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Original Research Article

Periodic Paralysis with Multiple Clinical Marker is it Sjogren's Syndrome

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Abstract

Sjogren's syndrome is a chronic, autoimmune disease. It can occur alone or in association with other autoimmune conditions like rheumatoid arthritis (RA) or lupus. Along with common glandular manifestations, Sjögren syndrome may also cause several extra glandular manifestations. We report a case of 38 year old female presenting to Neurology ICU having periodic paralysis with recurrent dental caries and intermittent waxing and waning purpuric non blanchable macular rash in bilateral lower limb. On evaluation, she was diagnosed as a case of Sjogren's syndrome with associated RA. Our case report illustrates the importance of recurrent dental caries and non-blanchable macular skin rash as clinical clue or biomarkers for etiological evaluation of cases with periodic paralysis or suspected Sjogren syndrome.

Keywords Sjogren's syndrome, Hypokalemic periodic Paralysis, Rheumatoid arthritis.

Introduction

Sjogren syndrome is one of the most common rheumatological diseases. The American College of Rheumatology(ACR)/European Alliance of Associations for Rheumatology (formerly European league against Rheumatism)/EULAR assist in the diagnosis of SS. [1]. SS is called secondary when it occurs in association with rheumatoid arthritis. [2] Patients having SS carries the higher risk for caries and higher dental expenses compared with healthy controls. [3,4] Cutaneous vasculitis has been reported in up to 30 percent of patients with SS. [5] The most common clinical manifestation of primary SS-associated cutaneous vasculitis is palpable and nonpalpable purpura occurring on the lower extremities. [6] Here we report a case of SS diagnosed with several years delay due to under recognition of dental caries and cutaneous manifestation as the first presentation of the disease.

Case Presentation

A female "in her 30s" presented to us with sub-acute onset, symmetrical weakness of all the four limbs involving proximal and distal muscles for four days. Patient had progressive dental erosions and cutaneous manifestation in the form of waxing and waning purpuric non blanchable macular rash in bilateral lower limb for the last 7 year. She had similar episode of weakness with severe hypokalemia and hyponatremia one year back treated with oral potassium chloride syrup. Patient suffered a cardiac arrest and revived in the same episode. She was discharged as a case of hypokalemic periodic paralysis but detailed etiological evaluation was not done. She had history of galactorrhea for one year. At the time of admission her BP was 110/60 mm of Hg, Pulse 106/min and respiratory rate was 22/min. Her general physical examination revealed severe dental carries with non palpable, non-blanchable purpuric macular rash in bilateral lower limb. Neurological examination revealed quadriparesis and areflexia. Remainder of the neurological examination was unremarkable. She was admitted in intensive care unit with power grade 1/5 (all the four limbs) with areflexia. Immediately, arterial blood gas monitoring was done that shows severe hypokalemia (serum potassium-1.5 meq/l) and hyperchloremic non anion gap metabolic acidosis while ECG shows tall T wave with prolonged QU interval. (Table1). She was managed aggressively with potassium

chloride syrup (maximum dose of 0.4 mg/kg body weight) through Ryle's tube every 30 minutes that was reduced to 15 meq every four hourly after power improved to MRC grade 4/5 and potassium reaches to 3.5 meq/l after 24 hours.

