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Case Report

Neuromyelitis Optica Mimicking Intramedullary Tumor

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Neuromyelitis optica (NMO) is considered to be a rarer autoimmune disease than multiple sclerosis. It is very difficult to make a diagnosis of MNO for doctors who are not familiar with its clinical features and diagnostic criteria. We report a case of a young female patient who had been suffering motor weakness and radiating pain in both upper extremities. Cervical MRI showed tumorous lesion in spinal cord and performed surgery to remove lesion. We could not find a tumor mass in operation field and final diagnosis was NMO. NMO must be included in the differential diagnosis of lesions to rescue the patient from invasive surgical interventions. More specific diagnostic tools may be necessary for early diagnosis and proper treatment.

Key Words : Neuromyelitis optica · Autoimmune disease · Spinal tumor.

INTRODUCTION

Neuromyelitis optica (NMO) is a severe demyelinating syndrome of the central nervous system by attacks of optic neuritis and myelitis[17,26]. The incidence and prevalence of NMO are unknown[1,9]. Moreover, NMO is considered to be a rarer autoimmune diseases than multiple sclerosis in Europe[7].

It is very difficult to make a diagnosis of MNO for doctors who are not familiar with its clinical features and diagnostic criteria. Some patients who present motor weakness with contiguous, intramedullary cervical spinal cord lesion without trauma history are misdiagnosed with intraaxial neoplasia of the spinal cord with other progressive disease. Often, when patients' motor weakness is progressive and deteriorated with newly appearing sensory symptoms, doctors, particularly surgeons, are apt to perform surgery[21]. Here, we present a case report on a patient who ultimately underwent cervical laminectomy for decompression and biopsy. The patient was treated with a high-dose steroid and physical therapy. After all, the diagnosis of her disease by histological, clinical, and radiological means was Neuromyelitis optica (NMO)[17].

Recent revised diagnostic criteria for neuromyelitis optica include two absolute requirements and 2 out of 3 supportive criteria. The two absolute requirements are optic neuritis and acute myelitis.

The three supportive criteria are contiguous spinal cord MRI lesion extending over ≤3 vertebral segments, brain MRI not meeting the diagnostic criteria for multiple sclerosis (MS), and seropositive NMO-IgG[4,11,13,27]. The diagnosis and prognosis of NMO are different from MS. Early aggressive treatment of NMO may be more efficient in preventing permanent neurologic deficit than early treatment of MS[3,26,28].

In most cases of acute neurologic deterioration with contiguous, intramedullary cervical spinal cord lesion, immediate surgical approach and decompression of the lesion are performed. However, when a sufficient and complete diagnosis of the lesion is made, as in our presenting case, decompressive operation can be avoided and achieve a better prognosis.

CASE REPORT

This 20-year-old woman had no other medical history. The patient had been suffering motor weakness in both upper extremities and radiating pain in both hands since 3 weeks ago. She experienced progression of pain, sensory changes, and lopsided motor weakness to the left side of the body. At the initial physical examination, a motor grade of both upper extremities was determined as grade 4 and no weakness was found in lower extremities. She was generally mildly hypertonic, although there was no Babinski sign or clonus. She complained of mild radiating pain in

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both hands, which was not one-sided with inexplicit dermatome. The patient underwent brain and whole spine MRI, which revealed a heterogeneously enhancing mass in the central posterior aspect of the spinal cord at the C1-6 level (Fig. 1). Brain MRI showed no specific findings. She was suspected of a tumorous condition, particularly ependymoma. Following this, she was admitted to our hospital for further evaluation of neurologic change. On the second hospital day, her motor function in both upper extremities suddenly deteriorated from grade 4 to 3. Cervical MRI was performed again, which revealed further extension of the hyperintense lesion on T2 weighted image and more intramedullary enhancing lesion on the T1 weighted image than that on the initial MRI image (Fig. 2). We hypothesized that the origin of neurologic deterioration was spinal cord compression by tumor bleeding. We immediately started intravenous steroid therapy and carried out total laminectomy C2-6 for decompression with surgical biopsy.

