

Multiple Cranial Nerve Enhancement on MRI in Primary Sjögren's Syndrome

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Abstract

A 77-year-old man showed bilateral abducens palsies and multiple cranial nerve enhancement on magnetic resonance images (MRI) and aseptic meningitis. He had xerophthalmia and xerostomia. Serum anti-SS-A and anti-SS-B antibodies were present. He had Sjögren's syndrome (SjS) and corticosteroid therapy ameliorated the symptoms. The cranial nerve enhancement and the cerebrospinal fluid findings were normalized. In patients with SjS, there have not been any reports of multiple areas of cranial nerve enhancement on MRI. We propose that in this case the aseptic meningitis and subsequent lymphocytic infiltration to the cranial nerves contributed to the multiple cranial neuropathy and multiple cranial nerve enhancement on MRI.

Key words: Sjögren's syndrome, multiple cranial neuropathy, cranial nerve enhancement, MRI

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Introduction

Sjögren's syndrome (SjS) is a systemic autoimmune disease characterized by xerophthalmia and xerostomia. A variety of neurological symptoms are described in SjS. Disease-associated neuropathy is one of the major neurological complications of SjS (1). Vasculitis in peripheral neuropathy and ganglionopathy in trigeminal or ataxic neuropathies have been reported as the main pathogenic etiology (1). However, the pathomechanism of disease-associated cranial neuropathies have never been clarified in detail. Recently, an immunologic cause was suggested to be involved in the etiology of cranial neuropathy in SjS (2, 3). We describe a patient with primary SjS complicated by bilateral abducens palsies, multiple cranial nerve enhancement and aseptic meningitis.

Case Report

A 77-year-old Japanese man was admitted to our hospital complaining of acute-onset bilateral abducens palsies and headache. Prior to admission, he was under treatment for diabetes mellitus and prostate cancer with bone metastasis. On examination, he showed bilateral abducens palsies. There

were no meningeal signs or symptoms of other cranial nerve involvement. The brain magnetic resonance images (MRI) with gadolinium enhancement disclosed multiple areas of cranial nerve enhancement, including the right oculomotor nerve, left trigeminal nerve, bilateral abducens nerves, and bilateral facial nerves (Fig. 1a, b). The cerebrospinal fluid (CSF) contained 69 mg/dL protein and 33/μL of mononuclear cells. The CSF level of prostate specific antigen (PSA) was not elevated (0.296 ng/mL). Cytology of the CSF was negative. Blood chemistry analysis was unremarkable except for elevation of the plasma glucose concentration (170 mg/dL) and an elevated erythrocyte sedimentation rate (ESR) (75 mm/h). The HbA1c was slightly elevated at 6.6%. Serum antinuclear, anti-SS-A and SS-B antibodies were positive. On ophthalmological examinations Schirmer's test was positive (0 mm of tear flow over 5 min). On dental examinations the unstimulated whole salivary flow was decreased (0.6 mL in 15 min). The salivary scintigraphy showed evidence of parotid gland involvement. Although a salivary gland biopsy did not demonstrate any sign of lymphocytic infiltration, he received a diagnosis of primary SjS (4). We administered methylprednisolone pulse therapy (1 g daily for 3 days in a week) 4 times. One month after starting treatment, abducens palsies were ameliorated. Follow-up

