Syringomyelia — Anatomy and Lesions of the Spinal Cord

Syringomyelia is characterized by progressive abnormal cerebrospinal fluid (CSF) accumulation in the spinal cord. It causes dissociated suspended anaesthesia that affects pain and temperature sensation, predominantly in the upper limbs. Etiology is unclear, although a number of theories have been proposed. The accepted consensus, however, is that syringomyelia is a manifestation of an underlying pathology rather than a disease itself. Treatment of the primary cause often leads to regression of the syrinx.

Definition of Syringomyelia

The term “syringomyelia” was introduced by Ollivier d’Angers in 1827 to describe *cystic cavitations of the spinal cord*. However, not every cystic formation of the spinal cord should be described as a syrinx. Syringomyelia is a *progressive accumulation of fluid inside the spinal cord*. This fluid may be localized inside the parenchyma or the central canal.

Other types of cystic formation of the spinal cord include:

- **Hydromyelia**: syrinx that is confined to the central canal
- **Holo-syrinx**: fluid-filled cavity extending through the entire spinal cord
Syringobulbia: the presence of syrinx in the brainstem

Epidemiology and Pathogenesis of Syringomyelia

The exact incidence of syringomyelia is unknown as many patients are asymptomatic.

As of now, no single unifying pathophysiological theory unequivocally explains syringomyelia. The following few facts are, however, universally accepted:

- Almost any pathology in the spinal canal or the craniocervical junction can potentially lead to the syrinx.
- The syrinx is usually a response of the spinal cord to 3 common abnormal phenomena: disturbance of cerebrospinal fluid (CSF) flow, spinal cord tethering or an intramedullary tumor.

Among the many theories proposed to explain syringomyelia, few prominent ones are as follows:

<table>
<thead>
<tr>
<th>Theory</th>
<th>Mechanism of syrinx formation</th>
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</thead>
<tbody>
<tr>
<td>Chiari et al.</td>
<td>Persistence of embryological state</td>
</tr>
<tr>
<td>Gardner et al.</td>
<td>Hydrodynamic theory-arterial pulsations lead to CSF accumulation</td>
</tr>
<tr>
<td>Williams et al.</td>
<td>Valsalva maneuvers lead to abnormal CSF accumulation</td>
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<tr>
<td>Oldfield et al.</td>
<td>Systolic pressure changes lead to CSF accumulation.</td>
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<tr>
<td>Milhorat et al.</td>
<td>Production of CSF in segments</td>
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<tr>
<td>Ball and Dyan</td>
<td>Dissection of fluid through Virchow-Robin spaces</td>
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<tr>
<td>Aboulker et al.</td>
<td>Fluid entry along nerve roots</td>
</tr>
</tbody>
</table>

Classification of Syringomyelia

Syringomyelia is a pathologic response of the spinal cord. It can be primary.
(congenital) or secondary (acquired). The most common example of primary congenital idiopathic syringomyelia is **Arnold-Chiari malformation (ACM)**. There are 4 types of ACM:

- **Type I**: Syringomyelia with tonsillar herniation greater than 5 mm
- **Type II**: Syringomyelia with neural tube defects with hydrocephalus
- **Type III**: Occipital encephalocele
- **Type IV**: Cerebellar hypoplasia

Acquired syringomyelia is secondary to a known primary cause. The different causes of secondary syringomyelia include:

- Post-traumatic (most common)
- Post-infective (post-tubercular, post-arachnoiditis)
- Secondary to a tumor in the spinal cord or, sometimes, due to intracranial tumor
- Spinal cord tethering and scarring
- Irritation by contrast agents such as metrizamide, which is used in myelography
- **Degenerative disc disease** (rare)
- After any spinal intervention (secondary to fibrosis and tethering).
- After any spinal intervention (secondary to fibrosis and tethering).

**Symptoms of Syringomyelia**

As noted, many patients are asymptomatic or minimally symptomatic. For those patients who are symptomatic, the most common symptoms include:
- **Dissociated anesthesia** due to the involvement of crossing *spinothalamic fibers* in the anterior white commissure. Pain and temperature sensations are lost, while posterior column sensations are largely preserved. Dysesthesias, paraesthesias, pain, tingling, and numbness are the most common presenting features.

- **Suspended anesthesia** or preferential involvement of upper limbs in a capelike distribution due to medially located cervical fibers.

- Early involvement of bowel/bladder.

- Local destruction of anterior horn cells at the site of the syrinx leading to a lower motor neuron-type of weakness in the involved segments. The symptom is characterized by wasting, weakness, and diminished reflexes.

- Disruption of the local microvascular system leading to *ischemia* at the watershed levels of the spinal cord. This occurs in the C8-T1 region, with borderline blood supply, and it commonly manifests as gross wasting of the small muscles of the hand.

- Further expansion of the syrinx causing the involvement of corticospinal tracts, descending sympathetics, and, lastly, the posterior column.

