

## Combined Central and Peripheral Nervous System Involvement Revealing Primary Sjögren's Syndrome

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## Abstract

## Case Report

**Background:** Primary Sjögren's syndrome (pSS) is a systemic autoimmune disease primarily characterized by lymphocytic infiltration of exocrine glands leading to sicca symptoms. However, neurological manifestations may occur and can occasionally precede the classical glandular features, posing significant diagnostic challenges. **Case presentation:** We report the case of a 57-year-old woman who presented with progressive headaches and lower limb paresthesia evolving over five months. Neurological examination revealed distal sensory deficits without motor impairment. Brain magnetic resonance imaging (MRI) demonstrated bilateral capsulo-thalamic lesions associated with non-specific subcortical white matter hyperintensities. Electroneuromyography revealed axonal sensory neuropathy of the lower limbs. Immunological investigations showed positive antinuclear antibodies (ANA, 1 :640) along with anti-SSA and anti-SSB antibodies. Histopathological examination of a minor salivary gland biopsy confirmed focal lymphocytic sialadenitis (grade IV). A diagnosis of primary Sjögren's syndrome with combined central and peripheral nervous system involvement was established. The patient was treated with high-dose corticosteroids followed by cyclophosphamide, resulting in partial clinical improvement. **Conclusion:** This case highlights the heterogeneous neurological spectrum of primary Sjögren's syndrome and emphasizes the importance of considering this diagnosis in patients presenting with unexplained neurological symptoms, particularly when associated with sicca features. **Keywords:** primary Sjögren's syndrome; neuro-Sjögren; peripheral neuropathy; central nervous system involvement; autoimmune disease.

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## INTRODUCTION

Primary Sjögren's syndrome (pSS) is a chronic systemic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands, leading to the classical manifestations of xerostomia and xerophthalmia. Beyond glandular involvement, a wide spectrum of extraglandular manifestations may occur, affecting organs such as the lungs, kidneys, skin, and nervous system [1,2].

Neurological involvement has been reported in approximately 10–25% of patients with pSS and may involve either the central nervous system (CNS) or the peripheral nervous system (PNS) [3,4]. Peripheral neuropathies are the most frequently reported neurological manifestations, particularly axonal sensory neuropathies. In contrast, CNS involvement is less common but may present with a broad range of clinical and radiological features [5].

CNS manifestations may include white matter lesions, focal neurological deficits, cognitive impairment, or inflammatory and demyelinating-like lesions that can mimic other neurological disorders such as multiple sclerosis or neuropsychiatric systemic lupus erythematosus [5,6]. The variability of these manifestations often complicates diagnosis and may delay appropriate management.

Here, we report a case of primary Sjögren's syndrome revealed by combined central and peripheral neurological involvement.

## CASE PRESENTATION

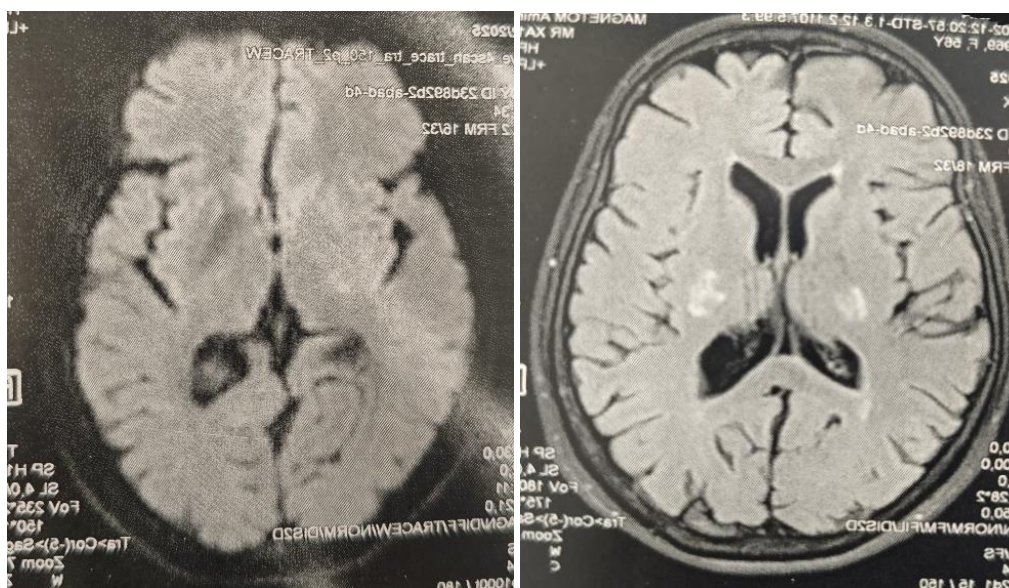
A 57-year-old woman with no significant past medical history except for a previous cholecystectomy was admitted for evaluation of suspected neurological involvement associated with Sjögren's syndrome.

