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Case Report

Atypical Guillain-Barré Syndrome Misdiagnosed as Lumbar Spinal Stenosis

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Guillain-Barré syndrome (GBS) is an acute inflammatory demyelinating polyneuropathy. In typical cases, the first symptoms of GBS are pain, numbness, paresthesia, weakness in the limbs. Autonomic involvement is common and causes urinary retention and ileus. Much of these symptoms overlap with those of lumbar spinal stenosis. Therefore, correct diagnosis of GBS in a patient with symptomatic lumbar spinal stenosis or in a patient with atypical manifestations of GBS can be difficult, especially early in the course of GBS. Here, we report on a case of atypical GBS in a 74-year-old previously healthy patient with lumbar spinal stenosis and discuss the differential diagnosis of the GBS and lumbar spinal stenosis.

Key Words : Guillain-Barre syndrome · Spinal stenosis · Polyradiculopathy.

INTRODUCTION

Guillain-Barré syndrome (GBS) is an acute inflammatory demyelinating polyneuropathy, an autoimmune disease affecting the peripheral nervous system that is usually triggered by an acute infectious process and is an important cause of acute neuromuscular paralysis[9,13]. In typical cases, the first symptoms of GBS are pain, numbness, paresthesia, and weakness in the limbs[3]. The weakness may initially be proximal, distal, or a combination of both and rapidly progressive[3]. Numbness and paresthesia usually affect the extremities and spread proximally. Decreased tendon reflexes or areflexia is found on examination[3]. Autonomic involvement is common and causes urinary retention and ileus[3]. Much of these symptoms and signs overlap with those of cauda equina syndrome. Lumbar spinal stenosis, which is prevalent in elderly population, is one of the causes of cauda equina syndrome in elderly[12]. In a typical patient with GBS, the diagnosis is usually straightforward[13]. However, in patients with asymmetric weakness, in those with weakness only in the lower extremities, in those with normoreflexia, or in those with symptomatic lumbar spinal stenosis, the diagnosis of GBS as a cause of cauda equina syndrome is challenging.

Here, we report on a case of cauda equina syndrome resulting from GBS in 74-year-old patient with symptomatic lumbar spinal stenosis and discuss the differential diagnosis of these two diseases.

CASE REPORT

A74-year-old previously healthy woman presented to the emergency department with low back pain, bilateral numbness and weakness in the lower extremities. Her symptoms had begun after riding in a car for 4 hours 1 day before and become worse. She had no history of recent infectious disease or vaccination. She had been treated for symptomatic LUMBAR SPINAL STENOSIS with medication and physiotherapy for 1 month. Physical examination revealed paresis of the left lower extremity with strength of 4/5 and of the right lower extremity with strength of 4+/5. Numbness in the lower extremities was not along the dermatomal distribution. Sensory function in the lower extremities was normal. Deep tendon reflexes including biceps jerk, knee jerk and ankle jerk were all hypoactive, and the Babinski sign was negative. Magnetic resonance imaging (MRI) of the lumbar spine revealed spinal stenosis at the levels L2-L3, L3-L4, and L4-L5, and disc protrusion was suspected at the L3-L4 level (Fig. 1). Post-contrast enhancement MRI showed contrast enhancement in the cauda equina (Fig. 2).

Initially she was managed with pain control and regular neu-

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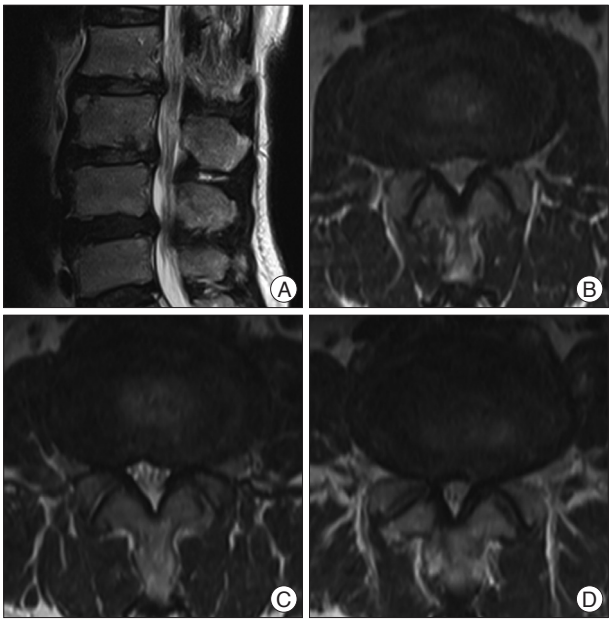

Fig. 1. Magnetic resonance imaging of the lumbar spine. A : Sagittal T2-weighted image shows lumbar spinal stenosis at L2-L3, L3-L4, and L4-L5 level. B : Axial T2-weighted image at L2-L3 level shows central canal stenosis and bilateral neural foraminal stenosis. C : Axial T2-weighted image at L3-L4 level shows central canal stenosis, bilateral neural foraminal stenosis, and disc herniation to the left side of the thecal sac. D : Axial T2-weighted image at L4-L5 level shows central canal stenosis and bilateral neural foraminal stenosis.

