Case 014: A man with loss of sensation in his fingers and weakness in his legs.

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A 53 year-old man presented to the clinic with complaints of weakness in all 4 limbs over the past month. He was on regular drug treatment for hypertension, gouty arthritis and chronic obstructive airway disease. He had surgery for carcinoma of esophagus 2 years ago with good recovery. On direct questioning he recalled that his trouble had begun more than 6 months ago as increasing clumsiness with his hands. He also noticed having lost pain sensation in his fingers – so much so that there was an episode of burn injury to the index and middle fingers of his right hand while holding a cigarette.

Physical examination revealed the following abnormalities bilaterally.

Upper extremities:

- Muscle wasting in the hands and forearms with some weakness in those muscle groups.
- Absent biceps and triceps tendon reflex.
- Loss of pain and temperature senses in C5 dermatome and below.
- Touch, vibration, and proprioception senses were unaffected.

Lower extremities:

- Minimal weakness.
- Slight spasticity.
- Brisk knee and ankle jerks with early clonus.
- The plantar reflex was extensor bilaterally (positive Babinski sign).
- There was no sensory deficit.
Cranial nerve examination and examination of coordination and gait were normal.

1. What is the differential diagnosis?

Normal cranial nerves and absence of cerebellar signs suggest that the lesion is at the spinal cord level. The differential diagnosis should include myelitis, intra- and extra-medullary spinal cord tumors, spinal cord trauma, syringomyelia, hemorrhagic or thrombotic lesions in the spinal cord, and progressive spinal muscular atrophy.

Magnetic resonance imaging (MRI) of his cervical and thoracic spinal cord was ordered and the diagram is a mid-sagittal image.

2. What is the diagnosis?

The MRI image shows a fluid-filled defect (syrinx) along the central axis of the spinal cord extending from the lower cervical to the upper thoracic segments. (Normal spinal cord appears dark. The syrinx is the white area between the red arrows.) This is consistent with a diagnosis of syringomyelia. There is no sign of cerebellum herniation at the foramen magnum, thus excluding Chiari malformation.

3. What is syringomyelia?

Syringomyelia is a condition in which a cerebral spinal fluid-filled cavity, called a syrinx, is formed in the center of the spinal cord causing dissociated sensory loss and lower-motor-neuron type weakness at the level of the involved segments together with upper motor neuron signs below the involved segments. Onset is
insidious and cervical segments are most commonly affected. In the early stages of the disease neurological deficit may be only unilateral or bilateral but asymmetric.

4. What is the etiology of syringomyelia?

It is believed that disturbances in the flow dynamics of cerebral spinal fluid (CSF) increase CSF pressure in the central canal of the spinal cord to cause the formation of syrinx. Syrinx may be found in patients who have altered CSF flow dynamics caused by narrowing of the cranio-cervical junction; it may form following spinal cord trauma; it may be associated with cord tumors. Syrinx formation is also associated with type 1 Chiari malformation, a congenital disorder in which the cerebellar tonsils are displaced downward below the foramen magnum. Many a time syrinx formation may be without a known etiological cause.

5. What is the anatomical basis for the symptoms and signs of syringomyelia?

Syringomyelia typically affects the lower cervical and the upper thoracic segments of the spinal cord. The most characteristic and early sign is dissociated sensory loss: loss of pain and temperature sensation with preservation of fine touch, two point discrimination, vibration, and proprioception. This pattern of sensory loss is “suspended” in the sense that dissociated sensory loss occurs only in segments affected by the syrinx; sensations of all modalities are normal above and below the level of the syrinx.

Sensory dissociation arises from the different courses taken by sensory fibers after entering the spinal cord. Second order neurons sub-serving pain and temperature decussate (cross the midline) in the ventral white commissure close to the level of cord entry. After descussation these fibers ascend in the spinothalamic tract. On the other hand neurons sub-serving fine touch, two point
discrimination, vibration, and proprioception do not cross the midline and ascends in the ipsilateral dorsal column (see diagram).

Diagram illustrating the effect of an enlarging syrinx (central grey area) on adjacent neurons.

With a syrinx (central grey area in the diagram) expanding outward from the center of the cord, it is the decussating fibers of pain and temperature that are first compromised. As the fusiform syrinx expands centrifugally, the motor neurons in the anterior horns are involved early, resulting in lower-motor-neuron-type weakness, wasting, and loss of tendon reflex in the upper extremities at the levels of the lesion. As the syrinx expands further outward, the descending lateral corticospinal (pyramidal) tract responsible for motor function is also compromised, producing an upper-motor-neuron-type weakness together with spasticity, exaggerated tendon reflex, clonus, and positive Babinski sign in the lower extremities below the levels of the lesion.
6. What is the treatment and prognosis of syringomyelia?

- Surgical correction of the underlying abnormality if one can be identified. Although corrective surgery may stop the progression of the disease, it does not necessarily reverse the neurological deficit.
- Drainage and decompression may still be attempted in the absence of an etiological cause.
- Prognosis is variable, ranging from deaths within a year of diagnosis to progressive incapacitation over several years. Spontaneous arrest of further deterioration has also been reported.

Further Reading