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# Cerebellar degeneration in primary Sjögren syndrome: a case report

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## Abstract

**Background:** Cerebellar degeneration is a rare and severe presentation of primary Sjögren syndrome. There are few case reports of cerebellar degeneration associated with different autoimmune diseases, especially with systemic lupus erythematosus and neuro-Behcet's disease. There are only six patients reported worldwide to be affected by cerebellar atrophy associated with primary Sjögren syndrome. In this report, we describe a patient with primary Sjögren syndrome who presented with ataxia due to cerebellar degeneration.

**Case presentation:** We report the case of a 37-year-old Chinese woman with primary Sjögren syndrome who presented with ataxia over 3 months associated with tremor of the limbs. Magnetic resonance imaging of the brain revealed bilateral cerebellar atrophy. Based on the presence of cerebellar signs with magnetic resonance imaging brain findings, she was diagnosed as cerebellar degeneration secondary to primary Sjögren syndrome. She was treated with methylprednisolone, hydroxychloroquine, and two cycles of monthly intravenous cyclophosphamide. Subsequently, she refused further treatment, and her neurological symptoms remained the same upon the last clinic review. Primary cerebellar degeneration is rarely associated with primary Sjögren syndrome. The pathogenesis of the neurological manifestations in primary Sjögren syndrome is unclear. Treatment involves corticosteroids and immunosuppressive agents with no consensus of a specific therapy for the management of primary Sjögren syndrome with central nervous system involvement.

**Conclusions:** Cerebellar degeneration is a rare presentation of primary Sjögren syndrome. Early diagnosis and treatment of this condition is needed to ensure a good outcome.

**Keywords:** Sjögren syndrome, Ataxia, Cerebellar atrophy, Cyclophosphamide

## Background

Primary Sjögren syndrome (PSS) is an autoimmune connective tissue disease, characterized by mononuclear infiltration and destruction of salivary and lacrimal glands leading to xerostomia and xerophthalmia. It predominantly affects female patients between 40 and 50 years of age [1].

SS may occur as primary or secondary to another connective tissue disease (mainly systemic lupus erythematosus, rheumatoid arthritis, or scleroderma). Approximately 35% of patients with primary Sjögren syndrome (PSS) suffer from systemic manifestations [2]. Neurological disorders are one of the extraglandular manifestations of the disease. In 25–60% of cases, the neurological symptoms preceded the diagnosis of PSS by 2 years. In the remaining patients, neurological disorders appeared 6–8 years after diagnosis [3].

The prevalence of neurological manifestations in PSS varies widely from 10% to 60% [4–6]. The most common neurological complication of PSS is peripheral neuropathy, particularly sensory polyneuropathy [7]. Central nervous system (CNS) involvement is much less common (2–25%) [8, 9]. Although ataxia due to PSS has also been

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