

Transverse Myelitis: Story of Fifteen Years of Dependence on Mechanical Ventilation

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This is a story of a patient with 27 years old; he had spent 12 years of his life enjoying a good health and living as a lovely child among his family, his father, mother and other brothers and sisters. Wail Hadi Hassan was looking forward to a life filled with happiness and hope and suddenly he found himself inside the hospital, he was admitted to the hospital and since that time he couldn't leave.

At the age of 12 years our patient was completely normal but at 7.0 o'clock a.m. in the 3rd of April 1995, the patient had developed severe cervical and back pain followed after few minutes with weakness in the left hand then right hand followed by vomiting twice then difficulty in walking progressed so rapidly to inability to walk completely.

In the causality when the patient was seen for the first time at 9 o'clock am, the patient was conscious quadriplegic with absent reflexes, bilateral small pupils and bradycardia, but within the following half an hour the patient had developed apnea and cyanosis and so, endotracheal intubation had been done with admission to the R.C.U.

Blood investigations had been done which were normal apart from atypical mononuclear cells and rouleaux formation. CSF was done twice during the first ten days and was normal. Cervical x-ray, myelography and brain C.T. scanning were normal, EMG showed signs of anterior horn cell disease of the upper limb.

Tracheostomy has been done on the 8th of April 1995 to provide tracheal access for ventilation and suction and since that time it has been changed every 2-3 months with meticulous chest physiotherapy and pulmonary toilet with frequent culture and sensitivity tests when there are signs of infections and to prescribe antibiotics if necessary. During the first few months, trials for spontaneous breathing and weaning from mechanical ventilation tried but with no benefit, the tidal volume stayed less 150ml.

At the end of the first week exaggerated reflexes with extensor planter reflex, patellar clonus and ankle clonus has been appeared on the lower limbs while the upper limbs has remained flaccid. The cranial nerves are intact except accessory nerve which shows normal movement of sternomastoid muscle and no movement of upper fibers of trapezius muscle with nystagmus of both eyes to all directions.

All the sensations were absent below C4 including vibration sense and sense of position. The urinary bladder was areflexic, retain urine and so catheterization was introduced but with good training voiding of urine improved and the patient now can pass urine with external catheter.

Stomach tube was introduced on 8th April 1995 but the patient managed after 40 days to swallow semi solid food and after 8 months the stomach tube has been removed and now the patient is on normal feeding. Bowel evacuation occurs every 3-4 days, sometimes helped by suppositories, illius and abdominal distention occurs frequently but usually relieved after passing motion.

Now, he is still with exaggerated reflexes with extensor planter reflexes, patellar clonus and ankle clonus still present on the lower limbs while the upper limbs have remained flaccid. Fundoscopic examination is normal. The patient still dependent on tracheostomy and ventilator.

Spinal cord Dysfunction, Pathophysiology & Non compressive etiology:

Spinal cord dysfunction, or myelopathy, can occur due to a lesion arising within the spinal cord or due to compression of the spinal cord by a lesion originating outside of it. Causes of noncompressive myelopathy from processes within the spinal cord include inflammatory or demyelinating lesions, spinal cord infarction, and vitamin B₁₂ or copper deficiency. Causes of compressive myelopathy include epidural tumors, epidural abscesses, and cervical or thoracic disks. The term transverse myelopathy refers to the clinical presentation of severe motor, sensory, and autonomic dysfunction below a spinal cord level due to any acute or subacute process affecting the spinal cord, whether compressive or noncompressive in etiology⁽¹⁾. The term myelitis is used when a myelopathy is presumed to be due to an inflammatory or demyelinating lesion within the spinal cord.

Clinical suspicion for spinal cord dysfunction should be particularly raised in patients whose clinical history suggests bilateral motor or sensory dysfunction in the extremities in the absence of any signs or symptoms referable to the brain (cognitive, language, or visual symptoms or headache) or brainstem (dizziness, diplopia, nausea, or vomiting). Neck or back pain is a common symptom in patients with epidural spinal cord compression from vertebral metastasis, although many patients with spinal cord disorders do not have associated pain.