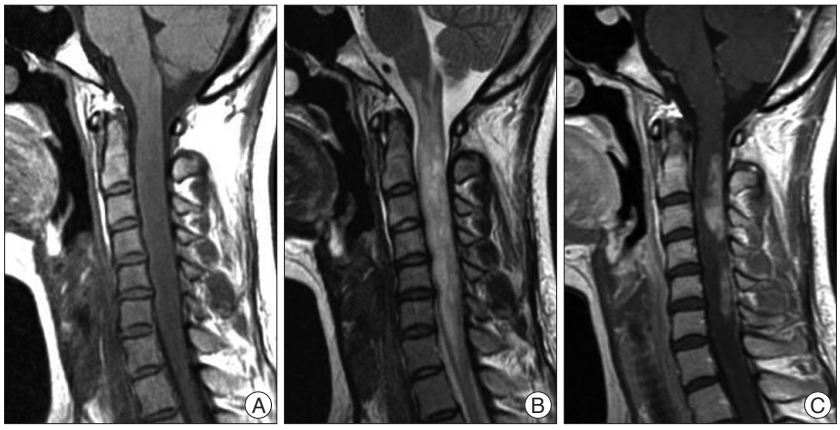

Fig. 1. Initial cervical MRI sagittal images show heterogenous enhancement of elongated shaped mass like lesion mimics tumorous condition from C1 to C6 levels. A : T1 weighted image. B : T2 weighted image. C : T1 weighted enhanced image.

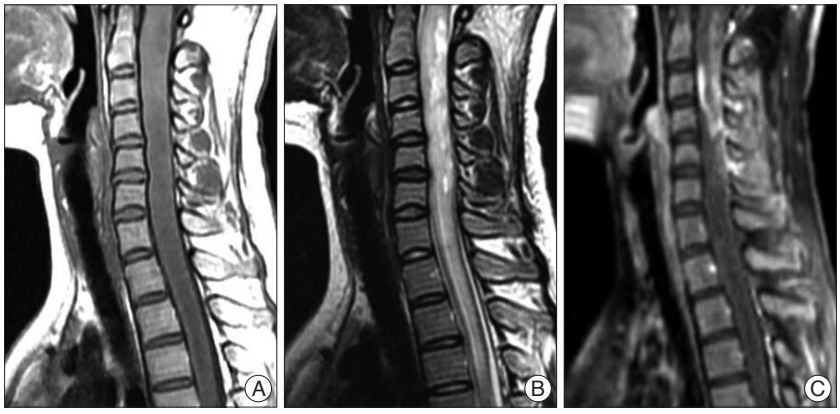

Fig. 2. Cervical MRI which was performed after neurologic deterioration shows newly appeared enhancing lesion in the anterior aspect of spinal cord from C2 to C4 level. A : T1 weighted image. B : T2 weighted image. C : T1 weighted enhanced image.

We performed C2-6 laminectomy and after opening the dura, the spinal cord was edematous but otherwise normal (Fig. 3). In the operation field, there was no tumor like lesion, discoloration or hypervascularization. We obtained frozen biopsy from a site near the lesion which was obtained consistent with astrocystic cell proliferation. Due to the gross normal finding and no tumorous frozen biopsy, we stopped the removal procedure of the mass lesion to prevent another potential spinal injury. The operation was closed by lateral mass screw fixation and fusion C2-6.

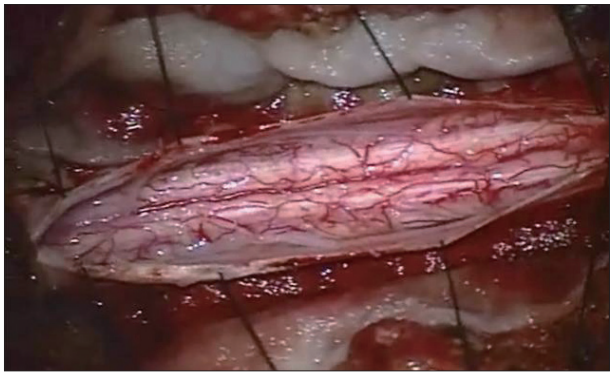

Fig. 3. Intraoperative picture shows no definite lesion which is suspected as tumorous condition except a mild edema.

