A rare case of bilateral trigeminal palsy presenting as a complication of Sjogren syndrome

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Sjogren syndrome is a chronic disorder affecting the exocrine glands of the body, mostly lacrimal and salivary glands, resulting in sicca symptoms. Diagnosis is aided by the presence of anti-Ro and anti-La antibodies and gland biopsy showing lymphocytic infiltration. Bilateral loss of facial sensation in Sjogren syndrome (SS) has not been reported before, although unilateral trigeminal palsy as a presentation of SS has been reported in literature. A 25-year-old lady presented in the neurology clinic with complete facial numbness and vision loss for 1 month. The vision worsened, leading to complete blindness. She had dry mouth and dry eyes for the past 3 months. She was diagnosed with SS using American–European consensus Sjogren criteria. Electrophysiological blink reflex testing confirmed complete trigeminal nerve palsy. SS was treated with prednisolone, 1 mg/kg/day, with a poor response. Symptomatic treatment was given for sicca symptoms, which improved significantly. Corneal transplantation was done for visual loss, but limited improvement was seen.

Keywords:
blink reflex, complete blindness, corneal transplant, Sjogren syndrome, trigeminal nerve palsy

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Introduction

Sjogren syndrome (SS), first described by M.D. Henry Sjogren, is a rare endocrine disorder characterized by chronic inflammatory disorder characterized by diminished lacrimal and salivary gland function [1–3]. SS can be classified as primary disease or secondary to other connective tissue disorders. The disease mostly affects middle-aged individuals and women (female : male ratio is 9 : 1) [4].

At least one-third of patients with SS present with systemic manifestations including neurological, renal, pulmonary, articular, and gastrointestinal [5]. Neuro-Sjogren is a rare disease that manifests with myelitis, hypophysitis, devics disease, autonomic, and peripheral neuropathy. We are presenting a case of complete facial sensory loss, which is secondary to bilateral trigeminal neuropathy that was later on diagnosed as having SS. This patient was having complete blindness due to neurotrophic corneal ulceration.

This case is rare; to the best of our knowledge, there has been no case of bilateral trigeminal palsy presenting as a complication of SS reported so far in PubMed. However, unilateral trigeminal palsy as a presentation of SS has been reported in literature. Hull *et al.* [1] in their article 'Sjögren's syndrome presenting as a severe sensory neuropathy including involvement of the trigeminal nerve' described the involvement of trigeminal nerve in SS; 'Sjogren's syndrome-associated neuropathy' was published by Koike *et al.* [2], and there are a number of articles in literature regarding trigeminal neuropathy and SS association, although, to best of our knowledge, in all these articles involvement of trigeminal nerve was unilateral.

Case history

A 25-year-old woman, with no known comorbidities, presented to neurology OPD with complete loss of vision for the past 2–3 days. She was in usual state of health 1 month back when she developed sudden, painless, bilateral loss of facial sensations and blurring of vision. The vision had been progressively worsening since, and for the past 2–3 days she could not see anything at all.

On further questioning, she said that for the past 3 months she had noticed gritty sensation and excessive watering of both eyes along with dryness of mouth. These symptoms had been worsening progressively since then. However, she did not seek any medical advice for these symptoms, nor used any medications, as it was not interfering with her daily life activity.

She denied having any similar symptoms before. She had no associated complaints of diplopia, eye pain, and

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redness of eye, dysphagia, hearing difficulty, headache, fever, rash, or focal weakness. There was no history of trauma before these symptoms. Her past medical and surgical history was insignificant; there was no previous hospitalization or recent travel history. She is unmarried, sexually inactive, and has no drug addictions. She has no known drug allergies and is not on any long-term medications. Her family history was insignificant for any long-term illness.

On physical examination, she appeared to be a young lady, alert, anxious, oriented in time, place and person, not in acute distress. She was vitally stable. Her neurological examination revealed bilateral loss of sensation on ophthalmic, maxillary, and mandibular division of trigeminal nerve. There was no facial asymmetry noticed; all other cranial nerve examinations were normal. She had normal tone, power, and bulk in all four limbs. Superficial and deep reflexes were intact and cerebellar signs were normal. Plantars were bilaterally down-going and gait was normal.

On eye examination, there was no conjunctival redness, and pupils were bilaterally round, regular, and reactive to light. However, corneal opacities were seen in both eyes. Corneal reflex was absent. On fundoscopy, red reflex was absent and hence fundus could not be visualized.

The following investigations were ordered. Complete blood picture, electrolytes, renal function tests, liver function tests, and thyroid profile were all within normal limit. Erythrocyte sedimentation rate was slightly elevated (39 mm/h). A complete autoimmune profile, as shown in Table 1 below, was advised; it showed raised anti-SSA (Ro) antibodies.

MRI brain with contrast was normal. Electrophysiological blink reflex testing was done, which showed complete trigeminal loss on both sides of the face; refer to Table 2 below. Facial nerve was normal.