Signs of autonomic dysfunction, including bowel, bladder, and sexual abnormalities, also may suggest a spinal cord process but are not always present, particularly early in the disease course. Some patients with cervical spinal cord lesions may have Lhermitte's sign, an electric shock-like sensation down the neck, back, or extremities that occurs with bending of the neck. When evaluating a patient for cervical spinal cord dysfunction, the history should include questions about this symptom. Examination findings that are particularly suggestive of a spinal cord lesion include bilateral weakness and upper motor neuron signs below the lesion, such as hyperreflexia and an extensor plantar response; significant sensory loss below the lesion, such as diminished pinprick sensation below a level in the trunk or abdomen; or severe loss of vibration sense in the legs.

Myelitis, or demyelinating or inflammatory conditions of the spinal cord, typically causes spinal cord symptoms that evolve over hours to days. Demyelination of the spinal cord commonly occurs in the setting of multiple sclerosis. Severe inflammatory demyelination of the spinal cord also is seen in neuromyelitis optica (Devic's disease), which is characterized by recurrent episodes of myelitis and optic neuritis, usually without evidence of the brain lesions typical of multiple sclerosis. In some cases of myelitis, the cause is unclear.

In addition, inflammatory dysfunction of the spinal cord may be associated with infectious or postinfectious processes. Related postinfectious processes include herpes viruses, enteroviruses, and *Mycoplasma*. Some systemic inflammatory diseases, including Sjögren's syndrome, systemic lupus erythematosus, and sarcoidosis, also may cause subacute inflammation of the spinal cord as part of their disease spectrum. High-dose intravenous corticosteroids are typically used as initial treatment for most causes of severe acute or subacute inflammatory or demyelinating disorders of the spinal cord, although supportive evidence from controlled trials is lacking⁽²⁾.

Vitamin B₁₂ deficiency causes subacute combined degeneration of the spinal cord, which develops because of the degeneration of the corticospinal tracts and posterior columns of the spinal cord. This condition is characterized by paresthesias, lower-extremity weakness, and gait instability and may not be associated with hematologic signs of vitamin B₁₂ deficiency. Physical examination findings in this setting may include paraparesis, vibration and position sense loss in the extremities, and sensory ataxia.

Subacute combined degeneration of the spinal cord is preventable or reversible if diagnosed early and treated with parenteral vitamin B₁₂ therapy. Therefore, evaluation for and exclusion of this condition is indicated for all patients with clinical suspicion for spinal cord dysfunction.

Diagnosis of subacute combined degeneration of the spinal cord typically is established by the presence of a decreased vitamin B₁₂ level. However, the presence of elevated levels of methylmalonic acid and homocysteine may be useful for detecting this condition in patients with a low-normal vitamin B₁₂ level.

Recently, copper deficiency also has been shown to cause spinal cord degeneration and a clinical syndrome mimicking vitamin B₁₂ deficiency⁽³⁾. In addition, copper deficiency has been associated with anemia and neutropenia in certain patients. In some patients, malabsorption, which may develop after gastrointestinal surgery among other conditions, causes copper deficiency. In other patients, copper deficiency occurs from excessive zinc ingestion. However, the cause of this condition occasionally remains unknown. Nevertheless, copper supplementation potentially may reverse myelopathy due to copper deficiency.

Infarction of the spinal cord is uncommon and presents as sudden spinal cord dysfunction. This dysfunction typically corresponds to the territory of the anterior spinal artery, causing weakness and pinprick loss below the level of the infarction but relatively sparing vibration and position sense. Spinal cord infarction may occur in the setting of vascular risk factors or aortic disease. However, in a recent study, >50% of the 28 consecutive patients with spinal cord infarction studied had no known cause for this condition⁽⁴⁾. There is no known treatment for spinal cord infarction. Prognosis is variable and primarily depends on the severity of the presenting deficit, but some patients do show clinical improvement.

Another rare cause of ischemic spinal cord dysfunction is dural arteriovenous malformations of the spinal cord. These malformations obstruct the venous outflow of the spinal cord, causing spinal cord dysfunction that may progress over months to years⁽⁵⁾. Surgical obliteration of the fistula may potentially reverse this condition.

References:

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