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Case Report

Guillain-Barré Syndrome Combined with Acute Cervical Myelopathy

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Authors describe a patient who developed a myelopathy associated with Guillain-Barré syndrome and cervical myelopathy. We provide radiological evidence of non-compressive herniated cervical intervertebral disc with cord signal changes and show the clinical and electrophysiological result of coexisting Guillain-Barré syndrome and cervical myelopathy. We tried to introduce and review the case of Guillain-Barré syndrome which was combined with cervical myelopathy to let us recollect the presumptive cause.

KEY WORDS : Guillain-Barré syndrome · Cervical myelopathy.

INTRODUCTION

Neck and arm pains with weakness are a common complaint that typically represents a spectrum of cervical spine disorders. In such patients with arm and neck pains with weakness, physicians must pay attention to the clinical history and examination of patients including diagnostic studies such as plain radiography, magnetic resonance image (MRI), and electrophysiologic study[5].

When patients complain the neck and arm pains with weakness of both upper and lower limbs, physicians should consider the various causes from trauma of whiplash injury to psychogenic origin. Sometimes, neurosurgeons concentrate too much on cervical spinal disorder or surgical disease, and may pass over the rare causes of internal or non-surgical disease.

Here, we present a case to inform the neurosurgeons knowing the possibility of association with the Guillain-Barré syndrome (GBS) and cervical myelopathy with reviewing the literatures of GBS, that could have been miserable, if we had surgery. Also, we intended to emphasize that intensive and scrupulous neurologic examination is prerequisite especi-ally in decision of operation.

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CASE REPORT

A 39-year-old woman without significant medical history was presented to emergency department, complaining the severe neck pain and both arm tingling sensation with weakness of lower limbs. Her lower limb weakness was started a few days prior to admission. She could stand by herself but required something leaning on when walking. And, she felt the neck pain and both arm numbness after severe coughing. On physical examination, the patient's vital signs were normal and there was no special past history except 1 week of upper respiratory infection at 1 month ago.

On neurologic examination, lower limbs motor powers were estimated to be grade 4, and both hands grasping motor power were slightly reduced with decreased deep tendon reflexes (DTR). Her numbness and tingling sensation of both arms was not exactly match to the dermatome of arm. Hoff-man's sign were detected in both hands.

Emergency MRI scans of cervical spine revealed the spinal cord signal change and disc protrusion at C5-C6 level on sagittal images, however we could not detect the severe compression which might lead to such a signal change of spinal cord on axial images of MRI (Fig. 1).

We first presumed that these findings would be closely

related with acute cervical disc herniation aggravated by neck flexion during the severe coughing. So, we planned to perform the surgical decompression. Preparing the surgery, we detected the increased level of plasma C-reactive protein (CRP). On blood examination, CRP was 4.5 mg/dL, and two days later reached to 17 mg/dL. We just guessed high CRP was related with her upper respiratory infection, though it was started 1 month ago. On same day, left side peripheral facial nerve palsy was developed with complains of severe headache. At that time we decided to postpone the surgery and tried to figure out the other combined or underlying cause of myelopathy. To reveal the other cause, brain MRI was performed, however nothing was revealed. Electrophysiologic studies showed slowed nerve conduction velocities, partial motor conduction block, which appropriated for Guillain-Barré syndrome. Immunoglobulin therapies were initiated, because patient was very nervous for progressive worsening of her symptom, instead of surgery. To make confirmative diagnosis, we tried cerebrospinal fluid (CSF) study, however, non-specific findings were detected. A few days later, we persuaded patient to have another CSF studies, but she refused because she felt better than before. We made the conclusion that Guillain-Barré syndrome was combined with cervical myelopathy.

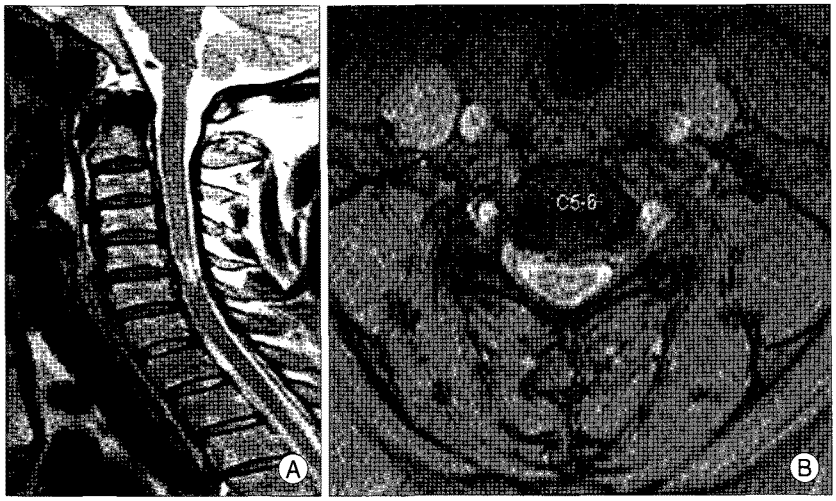

Fig. 1. This is cervical magnetic resonance image which was taken at symptom attack. A : Sagittal T1/T2WI show the non-compressive lesion of the spinal cord with signal change on C5/6 interspace. B : Axial T1/T2WI show no severe compression of spinal cord by protrusion of disc.

After 2 weeks of medical and physical treatment, symptom including pain was getting disappear and recovery was observed including motor power. She was discharged with full recovery after 3 weeks of treatment.

