Transient Oculomotor Palsy Correlated with Nerve Enhancement on MRI in Chronic Inflammatory Demyelinating Polyneuropathy

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Abstract

A 40-year-old woman was admitted to our hospital because of double vision combined with left ptosis. Although at 25 years of age she had already been diagnosed with limb weakness associated with chronic inflammatory demyelinating polyneuropathy (CIDP), she had never experienced double vision until her latest condition. Neurological examination revealed left oculomotor palsy without other cranial nerve involvement. Serial magnetic resonance imaging (MRI) studies demonstrated a temporal correlation between clinical severity of oculomotor palsy and segmental enhancement of the oculomotor nerve. Gadolinium enhancement on MRI may be a significant finding indicating relapse of oculomotor involvement of CIDP.

Key words: chronic inflammatory demyelinating polyneuropathy, oculomotor palsy, magnetic resonance imaging, gadolinium enhancement, immunosuppressive treatment

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Introduction

Magnetic resonance imaging (MRI) has revealed nerve root enlargement with or without enhancement in chronic inflammatory demyelinating polyneuropathy (CIDP) (1). These MRI findings have been reported not only in spinal nerve roots including the cauda equina (2) but also in cranial nerves (3). The MRI findings of oculomotor involvement in CIDP have also been reported previously (4). However, whether or not the MRI findings correlate with clinical severity of oculomotor palsy remains unclear. We report our observation of transient oculomotor palsy coinciding with segmental enhancement of oculomotor nerve on MRI in a patient with CIDP.

Case Report

A 40-year-old woman was admitted to our hospital because of double vision combined with left ptosis. The patient was diagnosed with CIDP at 25 years of age. The examination at age 25 revealed symmetric proximal and distal weakness [Medical Research Council (MRC) grade 3-4], distal dominant sensory deficit in both lower limbs, and reduced deep tendon reflexes. Albuminocytologic dissociation in CSF (total protein: 98 mg/dL; cell count: 3 lymphocytes/mm³) was present. Nerve biopsy demonstrated distinct demyelination, onion-bulb formation and mild lymphocyte infiltration in the hypertrophic cauda equina tissue. Nerve conduction study showed prolonged distal latency, slowed conduction velocity (10-20 m/s), delayed or absent F waves, and partial conduction block in the 4 limbs. These findings fully satisfy the diagnostic criteria for “definite CIDP” proposed by American Academy of Neurology Ad Hoc Sub-committee (5). Although she had already developed limb weakness associated with CIDP, she had never experienced double vision until her latest condition.

On admission, she was conscious and well-oriented. Neurological examination revealed left oculomotor palsy with ptosis, mydriasis and extraocular muscle weakness (Fig. 1).
Figure 1. Neurological examination showed left oculomotor palsy at onset. [The patient has given consent to the use of these photographs.]

Figure 2. Axial T1-weighted MR images enhanced by gadolinium at the cavernous sinus level: during relapse of CIDP with left oculomotor palsy (left), at 2 months (middle) and at 4 months (right) after treatment. The segmental enhancement by gadolinium (left arrow) eventually disappeared after treatment (right arrow). Note that the segmental lesion remained slightly enhanced even when clinical recovery of the left oculomotor palsy was observed at 2 months after treatment (middle arrow). The patient was instructed to tilt her head backward so that the entire length of the oculomotor nerve around the cavernous sinus could be scanned in the same slice.

She also complained of left eyeball pain during eye movement. Other cranial nerves were intact. Motor testing revealed distal dominant weakness in both lower limbs (MRC grade 3), more marked in the left peroneal nerve-innervated muscles (MRC grade 2). Deep tendon reflexes in the limbs were absent and flexor plantar response was observed bilaterally. Superficial sense was impaired in the lower limbs. Vibration sense was intact in the upper limbs and decreased in the lower limbs. These motor and sensory findings had not changed significantly during the recent course of her illness. Cerebellar and autonomic functions were not involved.

Serial MRI studies revealed marked enlargement of the spinal nerve roots, in particular a markedly enlarged cauda equina, without gadolinium-enhancement in any segment of the peripheral nerves. Previous MRI scans had never shown any abnormality in the cranial nerves until the oculomotor palsy occurred. The present MRI scan demonstrated segmental enhancement of the left oculomotor nerve along the cavernous sinus before treatment (Fig. 2). The other cranial nerves showed no abnormality on MRI.

Based on a diagnosis of relapsing CIDP, the patient was treated with intravenous steroid pulse therapy (methylprednisolone 1,000 mg/day for 3 days) followed by high-dose intravenous immunoglobulin. Left oculomotor palsy subsided gradually, and disappeared after 2.5 months from the start of treatment. Follow-up MRI showed disappearance of enhancement in the oculomotor nerve after successful treatment (Fig. 2).
Discussion

The correlation among oculomotor palsy, MRI findings and therapeutic effects in patients with CIDP remains to be elucidated. The present case showed a temporal correlation between disease activity and gadolinium enhancement on MRI in a segment of the oculomotor nerve. Previous MRI studies have shown that the facial nerve may be enhanced by gadolinium on MRI even in normal subjects (6), but gadolinium enhancement in a normal oculomotor nerve has not been reported although such enhancement was observed in patients with oculomotor nerve palsy and diverse underlying diseases (7). Therefore, segmental enhancement of the oculomotor nerve is regarded as “abnormal”. The possible mechanism for isolated oculomotor nerve involvement in the present case may be localized inflammatory demyelination. Localized enhancement may indicate breakdown of the blood-brain barrier by immunological inflammatory reaction in the acute relapsing stage of CIDP. Immunosuppressive treatment in the acute stage is expected to be effective to improve clinical condition and resolve gadolinium enhancement on MRI, and was observed in the present case. However, this therapy may be ineffective in the chronic stage characterized by hypertrophic nerve changes derived from Schwann cell proliferation and onion bulb formation by repeated segmental demyelination and remyelination.

Isolated demyelination is a characteristic feature of multifocal acquired demyelinating sensory and motor (MADSAM) neuropathy. Cranial nerve involvement, such as optic neuritis as well as oculomotor, trigeminal, and facial palsies, has been reported (8-10). In the present case, however, serial nerve conduction studies consistently demonstrated symmetric diffuse demyelinating findings in the upper and lower limbs, which negate a diagnosis of MADSAM neuropathy.

Nerve conduction study is useful to detect a newly developed conduction abnormality which may reflect relapse of CIDP (11). However, such electrophysiologic assessment is not always applicable to investigate cranial nerve involvement. Gadolinium enhancement on MRI may be a significant finding indicating relapse, at least for oculomotor involvement of CIDP.

References


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