Primary Sjögren's syndrome with polymyositis, a rare amalgamation

Harpreet Singh, Deepak Jain, Kiran B, Neeraj Kumar

Department of Medicine and Division of Rheumatology, Pandit Bhagwat Dayal Sharma University of Health Sciences, Rohtak, Haryana, India

Correspondence to Deepak Jain, MD, Department of Medicine, Pandit Bhagwat Dayal Sharma University of Health Sciences, Rohtak 124001, Haryana, India. Tel: +91-9416147887; fax: +911262211149; e-mail: jaindeepakdr@gmail.com

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Sjögren's syndrome is characterized by diminished lacrimal and salivary gland secretory function. This disorder is not strictly confined to the exocrine glands and its manifestations may extend to extraglandular sites, such as the lungs, kidneys, reticuloendothelial system, and the musculoskeletal system. Although muscular manifestations are very common with Sjögren's syndrome, true myopathy is very rare. Here, we report a case of a 45-year-old woman who presented with complaints of bilateral progressive weakness of upper and lower limbs associated with difficulty in neck holding with a history of dryness of the mouth and the eyes. The diagnosis of polymyositis associated with Sjögren's syndrome was established on the basis of clinical picture and diagnostic tests. True polymyositis is very rare in primary Sjögren syndrome and there are scarcely any cases of primary Sjögren's syndrome with polymyositis reported in the literature.

Keywords:
myopathy, polymyositis, Sjögren's, syndrome

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Introduction

Sjögren's syndrome is a slowly progressive, inflammatory autoimmune disease affecting primarily the exocrine glands [1]. It has a broad clinical spectrum extending from autoimmune exocrinopathy to extraglandular (systemic) disease affecting the various organs of the body. Musculoskeletal involvement in primary Sjögren's syndrome (PSS) has a diverse clinicopathological spectrum. Myalgias and muscular weakness have been noted in 33% of the patients with PSS [2]. Although there is a frequent association of subclinical myositis as evident by histopathological examination, true myositis is rare; however, polymyositis is even more very rarely associated with PSS. The literature on muscle biopsies in PSS has not yet provided a definite conclusion on the type of infiltrate and the degree of muscle damage.

Case report

A 45-year-old woman with no known chronic illness presented with weakness of the bilateral upper and lower limbs for the past 3 months that was insidious in onset, gradually progressive, and associated with difficulty in combing hair, difficulty in climbing stairs, and difficulty in standing from a squatting position. She also reported a history of dryness of the eyes and the mouth for the past 6 months. On examination, the patient was conscious and oriented, with a blood pressure 110/70 mmHg, pulse rate (PR) 88/min. There was no pallor, icterus, clubbing, cyanosis, pedal edema, or lymphadenopathy, and jugular venous pressure was normal. Neurological examination showed normal muscle bulk and hypotonia of both the upper and the lower limbs. Deep tendon reflexes were reduced in all four limbs. Power was decreased in the bilateral upper and lower limbs proximally (2/5), distal muscle power was normal, and planters were bilateral (B/L) flexor. The rest of the examination including other systems was normal. Patient consent form was obtained before the case report study after fully disclosing all points in local and English language.

Routine laboratory investigations including complete hemogram, blood sugar, and renal and liver function tests were normal and the rest of the investigations are presented in Table 1. Schirmer's Test was positive (2 mm B/Leye), with an abnormal ocular staining score of more than 5 (or van Bijsterveld score of ≥4). Needle electromyography showed myopathic potential with low-amplitude, polyphasic waves along with fibrillations. Muscle biopsy showed atrophic, degenerating, and regenerating myofibers in a patchy distribution. There was endomysial mononuclear inflammatory infiltration of lymphocytes and plasma cell features suggestive of polymyositis (Fig. 1). Salivary gland biopsy was also planned, but the patient refused this procedure. Sialography studies were not available in our institution. However, as our patient fulfilled the American College of Rheumatology/

European League against Rheumatism criteria for the diagnosis of PSS, these investigations were deferred.

The diagnostic criteria for PSS were fulfilled in our patient (New 2016 American College of Rheumatology/European League against Rheumatism classification criteria for PSS) (provided in Table 2) [3] and polymyositis was confirmed by the muscle biopsy. She was treated with intravenous methylprednisolone 1 g for 1 days, followed by oral prednisolone of 1 mg/kg tablet, pilocarpine, lubricating eye drops, and other supportive measures. The disease activity was monitored using manual muscle testing (MMT). The above treatment led to an improvement in the clinical symptoms and quality of life of this patient. Steroids were tapered gradually and the patient is under our regular follow-up. Now the patient is doing well with oral prednisolone of 5 mg/day, lubricant eye drops, pilocarpine tablet 5 mg thrice daily, and calcium supplements.

