:**

CVS: S1, S2 heard, no murmurs

RS: B/L NVBS ++, no added sounds

PA: Soft, non-tender, no organomegaly

CNS:

Higher Mental Functions: Conscious, oriented

Cranial Nerves: Intact

Motor System: Bulk Normal, Tone Normal, Power 0/5 in all limbs, Areflexia, Planter reflex was mute

Sensory System: Intact

Cerebellar Signs: Normal

Signs of Meningeal Irritation: Absent.

**Local Examination:** Multiple maculopapular rash present over both lower limbs, erythematous, red to violet color, non-blanching.

RBS: 136 mg/dl

LFT: Normal
Serology: Negative

Peripheral Smear: Normocytic, Normochromic with Neutrophilic Leukocytosis.

Platelet Count: 3.12 Lakh/ cu. mm.

Electrolytes:

Sodium: 144 mEq/L    Potassium: 2.9mEq/L

Chloride: 114 mEq/L

T3: 0.7 ng/ml

T4: 6.03 mcg/ml

TSH: 1.65 mcIU/ml

PT: 14.2 s

INR: 1.06

BT: 2 min

CT: 5 min

ABG:

pH: 7.115

pCO₂: 34.5
pO2: 236

HCO3: 10.6

Anion Gap: 5.3

Na: 139

K: 2.5

Cl: 123

ANA Profile:

nRNP/Sm: Not Detected

Sm: Not Detected

SS-A: Detected (3+)

Ro 52: Detected (3+)

SS-B: Detected (2+)

Scl-70: Not Detected

PM-Scl: Not Detected

Jo-1: Not Detected

CENP: Not Detected
PCNA: Not Detected

dsDNA: Not detected

Histones: Not Detected

Rib. P-Protein: Not Detected

AMA-M2: Not detected

Urea: 33 mg/dl

Creatinine: 0.9 mg/dl

Biopsy of Skin: shows features suggestive of Leucocytoclastic Vasculitis.

**FINAL DIAGNOSIS:**

1. Sjogren’s Syndrome

2. Hypokalemic Periodic Paralysis

3. Renal Tubular Acidosis

4. Leucocytoclastic Vasculitis

**TREATMENT:**
Quadriplegia reversed immediately after correction of Hypokalemia which includes infusion of KCl. 2 correction of Inj. KCl was given 1 day apart.

Syp. POTASSIUM CHLORIDE (POTKLOR) 10 ml 1-1-1

- Hypokalemia was caused due to Renal Tubular Acidosis which was treated with Sodium Bicarbonate and diuretics. Tab. SODIUM BICARBONATE (SODAMINT) 2-2-2

(ABG report included in investigations)

Tab. AMITRIPTYLINE (COTRIP) 0-0-1

Cap. VITAMIN B 12 (VICOBA) 1-0-0

Tab. HYDROXYCHLOROQUINE (HCQS) 400 mg 0-0-1

- LCV is treated with oral steroids along with Methotrexate which modulates the immune system and reduces the intensity of the disease.

- Sjögren’s Syndrome was treated symptomatically with eye drops, moisturizing lotion, saliva substitutes and immunomodulators like HCQS.

**DISCUSSION:**

Sjögren’s syndrome is divided into primary and secondary forms. The clinical hallmarks of Sjögren’s syndrome are keratoconjunctivitis sicca (dry eyes), xerostomia (dry mouth), and parotid gland swelling. Extraglandular features of primary Sjögren’s syndrome include fatigue, Raynaud’s phenomenon, polyarthralgia/arthritis, interstitial lung disease, neuropathy, and purpura. (1)
Hypokalaemia Periodic Paralysis (HypoKPP) is a disease that usually occurs at adolescence. Men are more often affected because of decreased penetrance in women. Attacks are often provoked by meals high in carbohydrates or sodium and may accompany rest following prolonged exercise.

Weakness usually affects proximal limb muscles more than distal. Ocular and bulbar muscles are less likely to be affected. Respiratory muscles are usually spared, but when they are involved, the condition may prove fatal. Weakness may take as long as 24 h to resolve. Life-threatening cardiac arrhythmias related to hypokalaemia may occur during attacks. A low serum potassium level during an attack, excluding secondary causes, establishes the diagnosis. (2)

Leucocytoclastic Vasculitis is a type of cutaneous vasculitis (Immune Complex mediated Vasculitis) which usually presents as a palpable purpura, but a variety of other skin lesions may be found including pustules, vesicles, urticaria, and small ulcerations. (1)

Most cases of idiopathic cutaneous leukocytoclastic vasculitis are mild and resolve with supportive measures such as leg elevation, rest, compression stockings, and antihistamines. In more chronic or resistant cases, a 4-6 week tapering dose of corticosteroids can be used. Rarely, immunosuppressive steroid-sparing agents such as methotrexate, azathioprine, mycophenolate mofetil, dapsone, cyclophosphamide, and intravenous immunoglobulin may be needed. (3)

ACKNOWLEDGEMENTS: None

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(1) Kelley Textbook of Rheumatology.
(2) Harrison’s Principles of Internal Medicine.

(3) Leukocytoclastic Vasculitis (Hypersensitivity Vasculitis)

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