A Rare Neurological Involvement in Sjögren's Syndrome: Abducens Nerve Palsy

Sjögren Sendromunda Nadir Görülen Nörolojik Bir Tutulum; Abdusens Sinir Felci

Abducens Nerve Palsy and Sjögren’s Syndrome

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Abstract
Sjögren’s syndrome (SS) is an autoimmune disorder characterized by lymphocytic infiltration of exocrine organs. Although neurological involvement occurs in approximately one quarter of patients, involvement of cranial nerves is a relatively rare occurrence. Here a rare case of cranial neuropathy related to SS is reported.

Keywords
Sjögren’s Syndrome; Cranial Neuropathy; Abducens Nerve Palsy

Özet

Anahtar Kelimeler
Sjögren Sendromu; Kranial Nöropati; Abdusens Sinir Felci
Introduction

Sjögren’s syndrome (SS) is an autoimmune disease characterized by chronic lymphocytic infiltration of exocrine glands. The prevalence of the disease ranges from 0.5 to 5%. SS can occur at all ages, with predominance in females between 40 and 50 years of age. The disease is nine times more commonly seen in females than in males. Primary Sjögren’s syndrome (pSS) accounts for approximately 50% of all SS patients. Extraglandular symptoms such as articular, neurological, pulmonary, and hematological involvement are common over the course of the disease [1]. Neurological involvement often manifests as sensorial polyneuropathy. The involvement of cranial nerves is relatively rare [2]. In the published literature, isolated abducens nerve palsy has been reported in only 1 patient [3]. The current report presents a case of pSS with the involvement of cranial nerves.

Case Report

A 70-year-old female patient presented to an ophthalmology outpatient clinic complaining of double vision, headache, and nausea. The physical examination of the patient revealed limitation in lateral and medial gaze and ptosis. The patient was referred to the department of neurology with the suspicion of abducens nerve palsy (Figure 1). The patient’s medical history was not remarkable, with the exception of preexisting hypertension and cranial diffusion on magnetic resonance imaging (MRI). MR venography was performed to exclude intracranial hypertension, sinus vein thrombosis, and intracranial space occupying lesions. Carotid-vertebral artery computed tomography (CT) angiography did not show any intracranial aneurysm, and Doppler ultrasonography of the temporal arteries revealed normal findings. Orbital MRI was also normal. The examination of the cerebrospinal fluid (CSF) did not show pathological findings (angiotensin converting enzyme (ACE), adenosine deaminase, Lyme serology, tuberculosis culture, or brucella). To exclude myelopathy, cranial nevus involvement, and hematological involvement are common over the course of the disease [1]. Neurological involvement often manifests as sensorial polyneuropathy. The involvement of cranial nerves is relatively rare [2]. In the published literature, isolated abducens nerve palsy has been reported in only 1 patient [3]. The current report presents a case of pSS with the involvement of cranial nerves.

Figure 1. Abducens nerve palsy in the patient

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Discussion

Sjögren’s syndrome is known to cause neurological involvement in approximately 25-30% of cases [2]. In the cohort of Jamiloux et al., neurological involvement occurred in 95 (22%) out of 420 patients with SS. SS may affect both the peripheral nervous system (PNS) and central nervous systems (CNS) (Table 1). The pathogenesis of neurological involvement has not been fully elucidated. Vasculitic lymphocytic infiltration of vasa nervorum has been the most commonly implicated mechanism in the involvement of the PNS regardless of the presence of necrosis [4]. Two hypotheses have been set forth to explain CNS involvement. The first hypothesis suggests direct infiltration of the CNS by mononuclear cells, whereas the second hypothesis suggests vascular involvement. Vasculopathy has been considered to be associated with the presence of anti-Ro and antineural antibodies [5].

Sensory polyneuropathy is the most common type of PNS involvement, occurring in approximately 20% of cases [2]. The patients with Sjögren’s often present with numbness, burning, and pain originating in distal parts of the extremities. Mononeuropathy is relatively rare in SS. Mononeuropathy encompasses mononeuritis multiplex, cranial neuropathy, and trap neuropathies. Cranial neuropathies show symptoms depending on the cranial nerve involved. Among cranial nerves, the trigeminal nerve and optic nerves are the most commonly involved [3]. Visual changes can be quite profound in Sjögren’s. The most worrisome is inflammation around or behind the eye, called optic neuritis or retrobulbar neuritis. This is treatable, but there is potential risk of loss of vision. This type of inflammation can also be seen in a variety of other neurologic conditions. There

Table 1. Neurological involvement in Primary Sjögren’s Syndrome

<table>
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<tr>
<th>PNS</th>
<th>CSS</th>
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<tbody>
<tr>
<td>Sensory neuropathy</td>
<td>Focal manifestations</td>
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<tr>
<td>- Small fiber neuropathy</td>
<td>Aseptic meningencephalitis</td>
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<tr>
<td>- Sensory ataxic neuropathy</td>
<td>Myelopathy</td>
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<td>Sensorimotor neuropathy</td>
<td>Headache</td>
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<td>Mononeuritis multiplex</td>
<td>Cognitive disorders</td>
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<td>Demyelinating polyradiculoneuropathy</td>
<td>Mood disorders</td>
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<td>Cranial nerve involvement</td>
<td>Seizure</td>
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<td>Autonomic neuropathy</td>
<td>Pyramidal signs</td>
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<td></td>
<td>Tranverse myelitis</td>
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PNS: Peripheral nervous system, CSS: Central nervous system
is a similarity that sometimes bedevils us between multiple sclerosis and Sjögren’s involvement in the nervous system. The aspect that makes differentiating between the diseases most difficult is involvement of the eye in optic neuritis.

The treatment usually involves the use of immunosuppressive medications such as corticosteroids, cyclophosphamide, and azathioprine. Intravenous immunoglobulin therapy is used in refractory cases. However, intravenous immunoglobulin therapy has some disadvantages due to indefinite dosage, the duration of therapy, and high treatment costs. There is a need for randomized controlled studies in order to establish the effectiveness of this therapy [6]. Rituximab, an anti-CD 20 antibody, offers promising results; however, further comprehensive studies are required in order to confirm its efficiency [7].

In conclusion, extraglandular organs can be involved in patients with Sjögren’s syndrome. In clinical practice, it should be considered that neurological involvement occurs in one quarter of SS cases. In fact, neurological complaints may be the first sign of SS. We emphasize that connective tissue disorders should be considered in patients presenting with similar complaints; it is prudent to evaluate these patients for the possibility of pSS.

**Competing interests**

The authors declare that they have no competing interests.

**References**


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