DISCUSSION

GBS is an autoimmune disorder and rapidly progressive, paralytic, inflammatory demyelinating polyradiculoneuropathy, causing progressive weakness of more than one limb, and reduced or absent tendon reflexes. Incidence is 1.6 to 2.3 cases per 100,000 populations, though its incidence depends on the region. And, it has been associated with antecedent bacterial and viral infections, administration of certain vaccinations, and other systemic illnesses. Two-thirds of patients develop neurologic symptoms 2-4 weeks after what appears to be a benign respiratory or gastrointestinal infection[1,4,8]. This would be one of the clues to our case. She had 1 week of upper respiratory infection at 1 month ago from attack of symptom.

The initial symptoms of GBS are paresthesias in the toes and fingertips, followed by lower extremity weakness that may ascend over hours to days to involve the arms, cranial nerves, and in severe cases the muscles of respiration. Symptoms and signs are relatively symmetric. Despite paresthesias, sensory deficits on examination are usually mild[4,8]. These symptomatic changes are not fully corresponding to our case, because her chief complaint was neck pain and tingling sensation of both arms, though progressive weakness of lower extremities and cranial nerve involvement was similar. In these points, we presumed that GBS was combined with cervical myelopathy. Reviewing the symptomatic changes, her lower limb weakness started first, before severe coughing which provoked the neck pain and both arm numbness. We assumed that she got GBS before cervical myelopathy was started, and at that time of meeting her at clinic, GBS and cervical myelopathy were coexisted.

Following description may be the clinical evidence of GBS. Progressive ascending lower limb weakness, peripheral facial palsy, decreased DTR and elcetrophysiologic findings. However, we could not make confirmative diagnosis by CSF study. This would be discussed later. On the other hand, cord signal changes of cervical MRI, Hoffman's sign and severe neck pain might be correspond to cervical myelopathy. Because Hoffman's sign could not be seen in such a demylinating disease and common MRI findings of GBS were mostly normal[8]. Most imaging studies are realized to exclude other conditions. Several reports have indicated that there was enhancement of the thickened nerve roots in the conus medullaris and cauda equina, although there are no abnormalities on precontrast images, because the spinal cord and nerve roots in the thecal sac generally do not take up much gadolinium due to the blood-nerve barrier or blood-brain barrier, so marked enhancement of nerve roots indicates a breakdown of the blood-

nerve barrier on enhanced MR imaging[2,4,6,8]. In current case, we should have had a whole MRI with enhancement to disclose the changes of nerve roots in the conus medullaris and cauda equine. Unfortunately, this was not done.

For diagnosing the GBS, electrophysiologic studies should demonstrate slowed nerve conduction velocities, partial motor conduction block, abnormal temporal dispersion, prolonged distal latencies. Elevated protein level in CSF may be observed, however, protein level may not yet be elevated or observed until 1-2 weeks after the onset of weakness, rarely they do remain persistently normal[2,5]. In our case, electrophysiologic study showed comparable to GBS, but we hesitated to make confirmative diagnosis of GBS because of normal CSF results. We assumed that normal CSF finding was due to acute stage and our urgent use of immunoglobulin. That would be vulnerable point of current case. We should have had actively persuade the patient to have another CSF study to make the confirmative diagnosis.

Regarding anamnesis of the patient in current case, we thought severe coughing made a forced neck flexion, which aggravated acute cervical myelopathy. Initial neurological examination and history results seemed like cervical myelopathy. Fortunately, we postponed the surgical decompression owing to absence of severe compression of spinal cord on axial image of MRI, increasing serum CRP, and facial palsy, which led us further examinations to found out the coexisting GBS. If we had performed the surgery, miserable results might have had occurred. In the review of literature, weakness of the muscles of inspiration and expiration (the diaphragm, intercostals, and accessory muscles) may result in inadequate lung expansion and leads to respiratory failure due to absence of adequate cough and secretion clearance and increase of the risk of aspiration and pneumonia[10]. GBS is the most common cause of acute paralysis and neuromuscular ventilatory failure. In spite of development of treatment with plasma exchange and immune globulins, 10% to 30% of patients require mechanical ventilation, 5% to 10% remain seriously disabled, and 3% to 8% die as a result of largely avoidable complications[7].

There is also case report of respiratory insufficiency after anesthesia due to muscle relaxant in patient of GBS[3,9]. In treatment of GBS, ICU support, plasmapheresis, intraveneous immunoglobulin, respiratory and physical treatment are recommended[1,3,8].

We had not paid much attention to upper respiratory illness history, which might have been an important clue for diagnosis of coexisting GBS. This would be our mistake. Considering this case, variable progression and symptoms of GBS may disturb the neurosurgeon in making proper diagnosis and planning surgery, and sometimes unawareness of underlying disease might lead to unintended, miserable results after operation with general anethesia.

Moreover, increase of vaccination such as H1N1 flu will make more frequent chance of meeting the GBS for neurosurgeon, although there is still controversial about influence of vaccination on GBS. We assume that neurosurgeon may need more information of demyelinating disease such as GBS.

CONCLUSION

Authors present a case of coexisting GBS with cervical myelopathy with review of the literature. In such a case, close neurologic examination is very important and essential in decision of surgery.

Authors hope that this case will help the neurosurgeon considering the possibility of underlying demylinating diseases in such clinical setting, especially when neurologic examination results do not fully correspond to cervical myelopathy.

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