Syringomyelia

Syringomyelia is a cystic cavity in the central canal of the spinal cord that often develops in association with Chiari malformations or after trauma to the spine. Clinical features are characterized by a cape-like distribution, meaning that the symptoms are felt predominantly in the arms, neck, shoulders, and upper back. They include bilateral loss of pain and temperature sensations as well as bilateral muscle weakness that starts distally in the upper extremities. Horner syndrome may develop if descending autonomic fibers are affected in the T1-4 region. Diagnosis often requires an MRI while management includes supportive care like physical therapy, muscle relaxants, and pain control. Surgery may be indicated in progressive or advanced cases.

PLAY PICMONIC

Characteristics

Cystic Cavity in Central Canal

Sisters in Cave

Syringomyelia is characterized by a fluid-filled cyst/cyst cavity (called a syrinx) that forms in the central canal of the spinal cord. It results from an abnormal circulation of cerebrospinal fluid (CSF). A syrinx usually gets larger over time, compressing nerve fibers around it.

Chiari Malformations

Cherry-malformed

Syringomyelias are often seen concomittantly with Chiari malformations. In a Chiari malformation, a small posterior fossa can cause part of the cerebellum to descend below the level of the foramen magnum. This obstruction can cause CSF back-pressure resulting in a syrinx.

Trauma

Trauma-spike

Post-traumatic syringomyelia commonly occurs within 5 years of the initial spinal cord injury (SCI). The exact pathogenesis is unclear, but is believed to be due to dural adhesion and scarring that forms a one-way valve phenomenon, thus blocking CSF flow. Another theory is the "Venturi effect", which states that a more rapid CSF flow in a stenotic region would pull the spinal cord laterally, resulting in a cystic cavity to develop centrally.

Clinical Features

Cape-like Symptoms

Cape

Expansion of the central canal disrupts the decussation of both lateral and anterior spinothalamic tracts. The most common area affected by syringomyelia is the cervical cord. Compression in this area causes bilateral loss of anterior spinothalamic tract functions resulting in a "cape-like" distribution of symptoms.

Loss of Pain Sensation

No-sign on Pain-bolts

The anterior expansion of a syrinx may compress the anterior white commissure, which is the decussation for fibers of the spinothalamic tract. This tract carries pain and temperature. Thus, bilateral loss of pain and temperature may be evident on physical exam. Radicular pain may still be felt, but external nociception (e.g. pinprick) will be dulled.

Loss of Temperature

No-sign on Thermometer

The anterior expansion of a syrinx may compress the anterior white commissure, which is the decussation for fibers of the spinothalamic tract. This tract carries pain and temperature. Thus, bilateral loss of pain and temperature may be evident on physical exam. Radicular pain may still be felt, but external nociception (e.g. pinprick) will be dulled.

Muscle Weakness

Weak Drooping-muscle

Extension of the syrinx anteriorly can cause compression and dysfunction of the corticospinal tract neurons. This can result in muscle weakness, atrophy, or even paralysis that usually starts distally and progresses proximally given the arrangement of fibers in the spinal cord. The dorsal (posterior) column is initially spared since the cystic cavity tends to expand anteriorly and laterally. However, in advanced cases, patients may being to lose dorsal column functions like proprioception and vibration sense beginning in the lower extremities.

Horner Syndrome

Horny-PAM

If the syrinx is around the upper thoracic region (T1-4), the hypothalamospinal tract, which carries sympathetic fibers to the face, may be affected. This interruption will result in Horner syndrome (a.k.a. oculosympathetic paresis), classically yielding unilateral miosis, ptosis, and facial anhidrosis.

Diagnosis

MRI

M-R-eyes Machine

MRI of spine and spinal cord with and without contrast allows visualization of syrinx in both sagittal and axial planes. It can show cystic lesions, their location, size, and extent. It can also show the degree of cerebellar tonsillar ectopia, arachnoid scarring, spinal tumors, and leptomeningeal enhancement (sign of infection). Repeated MRI scans are used to monitor the progression.

Management

Supportive Care

Supportive IV bags

Most syringomyelia patients will first undergo supportive care, which includes analgesics, physical therapy, and education to avoid activities that increase intracranial pressure (ICP) e.g. lifting heavy objects, jumping.

Surgery

Surgeon

If signs and symptoms of syringomyelia worsen or there is a surgical cause of syrinx development (e.g. tumor), surgery is recommended to reduce the pressure of the syrinx and to return the CSF circulation back to normal. The type of surgery depends on the underlying pathology. Some patients may only need decompression via CSF shunting while others may need syringotomy, laminectomy, or tumor resection.