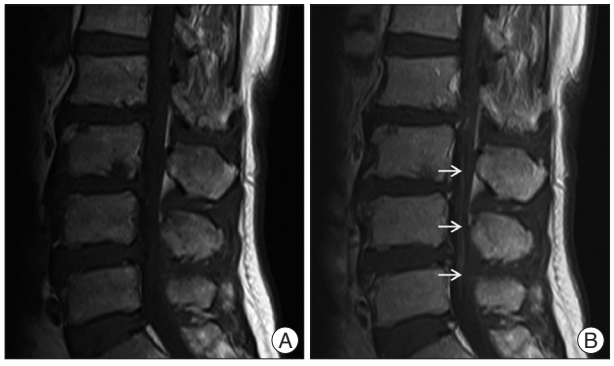

Fig. 2. Pre- and post-contrast enhancement magnetic resonance imaging of the lumbar spine. A : Pre-contrast enhancement sagittal T1-weighted image shows lumbar spinal stenosis at L2-L3, L3-L4, and L4-L5 level. B : Post-contrast enhancement sagittal T1-weighted image shows contrast enhancement in the cauda equina (white arrows).

rological assessment, but her pain and weakness got worse over the next 3 days. She became unable to stand without assistance with strength of 3/5 in the left lower extremity and of the right lower extremity with strength of 4/5. No changes in sensory function and in deep tendon reflex were found on examination. We suspected pain and paraparesis resulting from causes other than the lumbar spinal stenosis because of the rapidly progressive motor weakness. Electromyography (EMG) and nerve conduction study (NCS) 4 days after the onset of weakness of the lower extremities. EMG and NCS showed no findings of peripheral neuropathy. Two days after the initial EMG and NCS, she complaint of voiding difficulty and there was 600 cc of post-voiding residual urine in the bladder. However NCS repeat examination at that time again revealed no evidence of peripheral neuropathy.

Under the impression of cauda equina syndrome due to lumbar spinal stenosis with disc herniation, the patient underwent emergency operation with decompression of the three narrow levels. The dural sac was compressed by hypertrophic facets and a thick yellow ligament. However, her weakness and urinary retention remained unchanged postoperatively, and in addition, bilateral facial palsy, which was more severe in the left side, occurred 5 days after the operation. After referral to the department of neurology for the facial palsy, GBS was taken into consideration as main diagnosis. Electrodiagnostic evaluation, which was performed approximately 2 weeks from the onset of symptoms, presented electrodiagnostic findings suggestive of GBS. Owing to postoperative wound problems at lumbar region, and refusal of the patient to further invasive procedures, lumbar spinal cerebrospinal fluid (CSF) tapping, treatment was initiated solely under impression of GBS by clinical features, clinical course, and results from electrodiagnostic study. She took intravenous immunoglobulin in a regimen of 0.4 g/kg daily for 5 consecutive days.

Three weeks after the onset of symptoms, her weakness began to improve and she could walk with assistance at 5 weeks after the onset of weakness. One year after the onset of symptoms, her pain and weakness were much improved, urination was nearly normal, and no facial palsy was found. She could walk without assistance, but limping and numbness on the lower extremities remained.

DISCUSSION

GBS has remained a descriptive diagnosis of a disorder for which there are no specific diagnostic tests[13]. The combination of rapidly progressive symmetrical weakness in the arms and legs with or without sensory disturbances, hypoflexia or areflexia, in the absence of a CSF cellular reaction, remain the hallmarks for the clinical diagnosis of GBS[13]. In typical cases, the first symptoms are pain, numbness, paresthesia, or weakness in the limbs[3]. Pain was reported in the 2 weeks preceding weakness in 36% of patients, and 66% reported pain in first 3 weeks[10]. The most frequent location of pain was the extremities, less often low back or back[10]. The most frequent pattern of pain was muscle pain, less often painful paresthesia or radicular pain[10]. Numbness and paresthesia usually affect the extremities and spread proximally[3]. The weakness may initially be proximal, distal, or a combination of both, rapidly progressive, bilateral, and relatively symmetric[3,13]. Autonomic involvement is common and causes urinary retention and ileus in GBS patients[3]. However, atypical cases have been reported, in which the patients presented with asymmetric weakness, unilateral fascial palsy, normorelexia, or hyperreflexia[2,7,15].

The diagnosis of GBS itself is usually not difficult for the neurologist, but can be challenging for the physician of first contact who may not have seen a case[3]. In a typical patient with GBS, the diagnosis is usually straightforward[13]. However, in atypical patients, clinical manifestations of GBS can vary and the diagnosis of GBS may be challenging even to the neurologist. A thorough medical assessment may be needed to exclude "mimic disorders"[9,13]. In such cases electrodiagnostic study and CSF analysis can be helpful to confirm the diagnosis.