Ophthalmology consultation was sought. They diagnosed her with neurotrophic corneal ulceration

Table 1 The results of the autoimmune screening

Tests	Result	Reference
Anti-Sm antibodies	Negative (0.1)	<5–Negative5–10 – Equivocal>10 – Positive
Anti-RNP antibodies	Negative (0.1)	<5 – Negative5–10 – Equivocal>10 – Positive
Anti-SSA (Ro) antibodies	Positive (36)	<5 – Negative5–10 – Equivocal>10 – Positive
Anti-SSB (La) antibodies	Negative (0.1)	<5 – Negative5–10 – Equivocal>10 – Positive
Anti-Jo-1 antibodies	Negative (0.1)	<5 – Negative5–10 – Equivocal>10 – Positive
Anti-Scl-70 antibodies	Negative (0.1)	<5 – Negative5–10 – Equivocal>10 – Positive
Anti-Ku antibodies	Negative (0.1)	<5 – Negative5–10 – Equivocal>10 – Positive
Anti-PCNA antibodies	Negative (0.1)	<5 – Negative5–10 – Equivocal>10 – Positive
Histone antibodies	<0.1	<5 – Negative5–10 – Equivocal>10 – Positive
dsDNA antibodies	<0.1	<5 – Negative5–10 – Equivocal>10 – Positive
Nucleosome antibodies	<0.1	<5 – Negative5–10 – Equivocal>10 – Positive
c-ANCA antibodies	<0.1	<5 – Negative5–10 – Equivocal>10 – Positive
p-ANCA antibodies	<0.1	<5 – Negative5–10 – Equivocal>10 – Positive

Table 2 Direct facial nerve stimulation and blink reflex study

	Onset latency (ms)	Amplitude (mV)
Direct facial nerve stimulation		
Right	3.84	1.59
Left	3.30	1.97
	R1. latency/amplitude (ms/mV)	R2. latency/amplitude (ms/mV)
Blink reflex study		
Right		
Ipsilateral	NR	NR
Contralateral	NR	NR
Left		
Ipsilateral	NR	NR
Contralateral	NR	NR

Direct facial nerve stimulation test showed mildly prolonged-onset latency with normal amplitude. Blink reflex study shows no ipsilateral R1 and no ipsilateral and contralateral R2 on both sides. This is an abnormal study suggestive of complete lesion of trigeminal sensory nerve fibers, on both sides. NR, no response.

secondary to trigeminal nerve palsy. The patient was counselled regarding corneal transplantation. After consent, corneal transplantation was done, following which only mild improvement in her vision was seen.

She was also referred to a rheumatologist for opinion. She was diagnosed as having SS and started on symptomatic treatment. She was prescribed tear drops to help her with gritty eye sensation and for dryness of mouth she was started on oral secretagogue, both of which she found very helpful. She was also prescribed steroids (prednisolone×1 mg/kg/day×3 months), but as no improvement was seen it was tapered off.

Discussion

SS is a rare chronic autoimmune disorder characterized by diffuse lymphocytic infiltration of exocrine glands. Autoimmune exocrinopathy of salivary glands leads to dryness of mouth and that of lacrimal gland leads to dryness of eyes, both of which are chief symptoms of the disease [3].

American–European consensus Sjogren's criteria is the most widely used criteria for diagnosis of SS [6]. Table 3 below shows the diagnostic criteria.

Our patient was diagnosed with Sjogren disease according to the criteria mentioned in Table 3. She had ocular and oral symptoms for a few months, Schirmer's test was positive, and both anti-SSA and anti-SSA antibodies were positive.

SS can be classified as primary disease or secondary to other connective tissue disorders. The disease mostly affects middle-aged individuals and women (female : male ratio is 9 : 1) [4].

At least one-third of the patients with SS present with systemic manifestations including neurological, renal, pulmonary, articular, and gastrointestinal [5]. Neurological manifestations can be categorized into two: central nervous system (CNS) and peripheral nervous system. CNS involvement varies from headache to aseptic meningitis. Peripheral nervous system manifestations include sensory polyneuropathy, polyradiculopathies, autonomic neuropathies, and cranial nerve palsy. The mechanism behind CNS involvement in the disease remains unknown [6]. To the best of our knowledge, there has been no case of bilateral trigeminal palsy presenting as a complication of SS reported so far.

SS is associated with severe functional impairment and negatively affects the quality of life. Treatment comprises two parts: symptomatic relief and improvement in quality of life. Symptomatic relief includes use of lubricant eye drops and gustatory stimulating agents for dryness of eye and mouth, respectively. [7]. Studies have shown that rituximab is effective in improving systemic manifestations and quality of life in patients with primary SS [8]. High dose of steroids along with immunosuppressant is recommended in patients with CNS involvement. Managing patients with SS requires a multidisciplinary approach involving many specialities depending on systemic complications [9].

Table 3 The American–European consensus criteria for the diagnosis of Sjogren syndrome

Ocular symptoms	Any one of the following
	Dryness of eyes for ≥3 months
	Foreign-body sensation
	Use of artificial tears more than three times a day
Oral symptoms	Any one of the following
	Dryness of mouth ≥3 months
	Recurrent or persistent swelling of salivary glands
	Frequent need of liquids to help swallow dry food
Ocular signs	Any one of the following
	Positive vital dye test
	Abnormal Schirmer's test
Oral signs	Any one of the following
	Abnormal salivary scintigraphy or abnormal parotid sialography
	Unstimulated salivary flow ≤1.5 ml/15 min
Histopathology	Lip biopsy demonstrating focal lymphocytic sialoadenitis with a focus score of ≥1 per 4 mm^2
Autoantibodies	Any one of the following
	Anti-SSB (La)
	Anti-SSA (Ro)
Primary Sjogren syndrome	Any 4 out of 6 criteria above or any 3 of the last 4 criteria

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Conflicts of interest

There are no conflicts of interest.

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