- Patchy dysesthesias because of asymmetric expansion of the syrinx.

- **Charcot joints (neuropathic arthropathy)** in the shoulder joints due to loss of sensory fibers to the joint.

- **Scoliosis**.

- Abnormal abdominal reflexes in children.

The table below outlines the major tracts involved and their manifestations:

<table>
<thead>
<tr>
<th>Tract</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior spinothalamic tract decussation in the gray commissure</td>
<td>Loss of pain and temperature sensation in a predominant capelike distribution (involving bilateral upper limb and the neck).</td>
</tr>
<tr>
<td>Spinobulbospinal and reticulospinal pathways</td>
<td>Early involvement of bowel and bladder-upper motor neuron type (urgency, frequency, and nocturia)</td>
</tr>
<tr>
<td>Late involvement of corticospinal tract</td>
<td>Late motor weakness</td>
</tr>
<tr>
<td>Local anterior horn cells at the site of the syrinx</td>
<td>Local lower motor neuron type of weakness</td>
</tr>
<tr>
<td>Sparing of posterior columns</td>
<td>Sparing of vibration sense, position sense, and proprioception.</td>
</tr>
<tr>
<td>Ischemia of the watershed zone at C8-T1</td>
<td>Gross wasting of small muscles of the hand</td>
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</table>

**Progression**

If untreated, syringomyelia can spread rapidly. Long-term prognosis is often a culmination of the natural course of the primary pathology.

**Diagnosis and Differential Diagnosis of Syringomyelia**

The first symptoms of syringomyelia are related to the primary underlying local pathology. Hence, a detailed, accurate history can often point to the specific investigations needed to reach a diagnosis.

**Imaging studies** are often the first investigations performed. **Magnetic resonance**
Imaging of the spine (plain with contrast) is the most common and useful initial investigation. It helps to rule out cystic lesions of the spinal cord and any associated spinal tumors. Lepto-meningeal enhancement can pinpoint infection. Arachnoid scarring can sometimes be seen.

Cardiac gated Cine-MRI flow study is the most sensitive tool currently available to document CSF flow disturbances, as well as CSF flow around foramen magnum and around potential obstructions. It is also a key way to compare CSF flow changes post-operatively and to document objective improvement in the same.

Differential diagnosis

- Intramedullary tumor
- Intramedullary cysts
- Arachnoid cysts
- Myelomalacia
- Glioependymal cysts
- Dilatations of the central canal

Therapy of Syringomyelia

The fact that each syrinx is a manifestation of underlying pathology has significant repercussions for treatment. Treatment of the primary disease is the gold standard. The presence of syrinx should lead to a detailed and meticulous work-up to find the primary pathology. Not every syrinx requires treatment, however. Reversal of the inciting factor leads to regression of the syrinx.

The first causes to look for are intramedullary spinal tumors and spinal dysraphisms leading to tethered cord. Next in line are the causes of disturbed CSF flow, the most common being cranio-cervical junction disorders, Arnold-Chiari malformations, and arachnoid scarring. Treatment is then tailored to the causal factor.

The role of surgery can be summarized in the following manner:
- In patients with intramedullary tumors, the tumor should be removed.

- In patients with an ACM type I malformation, decompression of the foramen magnum is required.

- In patients with arachnoid scarring related to trauma, subarachnoid hemorrhages, infections, or other causes, the arachnoid scar should be resected and CSF flow established.

- In patients with spinal stenosis, the spinal canal must be decompressed.

- In patients with a tethered cord, the tethering must be released.

- In some patients, **progressive syringomyelia** or **re-tethering secondary to multiple spinal surgeries** requires direct treatment of the syrinx. A **theco-peritoneal shunt** is valuable in these patients. Caution should be exercised, as “once a shunt, always a shunt.” As well, shunt surgery has a **high rate of complications**. Programmable shunts may improve these rates in the future.

The benefits of other shunt surgeries such **syringo-peritoneal** and **syringo-subarachnoid shunts** do not clearly outweigh the risks and failures are not uncommon. Although a number of materials have been used to shunt fluid, the search for the best shunt type, material, and valve is ongoing.

**Complications of Syringomyelia**

The progressive nature of syringomyelia necessitates treatment. The complication rate for surgery is about 21%. The most common complication is **CSF fistula formation**. Other potential complications include:

- Hemorrhage
- Arachnoid scarring
- Re-tethering
- Infection
- Re-accumulation of syrinx
- Spinal cord contusion/edema/injury
- Nerve root injury/damage
- Shunt-related complications
- Added postoperative neurological deficits due to the treatment of the primary pathology

Summary

Syringomyelia is not a disease in its own right, but a manifestation of underlying pathology. Symptoms include patchy, dissociated suspended anesthesia; and early bowel/bladder involvement with late motor affection and relative preservation of posterior column sensations. Targeted treatment of the primary cause can lead to regression of the syrinx.

References


DeJong’s Textbook of Neurology

Youman’s Textbook of Neurological Surgery

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