Case 279

2. Neuromyelitis optica spectrum disorder

【Progress】

She was introduced to other major hospital where she was scheduled to get medical services from neurology department.

【Discussion】

Spinal spine composes of three spaces; subarachnoid space, subdural space and epidural space. Unlike brain, subdural space is minimum, no vessels present but fibrous tissue implying that subdural space of the spine is ignorable. As extraspinal tumors, meningioma and neurinoma are listed, while as intraspinal tumors, ependymoma, astrocytoma and hemangiosarcoma are listed. Further, arteriovenous malformation (AVM) and arteriovenous fistula existed. The difference between AVM and AVF is that AVM with nidus is located in spinal cord while AVF is located around spinal cord. Namely, AVM exists in both spinal cord and extraspinal cord while AVF exists in both subarachnoid space and epidural space.

Then, it is crucial to find the arising portion of the lesion: intraspinal, subarachnoid or epidural. Second, what type of lesion is to probe: tumor, inflammation or vessel malformation.

As inflammation of intraspinal cord, it is known that short cord lesion and long cord lesion; short lesion, multiple sclerosis, viral neuromyelitis, sarcoidosis and syphilis: long lesion, neuromyelitis optica spectrum disorder (NMOSD), myelin-oligodendrocyte glycoprotein antibody-associated disease (MOGAD) (1-6). In a bird's eye view, the inflammation of intraspinal cord, viral, bacterial and syphilis infection, and autoimmune disease including sarcoidosis, MS, NMOSD, MOGAD (1-7).

MS is one of immune diseases, a disorder of myelin covering central nerves which play an electric signal rapidly and smoothly. It damages causing brain and spinal cord, inducing short spinal cord lesion (7).

NMOSD is caused by IgG antibody for Aquaporin4 (AQ4) protein which binds to astrocyte (or glia cell). Astrocytes play a role to feed nutrients to neurotic cells and fluid from vessels via Aquaporin 4 pores. Ig G antibody for AQ4 impairs astrocyte, causing various symptoms in brain and spinal cord. As it was once called acute disseminated encephalomyelitis (ADEM), the symptoms vary and lesions spread more extensively than MS. MOGAD is similar as NMOSD but negative antibody for AQ4 but positive for antibody for myelin-oligodendrocyte glycoprotein antibody(1-6). Schwann cell and oligodendrocyte play the common role to support axon in peripheral nerve and central nerve of brain and spinal cord. MOGAD is a disease of disorder of oligodendrocyte. Meanwhile, sarcoidosis is considered one of immune disorder and histologically compose of accumulation of immune cells of macrophages, lymphocytes and plasma cell. Tuberculoma and syphilis are bacterial and spirochete infection, respectively.

Guillan Barre syndrome is also considered to be one of immune diseases. Although its

etiology is still unclarified, Schwann cells of peripheral cells are damaged. Gd-enhanced MRI depict marked enhancement of peripheral nerves and cauda equina nerve roots (8-10).

【Summary】

We presented a forty eight-year-old female presented in our hospital for feeling numbness at bilateral plantar region to bilateral lower extremity. Further, several days it worsened to sluggish and heavy on lower extremity, inducing hard to walk. MRIT2WI depicted a lesion in thoracic spinal cord with high signal intensity beyond three vertebrae, indicating long spinal cord lesion. It is borne in mind that long spinal cord lesions include neuromyelitis optica spectrum disorder (NMOSD), myelin-oligodendrocyte glycoprotein antibody-associated disease (MOGAD) and sarcoidosis. Short cord lesions include multiple sclerosis, sarcoidosis and infectious diseases (viral, bacteria, spirochete). NMOSD is an immune disease by IgG antibody for Aquaporin 4 adhered to Glia cells. MOGAD is the one by antibody for myelin oligodendrocyte glycoprotein. MS arises from a damaged myelin, causing a short spinal cord lesion. Guillan Barre syndrome is a disorder of Schwann cells of peripheral nerves.

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