Clinical Profile of Primary Sjogren’s Syndrome with Hypokalemic Periodic Paralysis

Nachiket Kulkarni1*, Arvind Chopra2

Abstract

Introduction: Primary Sjogren’s Syndrome (pSS) with Hypokalemic Periodic Paralysis (HPP) whether an association or a different clinical subset needs review.

Methods: Cross-sectional retrospective study of subjects of Primary Sjogren’s Syndrome with Hypokalemic Periodic Paralysis (HPP) identified from database maintained at Centre For Rheumatic Diseases, Pune since 1996 with records of over 50000 patients. The diagnosis was clinical. Clinical and investigations data was extracted pertaining to initial examination and follow up. Standard investigations & ELISA, immunoblot and nephelometry to assay autoantibodies (AAb) were done

Results: 16 patients of Primary Sjogren’s Syndrome (pSS) with Hypokalemic Periodic Paralysis (HPP) were identified in the period 2000-2014. Presenting feature was HPP in 86% with Dry eye (4%) and Arthralgias (10%) in remaining. Distal Renal Tubular Acidosis was identified in all. All were females with average age of 26 years. Symptomatic ocular sicca noted in 60% & Oral sicca in 50% patients. Other features – Arthralgias (91%), arthritis (42%), mucositis (38%), Neuropathy (30%), skin rash (20%) cytopenias (19%), Erosive arthritis (10%), interstitial lung disease (10%) and Raynaud Phenomenon (10%). 100% were positive for ANA. SSA was positive in 100%, SSB in 50% of patients & Rheumatoid Factor in 70 %. Hypothyroidism was associated in 70% patients.

Conclusion: We present a large series of Primary Sjogren’s Syndrome with Hypokalemic Periodic Paralysis (HPP) from India. Prominent features of female dominance, younger age of onset and SSA positivity noted in this cohort of patients on Routine clinical and serology phenotype suggests existence of a distinct subset. HPP was presenting feature in majority

Introduction

Sjogren’s Syndrome (SS) is reported as most common connective tissue disorder.1 SS can be primary or secondary associated with various connective tissue disorders (eg Rheumatoid Arthritis, Systemic lupus erythematosus, etc).2 Primary SS comprise about half of the cohort.1 SS is characterized by autoimmune exocrinopathy of salivary and lacrimal glands.1 Patients present with symptoms of dryness of eyes and mouth. SS is associated with multiple extraglandular features.2 Association of pSS and Hypokalemic Periodic Paralysis (HPP) first described in 19813 has been infrequently reported. First description from India dates back to 1996. Whether pSS and HPP occurrence is an association or different clinical subset needs review. Current study describes clinical phenotype of this co-expression.

Material and Methods

Aim: To study clinical phenotype of co-occurrence of Primary Sjogrens Syndrome and Hypokalemic Periodic Paralysis.

Design: The study was a retrospective cross sectional analysis design using a rheumatology database.

Site: Single center study. Data of patients attending a popular community based rheumatology clinic [Centre For Rheumatic Diseases (CRD) based in Pune metropolis in West India (www. rheumatologyindia.org) was assessed. A comprehensive patient data base is maintained in CRD since 1998.

Selection: The principle criteria for inclusion in the study was a clinical diagnosis of Primary SS made by a senior rheumatologist (AC). Occular sicca was confirmed by Schirmer’s test and tear film breakage time by Ophthalmologist. Oral sicca was a clinical assessment on patient symptom confirmation. Salivary functional tests could not be conducted All patients were required to have been followed for at least one year after the initial diagnosis. The current study cohort was identified from the database during period of 2002 to 2014

Examination, Follow up: All patients were examined by senior rheumatologists (AC). A standard rheumatology case record form recorded detail history and clinical evaluation. A comprehensive laboratory and plain Skiagram evaluation results were available. Blood was tested for Rheumatoid Factor (RF, nephelometry with cut off at 40 IU/ml) and Anti Cyclic Citrullinated Polypeptide (ACCP, second generation, ELISA with cut off at 5 RU/ml ) and anti-nuclear antibody (ANA, ELISA) and ANA profile analysis (ANA Blot, ELISA) were conducted along with all routine hemogram, biochemistry and urine analysis. Hypokalemic periodic paralysis (HPP) was identified by Neurologist. Distal renal tubular acidosis was identified in each patient.

All other rheumatic diagnosis were clinical though standard classification criteria were referred to.

Data and Statistics: Data was extracted from the database and entered into an Excel (MS) worksheet

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Received: 06.06.2016; Accepted: 10.10.2017
stopped potassium supplementation. Paralysis was noted in 3 patients who had Raynaud’s Phenomenon. Relapse of attacks was noted in 2.6 meq/dl during the attacks. The age of onset of Primary Sjögren’s Syndrome has high female dominance as reported in western and Indian literature. This cohort though is characterised by only female presence. Case reports of Primary Sjögren’s Syndrome presenting feature in majority. This cohort displayed multiple extra-glandular features. All patients had Anti SSB antibody.

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Renal involvement in Primary Sjögren’s Syndrome has been well described. Interstitial nephritis and Glomerulonephritis are two main forms. Interstitial Nephritis which can be seen in about one third patients can produce latent or complete renal failure. Hypokalemic Periodic Paralysis is associated with distal renal tubular acidosis and rarely proximal. Hypokalemic Periodic Paralysis is associated with distal renal tubular acidosis. Hypokalemic Periodic Paralysis was positive for Anti SSB. 1 patient was positive for SSA, SSB, RF, ACCP, Anti dsDNA, Histone. Absence of Sm, n-RNP, SCL-70, CENP, PM-SCI, JO-1, PCNA, Nucleosome, ribosomal P-protein was noted in ANA profile in this cohort.

Discussion

We present a case series of patients with Primary SS and HPP. This retrospective analysis highlighted some unique features of this cohort. This association was noted only in females. Young age of onset was common. HPP was presenting feature in majority. This cohort displayed multiple extra-glandular features. All patients had Anti SSA antibody.

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Acknowledgement

The authors acknowledge the efforts and support of all doctors, paramedical and support staff of Centre for Rheumatic Diseases, Pune. We would like to thank the laboratory staff for their full-hearted support. Lastly we would like to thank the patients.

References


Table 1: Extra-glandular features

<table>
<thead>
<tr>
<th>Extra glandular features</th>
<th>Frequency (%)</th>
</tr>
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<tbody>
<tr>
<td>Arthralgias</td>
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<td>Arthritis</td>
<td>20</td>
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<tr>
<td>Mucositis</td>
<td>40</td>
</tr>
<tr>
<td>Neuropathy*</td>
<td>30</td>
</tr>
<tr>
<td>Skin Rash**</td>
<td>20</td>
</tr>
<tr>
<td>Cytopenias</td>
<td>20</td>
</tr>
<tr>
<td>Intersitial Lung Disease</td>
<td>10</td>
</tr>
<tr>
<td>Raynaud’s Phenomenon</td>
<td>10</td>
</tr>
</tbody>
</table>

*Identified on Nerve Conduction Velocity, **Rash characteristic of Connective Tissue Disorders

for demographics and other variables Frequency percentage of these parameters were calculated.