On the first postoperative day, her vital signs were stable, and suffered from nuchal pain, but her motor weakness remained the same. However, from the second postoperative day, motor weakness in both upper extremities became gradually better along with improvement of other sensory symptoms. On the twelfth postoperative day, permanent biopsy revealed abundant histiocytic collection, perivascular lymphocytic inflammation and reactive astrocytic proliferation. There was no evidence of a neoplastic process and no oligoclonal bands. The pathologist strongly suggested the diagnosis of neuromyelitis optica. Only then did we investigate her ophthalmic symptoms with a suspicion of another disease. She recalled her visual disturbance 5 years ago. Since then, there was no symptom until visit our department. We referred her to ophthalmology and revealed optic atrophy with disc pallor with optic neuritis on her left eye. Finally, her diagnosis was determined on NMO.

On the fourteenth postoperative day, her motor grade was improved to grade 5 and the patient could perform her daily tasks including eating food. We continued postoperative steroid treatment and started rehabilitation. After rehabilitation, she regained most of her upper-extremity motor functions and her ability to walk. Her pain significantly disappeared. When she

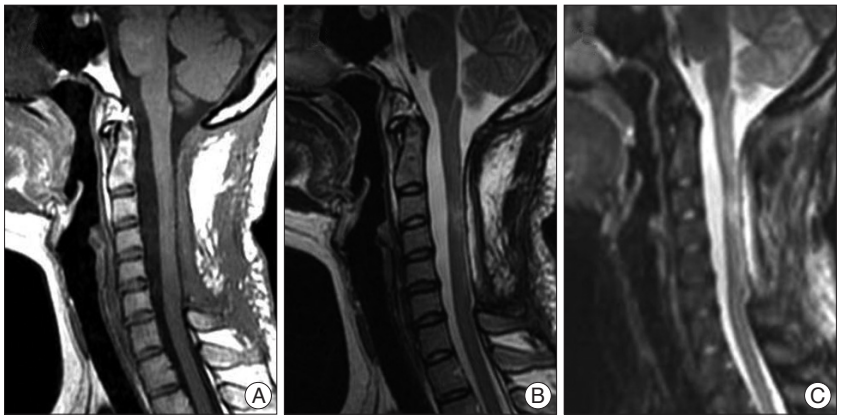

Fig. 4. Postoperative cervical MRI (after 3 months from surgery) shows improvement of cord swelling and remaining focal high signal foci on T2 weighted image. A : T1 weighted image. B : T2 weighted image. C : T1 weighted enhanced image.

was discharged from the hospital, there remained only mild nuchal pain. After 3 month from surgical intervention, her follow-up cervical MRI showed dramatic improvement (Fig. 4).

DISCUSSION

Many non-neoplastic lesions are misdiagnosis as intramedullary spinal cord tumors through radiographic findings and clinical symptoms. More specifically, the following misdiagnoses are made : infectious disorders (tuberculosis, fungal, bacterial), demyelination (MS), granulomatous disease, vascular lesions and syringomyelias[20].

It is difficult to distinguish these lesions from neoplastic spinal cord tumors from only case history and physical examination[10]. In substance, we can get conclusive evidence through biopsy of the lesion, but it can be very invasive and cause irreversible neurologic injury. Thus, in our case, the overall comprehension of NMO was helpful to manage the patient who was likely to be misdiagnosed. A sound knowledge of the differential diagnosis between NMO and MS may also be of additional assistance.

In 1894, Devic described the clinical symptoms of NMO for the first time : optic neuritis and acute transverse myelitis. His patients had monophasic or relapsing courses of neuromyelitis optica[19]. Improvements in biomarkers and immunoassay techniques gave rise to the revised definition of neuromyelitis optica[27]. The clinical presentation of optic neuritis relapse is more commonly unilateral than bilateral with ocular pain, and relapse of optic neuritis and myelitis usually appear gradually rather than rapidly. Most relapses worsen over the course of few days and then slowly improve after severe neurologic deterioration[26]. The followings are typical symptoms of relapse : ocular pain with visual loss, myelitis with severe symmetric paraplegia, sensory change below the lesion and bladder dysfunction[26].

