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Internal Medicine

A RARE PRESENTATION OF SJOGREN'S SYNDROME: SJOGREN'S SYNDROME WITH DYSAUTONOMIA – A CASE REPORT

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ABSTRACT Sjogren's syndrome is a chronic systemic autoimmune disease characterised by inflammation and dysfunction of the exocrine gland (6). Patients may also experience neurological symptoms characterized by paresthesia , pain, dizziness, palpitations and presyncope. In fact, patient may present initially with pure neurological symptoms (6). Thus the diagnosis of sjogren's syndrome remains a clinical challenge. We report a case of a patient with neurological symptoms diagnosed as Sjogren's syndrome with dysautonomia. A 30 year old male patient presented with generalised fatigue, palpitations and dizziness. On examination, postural orthostatic tachycardia was noted, started on beta blockers. Patient had improvement in symptoms. There was also progression of his sjogren's disease, had PSVT – radiofrequency ablation was done and disease progression was managed with rituximab and IVIG therapy. The diagnosis of neuropathy in sjogren's syndrome requires a combination of clinical signs and symptoms (5). This case report mainly focuses on autonomic dysfunction associated with sjogren's syndrome. We aim at sharing awareness regarding autonomic symptoms in patients with sjogren's syndrome and alert physicians that it could be the initial presentation of the systemic disease.

KEYWORDS : Sjogren's syndrome , Histopathology of minor salivary gland, Dysautonomia.

INTRODUCTION:

Primary sjogren's syndrome is an autoimmune disease affecting exocrine glands, giving rise to hypofunction, especially of lacrimal and salivary glands. (1). The classical feature of primary sjogren's syndrome include oral and ocular dryness , pain and fatigue (3). Fatigue is often the predominant symptom experienced by patients with sjogren's syndrome (3).

According to estimation, approximately 70% of patients with sjogren's syndrome complain of significant fatigue with many making it as the most disabling symptom of the disease(2). Many studies have reported autonomic nervous system dysfunction in primary sjogren's syndrome and that ANS dysfunction and fatigue are strongly associated (3). Immunemediated destruction of Salivary and lacrimal gland , has been largely attributed to etiology of primary sjogren's syndrome. However, the degree of destruction of exocrine gland and exocrine function are poorly correlated (3). According to AECC, for diagnosis of Primary sjogren's syndrome , some evidence of autoimmunity (ie) anti-SSA / anti-SSB seropositivity and / or Positive lip biopsy is mandatory (1).

Here, we report a case of Primary sjogren's syndrome presenting with dysautonomia.

CASE REPORT:

A 30 year old male with marfanoid habitus (181cm / 61.2kg) came with outpatient department with chief complaints of generalized weakness and fatigue for past 2 years. History of palpitations and orthostatic light headed ness for past 2 years.

On examination, His supine BP and PR- 124/84mmhg and 84/min respectively. Standing BP and PR- 124/90mmhg an d 116/min. He was evaluated further with tilt table test, it showed increase in heart rate in upright position. In Nerve conduction study , both sural SNAP showed reduced amplitude (axonal neuropathy). Sympathetic skin response in Autonomic function test could not be elicited from lower limb , which suggests small fibre involvement. Nerve biopsy showed chronic axonopathy, he was diagnosed with Postural orthostatic tachycardic syndrome and dysautonomia, started on Beta- blockers. Patient had persistent fatigue , on further evaluation, he was found to have ANA 2+ (speckled).

Young male with persistent fatigue, dysautonomia and ANA 2+, lip biopsy was done. Lip biopsy showed chronic lymphocytic sialadenitis with focus score > 2.5. Hence, he was diagnosed with sjogren's syndrome and started on MMF 500mg BD, HCQS 200mg BD and Deflazacort 6mg OD. Meanwhile, patient had tachycardia upto 200 beats per minute, ECG showed PSVT , reverted back with adenosine. Again patient had 2to 3 similar episodes , hence EPS with RFA has been done and continued with maintanence beta blockers. Patient had persistent fatigue, hence rituximab and IVIG infusions.

