Atypical Guillain–Barre Syndrome Caused by Primary Diffuse Large B-cell Lymphoma Originating from Dorsal Root Ganglion

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Guillain–Barre syndrome (GBS) is a common condition in neurological clinical practice. Generally, it is characterized by almost symmetrical flaccid paralysis, extending from the lower limb to upper limb or vice versa. Gloves and socks paresthesia may precede the motor symptoms, but objective sensory loss seldom occurs during the course. Bladder and bowel functions are normal, which is often considered the main discrimination from acute myelitis. Protein-cellular dissociation in the cerebro-spinal fluid is another feature in the diagnosis [1]. Although GBS often takes the form of acute or subacute polyneuropathy and is preceded by viral or ambiguous infection, some types of such neuropathy are actually a manifestation of the remote effect of cancer or are directly associated with non-Hodgkin lymphoma of both T- and B-cells [2]. When making a diagnosis and adopting treatment procedures, a neurologist must perform complete examinations on the central nervous system in case of omitting possible severe conditions.

A male Chinese man at the age of 65 complained of numbness and weakness of the four limbs for about 8 days. His previous history was unremarkable. On neurological examination, he was conscious and mentally normal. His cranial nerves were normal with the exception of the right Horner syndrome. The joint and vibration sensations on both sides were normal, but the pain and temperature sensation on the left limbs were slightly reduced than on the right. The grasping power of the two hands was reduced (grade of 4/5), and he was able to stand up continuously for 5 times. He adopted a steppage gait when walking. The knee and the ankle reflexes on both sides were markedly diminished. The plantar reflexes were flexor. His bladder and bowel functions were normal. According to these symptoms and signs, GBS was considered, but it is obviously not typical because the patient has right Horner sign and the pain–temperature sensation on the left reduced than that on the right. The patient was transferred to the Department of Emergency, waiting for lumbar puncture. Subse-

quent cerebro-spinal fluid showed that the protein was 0.71 g/L and the total number of cells was $2250 \times 10^6/L$, but the number of white blood cells was only 2. Unfortunately, on the second day after the lumbar puncture, his weakness worsened and he suffered from urinary retention, so catheterization was adopted.

At this time, the spinal cord involvement was suspected. About 500 mg of methylprednisolone was intravenously used, and cervical spine MRI was performed immediately. To our surprise, the MRI revealed that the cervical cord was not with myelitis but with a tumor. On MRI, the tumor was epidural at the segment of C7 and the upper cord was swollen. It was homogeneously enhanced after Gadolinium contrast agent was used (Figure 1). Then, the patient was admitted to the Department of Neurosurgery. On operation, it was found that the abnormal tissue originated from the dorsal root ganglion and invaded into the canal through the C7-T1 intervertebral foramen. The tumor was subdural with a clear edge. The corresponding cord was severely compressed. After the tumor was carefully removed and gently scraped from the dorsal root, the tissue was examined pathologically. Immunohistochemical results indicated the diffusion of large B-cell lymphoma (Figure 2). The whole body PET-CT showed normal signs except for the focal neoplasm residue at the site of operation, and the bone marrow biopsy demonstrated only reactive proliferation. All these facts predispose to a primary lymphoma.

To our knowledge, this is a rare case of lymphoma initially presenting with flaccid paralysis [3]. Firstly, this condition is a tumor, and so, theoretically it should experience a chronic course. In fact, the patient only felt ambiguous pain on the right arm one year before and this symptom disappeared soon. The patient was nearly able to walk normally. The motor symptoms worsened quickly only several days before he saw the doctor. Secondly, this condition is undoubtedly a spinal cord compression at the level of C7, and so, theoretically it should have the features of upper motor neuron damage, such as brisk knee tendon reflex and Babinski

Figure 1 There is a tuberos mass in the canal at the level of C7, which traverses out of the right C7-T1 intervertebral foramen, taking the shape of “dumbbell”. On T₁W imaging, the mass signal is identical (**A**, **D**), and on T₂W imaging, the signal is slightly high (**B**). After enhancement, the mass is homogeneous and explicit (**C**, **E**). The cord at the corresponding level is compressed severely and shifted to the left (**E**). Above the mass, the cord is swollen with high signal on T₂W imaging (**B**).

Figure 2 Histopathological examination revealing diffuse, large, and abnormal lymphocytes, with their nuclei highly heterogeneous (**A**, HE staining), and immunohistochemical staining was positive for CD20 (**B**), CD79 (**C**), and Pax-5 (**D**).

sign. However, he did not possess these features when seeing the doctor on the first day. These facts led to the suspicion of GBS and subsequent lumbar puncture.

A question in the differential diagnosis is the tendon reflex. Why did the knee reflexes diminish in such a cervical cord compression? Was the patient on the brink of spinal shock? One explanation may be true, that is the patient did suffer from peripheral neuropathy. The malignant cells in the spinal cord may

directly infiltrate the nerve roots in the canal, or the remote effects of lymphoma had influenced the peripheral nerves.

This case indicates that the clinical presentations of malignant spinal cord compression may be different from those of benign ones. Before lumbar puncture was performed, the whole cord should be imaged by MR. We wondered whether the lumbar puncture precipitated the symptoms, as the paralysis of the limbs worsened and urinary retention happened on the second day. This may

be true because the outflow of CSF can break the pressure balance above and under the compression, which may pull the spinal cord and resultantly damage the neural fibers in the cord. The emergent operation in this case undoubtedly decompressed the cord and facilitated the pathological verification of the diagnosis.

Because the malignant spinal cord compression is a serious and often disabled condition, it is very necessary to emphasize the emergency. In fact, the British National Institute for Health and Care Excellence (NICE) has made detailed procedures for this condition [4,5]. A neurologist or a neurosurgeon should confirm the diagnosis as soon as possible if a patient has definite neurological signs or one of the following symptoms:

- limb weakness;
- difficult in walking;
- sensory loss;
- bladder or bowel dysfunction;

- cervical or thoracic pain;
- pain that is increased by straining and nocturnal spinal pain [6,7].

Once the diagnosis is established, emergent operation and simultaneous methylprednisolone are recommended unless contraindicated, followed by CHOP chemotherapy and radiotherapy.

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Conflict of Interest

The authors declare no conflict of interest.

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