We can typically see central scotoma, although other visual changes such as blindness, bitemporal hemianopsia are also detected by the visual field test in NMO patients[2,14].

The symptoms of transverse myelitis are para- or tetraparesis below the lesion, an almost symmetrical sensory change and sphincter dysfunction[15].

In our case, the patient's NMO visual symptoms were not clear. Three months later, she said that had experienced visual blurring 3 years-ago. She had normal motor power, no sensory deficit (even nuchal pain), and no visual symptoms.

Most patients with NMO have relapsing episodes of optic neuritis and myelitis, rather than a monophasic course. Relapse occurs within 1 year in 60% of patients and within 3 years in 90%[17,26,28].

Recovery is usually incomplete, and most patients follow a course of early irreparable disability due to frequent and severe relapses. Within 5 years of disease onset, more than 50% of patients with relapsing NMO become blind in one or both eyes or require ambulatory help. Risk factors of a poor prognosis are the number of relapses in the first 2 years of disease activity, the severity of the first attack and pre-existing autoimmune disorder.

In studies of broad spectrum NMO related disorders, the 5-year survival rate of NMO was 68%. The most frequent cause of death is neurogenic respiratory failure[4]. Luckily, our patient did not undergo relapsing episodes after the first attack and on her visual field revealed no central scotoma.

Spinal cord lesions contiguous over 3 vertebral levels are the most convincing findings for the diagnosis of NMO, and spinal cord lesions contiguous over 2 more vertebral segments are rarely found in MS[22,27]. The localization of cord lesions in the axial plane has various diversities, but is predominantly centrally located in AQP-4 (aquaporin-4) antibody positive patients[3].

At an early stage of NMO, spinal MRI may show typical patterns of NMO, such as snake-eye or owl-eye indicating spinal artery ischemia[8]. In brain MRI, most lesions are non-specific and asymptomatic in NMO, except for optic nerve enhancement of optic neuritis[5,23]. But, in MS, MRI findings of brain is mostly periventricular white-matter abnormal lesions.

As NMO-IgG, Aquaporin-4 (AQP-4) antibody in the serum is directed against AQP-4, osmosis-driven, and bidirectional water channel expressed on the foot processes of astrocytes in the CNS. Positive for NMO-IgG/AQP4 antibodies secures credible evidence for the diagnosis of NMO (more than half the cases for NMO patients) and is a prognostic factor for high-risk disease[6,16,18]. For treatment of NMO, we must bear in mind two mainpoints : one is to manage the inflammatory damage in acute sudden attacks and the other one is maintenance treatment to avoid relapses. The first depends on high dose intravenous corticosteroids and plasmapheresis and the second on low-dose corticosteroids and immunossupressants[12,24,25]. Additionally, low AQP-4 antibody titers by immunosuppressant treatment can reduce the occurrence of relapses[25,29]. Frequent relapsing

NMO attacks are related to permanent neurologic disability. Thus, it is momentous to commence treatment as early as possible to avoid new relapses and further disability.

The serum sample was referred by another research institute, but the titer was low and not specific for diagnosis. A diagnosis of non-neoplastic disease on cervical lesion must have underlying laboratory investigations and diagnostic procedures before invasive surgical intervention. However, by the surgeon's inclination, surgery may be considered as an alternative choice of treatment, if the patient's neurologic deficit is rapidly deteriorated. We should closely observe changes of the patient's trivial symptoms and be aware of a minor possibility of rare diseases such as NMO that can cause catastrophic results. Intense studies regarding biomarkers and immunoassay techniques are required for noninvasive, cost-effective diagnosis

CONCLUSION

A sudden motor deterioration with contiguous, intramedullary cervical spinal cord lesion could be misdiagnosed as a tumorous condition, but may actually be the symptoms of demyelinating diseases, such as MS and NMO. Early diagnosis and treatment of NMO determine the prognosis and sequelae of the disease. Thus, appropriate diagnosis results in suitable medical treatment (although it may require surgical approach), if careful attention is drawn to clinical clues, including visual symptoms and relapsing neurologic events. NMO must be included in the differential diagnosis of these lesions to rescue the patient from invasive surgical interventions. More specific diagnostic tools may be necessary for early diagnosis and proper treatment.

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