Table 1: Showing various laboratory parameters

Parameters	First value	Second value	Normal Range
Arterial blood gas			
PH	7.0	7.1	7.35-7.45
K+	1.5	4.0	3.5-4.5 meq/l
HCO3-	10.5	11.0	22-26 meq/l
PCO2	37.5	24.9	38-45 mm Hg
Anion gap	10.4	9.2	
Chloride	120	125	98-110 mmol/l
Serum			
Hemoglobin	9.5		13-17 gm/dl
Total Leucocyte Count	19700		4000-10000 cells/mm3
Platelet Count cells/mm3	154000		50000- 400000
Sodium	131.5	155.2	137-150 meq/l
Potassium	2.48	5.19	3.5-5.3 meq/l
Urea	31		13-43 mg/dl
Creatinine	1.28		0.8-1.3 mg/dl
TSH	1.10		0.35-4.94 uIU/ml
ALT	31		0-45 U/L
AST	41		0-40 U/L
FSH	1.87		0.95-11.95 mIU/l
LH	1.17		1.14-8.75 mIU/ml
Prolactin	43.37		3.46-19.4 ng/ml
CRP	3.8		0-6 mg/L
RA Factor	111.2		Up to 20
Viral Marker			
HCV		Negative	
HIV		Negative	
HbsAg		Negative	
ANA ENA Combo Panel			
Antinuclear antibody Hep-2		Positive	
Pattern		Nuclear Speckled	
Intensity		3+	Significantly positive
Primary titre/dilution		1:100	
Endpoint titre/dilution		1:1000	
Anti -ds DNA antibody		33.64	0-200 IU/ml
SS-A /Ro		4+	Strong Positive
SS-B / La		4+	Strong Positive
Vasculitis Panel			
p-ANCA; Anti Myeloperoxidase 3.35 <20 U/ml			
c-ANCA; Serin Proteinase 3		5.29	<20 U/ml
Urine analysis			
Calcium, 24 hours urine		302.50	100-300 mg/day`
Urine culture and sensitivity		sterile	
Schirmer test		<5 mm in bilateral eye	<10 mm-dry eye