NCS and EMG play a very important role in diagnosis, subtype classification, and confirmation that the disease is a peripheral neuropathy, but in up to 13% of cases the initial NCS are normal[3,9]. In these cases, retesting in 1 to 2 weeks might be required to confirm the diagnosis[3,9]. In the present case, initial and repeated electrodiagnostic test did not suggest GBS, but the tests were performed too early, that is within 1 week after the onset of symptoms. The third electrodiagnostic test checked about 2 weeks after the onset confirmed GBS.

In addition to NCS and EMG, CSF analysis may confirm a diagnosis of GBS[9]. CSF protein concentrations in patients with GBS are often normal in the first week, but increased in more than 90% of the patients at the end of the second week[13]. Therefore, neither of these tests is highly sensitive early in the course of the disease[8]. We did not examine CSF because we did not consider the possibility of GBS initially. However, even if diagnosis with GBS was considered and subsequent CSF evaluation was performed at the time of arrival at the emergency department or on admission, GBS would not have been accurately diagnosed due to the fact that CSF profiles are usually normal in the early stage of GBS. In the present case, lower extremity weakness was not improved following surgery, and in addition, although asymmetric, bilateral facial palsy developed. After referral to the department of neurology for the facial palsy, GBS was taken into consideration as main diagnosis. Electrodiagnostic evaluation, which was performed approximately 2 weeks from the onset of symptoms, presented electrodiagnostic findings suggestive of GBS. Owing to postoperative wound problems at lumbar region, and refusal of the patient to invasive procedures including lumbar spinal CSF tapping, treatment was initiated solely under impression of GBS by clinical features, clinical course, and results from electrodiagnostic study.

Some authors suggested spinal MRI is a sensitive diagnostic test and should be considered an additional diagnostic tool[8,14]. Spinal MRI with gadolinium frequently indicates enhancement of the spinal nerve roots early in the course of pediatric GBS, and aids clinicians in establishing a diagnosis early in the course of the disease, when other diagnostic tests may still produce normal results[8]. However, in patients with lumbar spinal stenosis, enhancement of compressed nerve roots can be visualized in MRI and this enhancement is thought to reflect either obstructed periradicular veins, indicating venous stasis, intraradicular edema, or breakdown of the blood-nerve barrier, a sign of chronic compressive radiculitis[1,6,11]. In the present case, enhancement of the cauda equina was found in the MRI of the lumbar spine, but we thought it resulted from compressed nerve roots in lumbar spinal stenosis rather than GBS. We could find no report on the differences in enhancement patterns in the MRI between GBS and lumbar spinal stenosis.

The classic clinical presentation of cauda equina syndrome is characterized by low back pain, bilateral sciatica, saddle anesthesia, weakness of lower extremities that may progress to paraplegia, bowel and bladder dysfunction, and impairment of the ankle jerk[12]. These signs and symptoms are typically bilateral but may be asymmetric[12]. Although evaluating a patient with back pain who present with "classic" cauda equina syndrome typically does not present a diagnostic challenge, evaluating a patient with back pain who has early and incomplete signs and symptoms of cauda equina syndrome are difficult problems[12]. Symptoms and signs of GBS and cauda equina syndrome resulting from lumbar spinal stenosis are similar in many ways. Thus, the diagnosis of GBS can be difficult, particularly in patients with pre-existing symptomatic lumbar spinal stenosis who presented with low back pain, asymmetric weakness of the lower extrimities, bilateral numbness or paresthesia in the lower extrimities, and urinary retention as in our patient.

Lumbar spinal stenosis can be a cause of the cauda equina syndrome[12]. But, lumbar spinal stenosis per se is seldom a cause of the cauda equina syndrome and is only found in exceptional cases in spite of the fact that this disease causes a compression of the dura[5,11]. In one study of 163 patients with lumbar spinal stenosis in southern Sweden in the period 1982-1991, an annual incidence of lumbar spinal stenosis of about 50 cases per million inhabitants was found. During this 10-year period, two cases of cauda equina syndrome were diagnosed[4]. In another study, 340 cases of lumbar spinal stenosis were diagnosed in a county in Denmark in the period 1996-2000. During a 5-year period, only one patient with acute cauda equina syndrome in patients with lumbar spinal stenosis was diagnosed[5]. While a disc herniation in a narrow spinal canal may start the cauda equina syndrome, sudden onset of a cauda equina syndrome resulting from a large herniated disc is different from the insidious onset of a cauda equina syndrome from lumbar spinal stenosis[5,12]. In the present case, disc herniation at the L3-L4 level was found in the MRI of the lumbar spine. We thought the pain, progressive paraparesis, and urinary retention resulted from the disc herniation with underlying lumbar spinal stenosis because no other causes were found on the electrodiagnostic study. This led us to urgent decompressive surgery. But in retrospect, the patient presented with rapidly progressive paraparesis and this was more suggestive of GBS rather than lumbar spinal stenosis. Our hasty decision led us to unnecessary surgery.

CONCLUSION

Atypical symptoms and signs of GBS can be confused with

those of lumbar spinal stenosis. No tests are highly sensitive, especially early in the course of the disease. Careful observation of the clinical course and the awareness and suspicion of a possibility of GBS are essential for the proper management and for avoiding unnecessary surgery for lumbar spinal stenosis.

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