INVESTIGATIONS:

INVESTIGATION	VALUE	NORMAL RANGE
HEMOGLOBIN	14.8 g/dl	13-18 g/dl
WBC	8000/cubic mm	4000-11000/ cubic mm
PLATELET COUNT	2,19,000/cubic mm	1,50,000-4,50,000/cubic mm
UREA	24 mg/dl	13-43 mg/dl
CREATININE	0.9 mg/dl	0.7-1.3 mg/dl
TOTAL BILIRUBIN	0.9 mg/dl	0.2-1.2 mg/dl
ALP	61 U/l	< 128 U/l
AST	25 U/l	< 35 U/l
ALT	13 U/l	< 45 U/l
BLOOD SUGAR	105 mg/dl	< 140 mg/dl
SODIUM	142 mEq/L	136-145 mEq/L
POTASSIUM	3.8 mEq/L	3.5-5.1 mEq/L
COMPLEMENT 3	85.6 mg/dl	84-164 mg/dl
COMPLEMENT 4	10.2 mg/dl	15-48 mg/dl
ANA	2+ (speckled)	
dsDNA	NEGATIVE	
Anti-MPO	NEGATIVE	
Anti-PR3	NEGATIVE	
ECHO	NO RWMA; EF = 63%	
HIV ELISA	NEGATIVE	
HBsAg	NEGATIVE	
TSH	1.5 micro IU/ml	0.42-4.2 micro IU/ml
Anti-SS-A / Anti-SS-B	NEGATIVE	

NERVE CONDUCTION STUDY	Lower limb motor conduction study showed reduced CMAP amplitude from right tibial. Sensory conduction showed reduced SNAPs amplitude from bilateral sural.
AUTONOMIC STUDY	Absent lower limb Sympathetic skin response suggests small fibre involvement.
HISTOPATHOLOGY OF LIP BIOPSY	LCA highlights the periacinar, perivascular and periductal lymphocytic infiltrate. Focus score = 2.5 (>1). Chronic sialadenitis. Features considered to be suggestive of sjogren's syndrome.

DISCUSSION:

Sjogren's syndrome is a systemic autoimmune disease that affects exocrine gland, characterised by dry eyes , dryness of mouth (5). It can

also affect other organs including nervous system , which may contribute to wide range of symptoms and functional burden (3). The clinical symptoms of sjogren's syndrome can be divided into two broad categories as exocrine glandular feature and extra glandular feature (5).

Neurological symptoms are the common characteristics of extra glandular manifestations and it occur in approximately 20% of patients with sjogren's syndrome (5). Onset of neurological symptoms can precede diagnosis of primary sjogren's syndrome in 40-80% cases (6). Among that, peripheral neuropathy is the major neurological manifestations.

One of the most common peripheral neuropathy in primary sjogren's is axonal sensory and sensorimotor polyneuropathy (5). Multiple neuropsychologically domains are involved in primary sjogren's syndrome , Anxiety and depression are more prevalent in patients with primary sjogren's syndrome (6).

Autonomic neuropathy is also a subset of peripheral neurological manifestations in patients with primary sjogren's syndrome (5). It is characterized by abnormal cardioreflex testing such as orthostatic hypotension, Postural orthostatic tachycardia syndrome , abdominal pain, diarrhoea and abnormality in micturition (5).

Focal lymphocytic sialadenitis is the characteristics lesion of sjogren's syndrome (7). Minor salivary gland biopsy is the valuable tool in diagnosis of sjogren's syndrome with focus score more than or equal to 1 (4).

Patient with neuropathy symptoms should proceed with autonomic study and nerve conduction study (5). The management of autonomic dysfunction is complex and no definitive cure (5). Symptomatic management can be made with beta blockers and steroids. In case of severe forms, could be managed with either IVIG or Rituximab infusion as done in our patient.

Our patient reported fatigue, orthostatic light headedness and palpitations, on examination, he had Postural orthostatic tachycardic syndrome, nerve conduction study showed axonal neuropathy, sympathetic skin response absent in lower limb, nerve biopsy showed chronic axonopathy, ANA 2+ and lip biopsy showed chronic lymphocytic sialadenitis. Thus our patient was diagnosed with sjogren's syndrome with autonomic dysfunction. He was started on beta blockers, immunosuppresants, steroids and received Rituximab, IVIG infusions. During followup, his symptoms were less.

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