Table 1 Specific investigations of patient

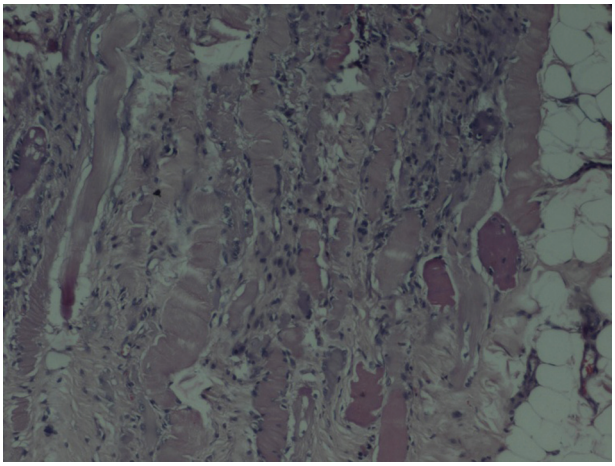
Investigations	Results
Erythrocyte sedimentation rate	66 mm in first hour (0–20 mm/h)
CRP	6 mg/l (positive)
Indirect immunofluorescence assay	1 : 3200 (speckled pattern)
Total CPK	9509 IU/l (normal: 38–176 U/l)
Anti-Ro/SSA	120.18 IU/ml (normal: <3 IU/ml)
Anti-La/SSB	62.64 IU/ml (normal: <3 IU/ml)
Anti-U1-RNP antibody	<20 U (negative)
Anti-Jo antibody	<20 (negative)
Anti-dsDNA	18.2 IU/ml (negative)
Complement component 3 (C3)	97.40 (normal: 90–180 mg/dl)
Complement component 4 (C4)	18.50 (normal: 10–40 mg/dl)
Serum ACE	44 U/l (normal: 8–65 U/l)
HIV, HBsAg, anti-HCV antibody	Negative
Triidothyronine (T3)	1.1 ng/dl (normal: 0.60–1.81)
Thyroxine (T4)	8.4 μg/dl (normal: 5.01–12.45)
Thyroid-stimulating antibody	2.6 μIU/ml (normal: 0.5–5)
Cryoglobulins	Negative

ACE, angiotensin-converting enzyme; CPK, creatine phosphokinase; CRP, C-reactive protein; HBsAg, hepatitis B virus surface antigen; SSA, Sjogrens syndrome related antigen A; SSB, Sjogrens syndrome related antigen B.

Discussion

Sjögren's syndrome is characterized by diminished lacrimal and salivary gland secretory function. This disorder, not strictly confined to the exocrine glands, may extend to extraglandular sites, such as the lungs, kidneys, reticuloendothelial system, and the musculoskeletal system. The prevalence of PSS ranges from 0.1 to 4.6%. Nearly three quarters of the patients with PSS manifest signs and symptoms of extraglandular disease. Extraglandular involvement is more likely to

Figure 1



Muscle biopsy showing features of polymyositis.

Table 2 American College of Rheumatology/European League against Rheumatism classification criteria: primary Sjögren's syndrome

Items	Scores
Labial salivary gland with focal lymphocytic sialadenitis and a focus score of ≥1 foci/4 mm^2	3
Anti-Ro/SSA	3
Ocular staining score ≥5 (or van Bijsterveld score ≥4) in at least 1 eye	1
Schirmer's test ≤5 mm/5 min in at least 1 eye	1
Unstimulated whole saliva flow rate ≤0.1 ml/min	1

The ACR/EULAR classification of primary Sjögren's syndrome applies to any individual who fulfills the inclusion criteria, does not have any conditions listed in the exclusion criteria, and has a score of more than or equal to 4 when the weights from the five following criteria items are summed. Inclusion criteria are applicable to any patient with at least one symptom of ocular or oral dryness, defined as a positive response to at least one of the following: (a) Have you had daily, persistent, troublesome dry eyes for more than 3 months? (b) Do you have a recurrent sensation of sand or gravel in the eyes? (c) Do you use tear substitutes more than three times a day? (d) Have you had a daily feeling of dry mouth for more than 3 months? (e) Do you frequently drink liquids to aid in swallowing dry food? or in whom there is a suspicion of Sjögren syndrome. Exclusion criteria include a previous diagnosis of any of the following, which would exclude the diagnosis of Sjögren syndrome and participation in Sjögren syndrome studies or therapeutic trials because of overlapping clinical features or interference with criteria tests: (a) history of head and neck radiation treatment, (b) active hepatitis C infection (with confirmation by PCR), (c) AIDS, (d) sarcoidosis, (e) amyloidosis, (f) graft-versus-host disease, and (g) IgG4-related disease. ACR/EULAR, American College of Rheumatology/European League against Rheumatism; IgG, immunoglobulin G; SSA, Sjogrens syndrome related antigen A.

occur in the patient with positive serum anti-Ro and anti-La antibody. In our patient, the diagnostic criteria for PSS were fulfilled and she also fulfilled the requirements for a diagnosis of polymyositis [3,4]. Cryoglobulins and serum complements were normal, which are predictors of future malignancy [4], and there were no signs of infectious, neoplastic, or endocrinological disease that could have explained the various clinical features presented. Before disease onset, no drugs had been ingested by the patient.

Involvement of the muscular system in Sjögren's syndrome appears to be rather frequently encountered. The majority of patients with Sjögren's syndrome have muscular and articular pain [5–7]. Mayalgias are found in 33% of patients with PSS; however, myositis was only found in 3% [8]. The majority of reported cases with myositis are subclinical with histological evidence of focal myositis and there are some reported cases of inclusion body myositis with PSS. However, true polymyositis is very rare. There are scarcely any cases of PSS with polymyositis reported in the literature.

There is paucity of knowledge on the prognosis of this combination of diseases, but our patient responded favorably to oral corticosteroid therapy. Regular follow-up is required in such patients as both diseases are associated with the risk of hidden malignancy and long-term follow-up will enable us to study the final prognosis of such a patient.

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Conflicts of interest
There are no conflicts of interest.

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