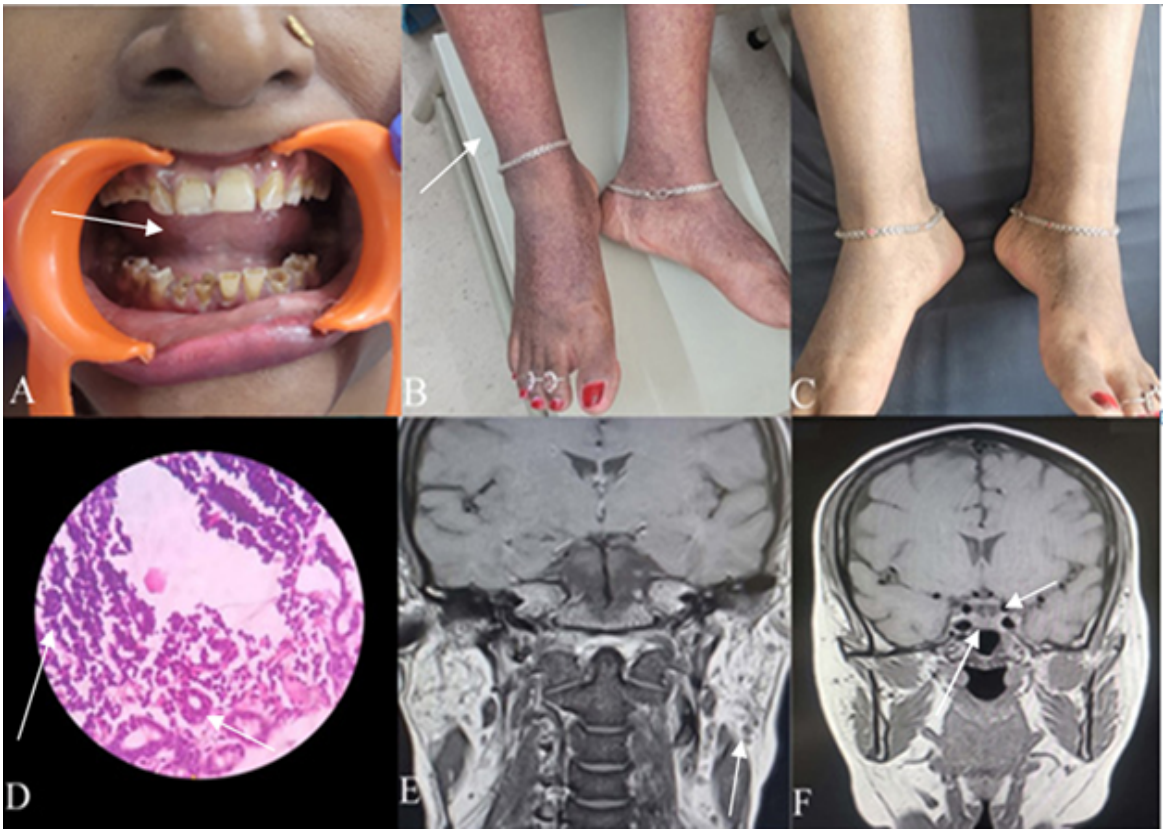

Figure 1

(A). Oral examination showing generalised attrition, multiple root stumps with multicarious teeth. **(B)** Purpuric, non-blanchable macular rash in bilateral lower limb. **(C)** Rash disappears on treatment **(D**) Lip biopsy: hematoxylin and eosin (H&E) stain showing serous acini (small arrow) and lymphoid aggregates (large arrow) with low power view 10x. **(E)** Bilateral Parotid gland coronal T1sequence MRI shows partly indistinct margin with diffusely altered signal intensity suggestive of fatty infiltration.**(F)** Larger arrow showing bulky and asymmetrically enlarged pituitary with smaller arrow showing slight deviation of pituitary stalk towards left on T1 coronal sequence.

Differential

Patient was managed as a case of recurrent hypokalemic periodic paralysis and etiological workup for the same was done including hyperthyroidism, AIDS, Amyloidosis, HCV, Sarcoidosis.

Treatment

Our patient was treated was treated acutely with oral potassium chloride syrup and discharged on oral glucocorticoids, bicarbonate and potassium repletion oral tablet. She has been advised to avoid diuretics, antidepressants, anticholinergics. Apart from this, she is advised for sugar free candies, frequent gargling during day time, using fluoride containing toothpaste, frequent mouth wash with chlorhexidine.

Follow up

During the course of follow up with improvement in her weakness and cutaneous manifestation she was started on Azathioprine 50 mg once daily dose that was escalated to 100 mg dose. Our plan is to taper steroids after 6 months.

Discussion

SS may present with wide spectrum of extra-glandular manifestations including gastrointestinal, pulmonary, neurologic, cutaneous, oral manifestations along with associated endocrine manifestations in addition to sicca symptoms. The extra-glandular manifestations such as recurrent dental caries and cutaneous features are underestimated and poorly looked symptoms of the disease. Our patient presented with PP having history of recurrent dental caries as the first clinical manifestation which was treated by dentist with little success. There are very rare case report that reported dental caries as the first manifestation of SS. Ahmadi E et al [7] reported a case of primary SS with dental caries as the first manifestation of the disease. In his patient, there was associated difficulty in swallowing dry food with foreign body sensation in bilateral eye. Our patient had no history suggestive of any primary difficulty in swallowing dry food nor there was any foreign body sensation in her eyes. There are case reports that have reported salivary gland hypofunction in SS with increased prevalence of dental caries but not as the first

presentation of the disease. In our patient, cutaneous manifestation was the second presentation in the form of waxing and waning purpuric non blanchable macular rash in bilateral lower limb. There are very few case report that presented solely with cutaneous feature as first presentation of the disease. Sneha Centala et al [8] reported case of 56yearold Asian female presenting with preauricular erythematous lesion as the rare first manifestation of the disease. Our case is a very rare one as these manifestations are reported in literature but not as the initial presentation of the disease. In our case, there was whitish discharge from breast with associated hyperprolactinemia. These results are in concordance Gutierrez [9], Haga and Rygh [10] that showed significant increase in mean serum prolactin levels in primary SS. Our patient had bulky pituitary with asymmetrical enlargement of the gland which might be an incidental finding.

Our case of SS with associated RA with extra-gladular manifestation such as recurrent dental caries and cutaneous manifestation as the first manifestation of the disease with associated PP has not been reported in literature to the best of our knowledge.

Conclusion

Dental and dermatological manifestations are underestimated for SS diagnosis in general.

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