CASE REPORT

Unilateral Optic Neuritis in Primary Sjögren Syndrome Onset – a Case Report

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Abstract

Introduction. *Primary Sjogren's syndrome (pSSj) is a chronic, autoimmune disease that predominantly affects the lacrimal and salivary glands but is also responsible for extra-glandular disturbances. Although rare, optic neuritis (ON) may appear as initial manifestation and may be responsible for visual loss if diagnosis is delayed. The aim of the paper is to report a case of pSSj-induced ON as disease debut.*

Case presentation. *A 35-year-old patient with xerophthalmia presented an episode of sudden decrease in visual acuity of the left eye. Examination of the fundus revealed papillary edema, tortuous retinal vessels, non-reflex macula improved after methylprednisolone pulse-therapy. Immunological testing revealed high titer antinuclear antibodies (ANA) and intensely positive anti-Ro antibodies. Schirmer test was positive and parotid gland ultrasound was compatible with pSSj. Central nervous system damage is heterogeneous as clinical phenotypes in patients with pSSj. Prompt distinction between non-specific ON and pSSj-related eye involvement is mandatory for successful therapeutic strategy.*

Conclusions. *pSSj is a rare cause of ON that requires a detailed medical history, autoantibody determination, and minor salivary gland biopsy for confirmation. The ON response to methylprednisolone is promising and may improve patients' prognosis.*

Keywords: optic neuritis, xeropthalmia, primary Sjögren's syndrome, autoimmunity, autoantibodies.

Introduction

Primary Sjogren's syndrome (pSSj) is a chronic, progressive, autoimmune disease that predominantly affects the exocrine glands, namely the lacrimal and salivary glands, but almost half of patients can also experience extra-glandular manifestations [1].

The mechanism responsible for the glandular damage is the inflammatory process induced by lymphocytic infiltration and B cell hyperactivity [2].

SSj has a prevalence ranging from 1 to 3% in the general population. It is classified as primary (pSSj), if the sole condition or secondary (sSSj) if accompanied by other chronic conditions like rheumatoid arthritis, systemic lupus erythematosus or autoimmune thyroiditis [3]. However, both conditions mainly affect female patients aged over 50 years old, with an estimated gender ratio of 9:1 [4].

Systemic manifestations can target the skin, lungs or kidneys but also the peripheral nervous system, leading to the widely