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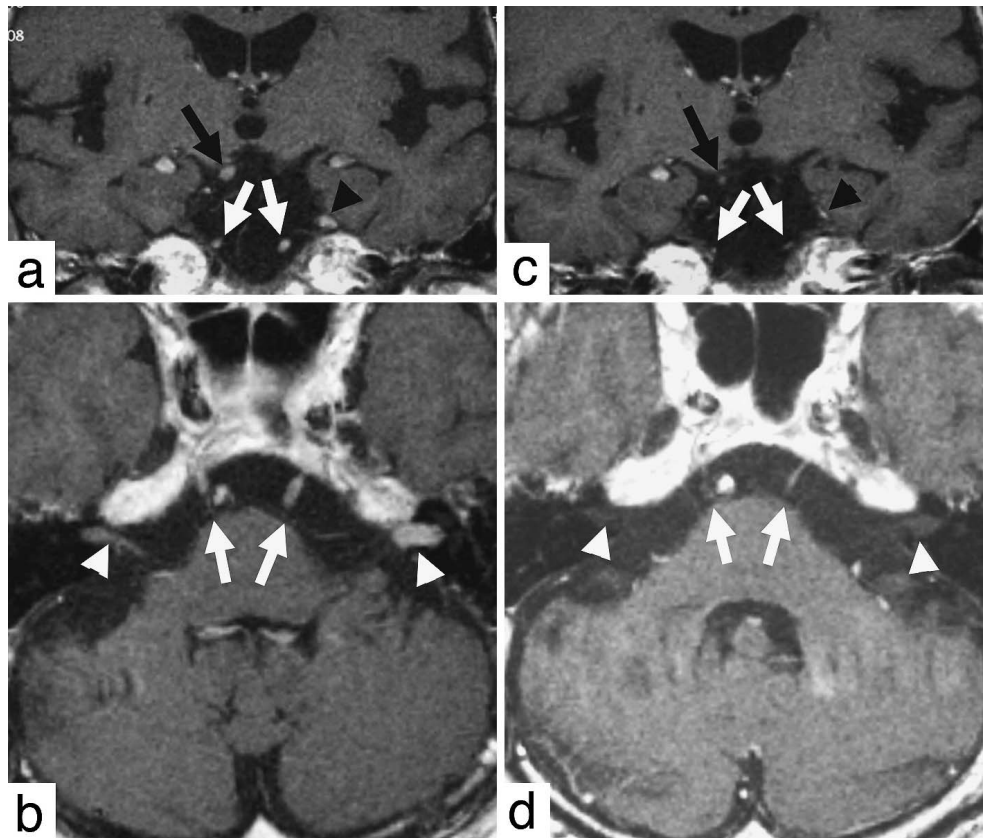


Figure 1. Magnetic resonance images (MRI) before treatment (a, b) displayed swelled and gadolinium-enhanced cranial nerves. MRI with gadolinium enhancement performed after treatment (c, d) disclosed improvement of the findings of the cranial nerves. Right oculomotor nerve, black arrow; left trigeminal nerve, black arrowhead; bilateral abducens nerves, white arrows; bilateral facial nerves, white arrowheads. Coronal sections (a, c); axial sections (b, d).

MRI no longer showed cranial nerve enhancement (Fig. 1c, d), and the CSF findings were normalized. He had no episode of relapse for a year after clinical remission.

Discussion

We described a patient with primary SjS showing multiple cranial neuropathies, multiple cranial nerve enhancement and aseptic meningitis. Although aseptic meningitis is a common neurological complication in patients with SjS (5, 6), multiple cranial neuropathy is relatively rare (1). There have not been any reports of multiple regions of cranial nerve enhancement on MRI in patients with SjS. Several conditions, such as inflammation, demyelination, tumor, meningoencephalitis and diabetes mellitus, may account for the cranial nerve enhancement (7). Although diabetic patients may have paralysis of multiple cranial nerves, these paralyzes are not responsive to corticosteroid therapy (8). The present patient also had prostate cancer, however, there was no evidence of metastasis of the prostate cancer in the brain. In addition, there was no episode of relapse for a year after clinical remission. Improvement of the abducens palsies as well as both MRI and CSF findings after corticosteroid therapy indicated that SjS was the main etiology of cranial neuropathy in this patient. The pathomechanisms under-

lying cranial neuropathy in SjS have not yet been elucidated, except for trigeminal neuropathy due to ganglionopathy. Vasculitis and subsequent axonopathy could lead to multiple cranial neuropathies (1). Recently, two possible mechanisms, vascular origin with damage to the vasa nervorum and an immunologic cause inducing lymphocytic infiltration of the nerve, have been suggested in oculomotor nerve palsy related to SjS (2). Lui et al reported a SjS patient with acute-onset left oculomotor nerve palsy showing rapid and almost complete recovery from nerve palsy after 2 months of corticosteroid and azathioprine treatments, and suggested that lymphocytic infiltrate, rather than a vasculitic process, was the cause of cranial neuropathy (3). The present patient showed rapid and complete recovery from abducens palsies and the MRI findings improved after corticosteroid therapies. It is proposed that aseptic meningitis and the subsequent lymphocytic infiltration to the cranial nerves contributed to the multiple cranial neuropathy and multiple areas of cranial nerve enhancement on MRI.

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