The patient reported a five-month history of progressive diffuse headaches accompanied by paresthesia affecting the lower limbs. Neurological examination revealed distal hypoesthesia predominantly involving the left lower limb. No motor deficit or cranial nerve abnormalities were observed. Clinical evaluation also revealed a complete sicca syndrome characterized by both xerostomia and xerophthalmia.

Immunological investigations demonstrated positive antinuclear antibodies (ANA) at a titer of 1:640,

as well as positive anti-SSA and anti-SSB antibodies. Histological analysis of a minor salivary gland biopsy revealed focal lymphocytic sialadenitis classified as grade IV.

Brain MRI showed bilateral capsulo-thalamic lesions associated with non-specific subcortical white matter hyperintensities. Electroneuromyography confirmed the presence of axonal sensory neuropathy affecting the lower limbs (figure 1).



**Figure 1: Axial brain MRI (FLAIR and diffusion-weighted sequences) showing white matter signal abnormalities, suggestive of inflammatory involvement of the central nervous system in the setting of Sjögren's syndrome**

Laboratory tests revealed moderate lymphopenia with complement consumption or evidence of other systemic organ involvement.

The patient received high-dose intravenous corticosteroid therapy followed by cyclophosphamide. Partial clinical improvement was observed during follow-up.

## DISCUSSION

This case illustrates a relatively uncommon presentation of primary Sjögren's syndrome characterized by combined central and peripheral nervous system involvement.

Neurological manifestations of pSS are highly heterogeneous and may precede glandular symptoms in a significant proportion of cases, potentially delaying the diagnosis [2,3]. In the present case, neurological symptoms represented the initial manifestation of the disease, a pattern that has been increasingly recognized in recent studies.

Central nervous system involvement in pSS remains poorly understood and may present with non-specific radiological abnormalities. White matter lesions

are among the most commonly reported findings and may resemble those observed in demyelinating disorders such as multiple sclerosis [5,6]. The bilateral capsulo-thalamic lesions observed in our patient are consistent with previously reported imaging findings in neuro-Sjögren.

Peripheral neuropathy represents the most frequent neurological complication of pSS and typically manifests as axonal sensory neuropathy [9]. The coexistence of both CNS and PNS involvement, although relatively rare, has been documented in several clinical series [7].

The diagnosis of pSS relies on a combination of clinical features, immunological markers, and histopathological findings. In this case, the presence of sicca symptoms, positive anti-SSA and anti-SSB antibodies, and confirmatory salivary gland biopsy supported the diagnosis. Systemic lupus erythematosus was considered in the differential diagnosis but was excluded due to the absence of additional clinical and biological criteria.

Therapeutic management of neurological involvement in pSS is not yet standardized.

Immunosuppressive therapies such as cyclophosphamide or rituximab are often used in severe or progressive neurological forms [8,10]. The partial clinical improvement observed in our patient following corticosteroid and cyclophosphamide therapy is consistent with previously reported outcomes.

This case highlights the importance of considering primary Sjögren's syndrome in patients presenting with unexplained neurological manifestations, particularly when associated with sicca symptoms.

## CONCLUSION

Neurological involvement in primary Sjögren's syndrome is heterogeneous and may affect both the central and peripheral nervous systems. Combined involvement remains uncommon but should be recognized as a possible presentation of the disease. Early diagnosis and appropriate immunosuppressive therapy are essential to improve functional outcomes and prevent disease progression.

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