Syringomyelia — Anatomy and Lesions of the Spinal Cord

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Syringomyelia is characterised by progressive abnormal CSF accumulation in the spinal cord. It causes dissociated suspended anaesthesia affecting pain and temperature sensation predominantly in the upper limbs. Etiology is unclear. There are many theories proposed but the accepted fact is that it is a manifestation of an underlying pathology rather than being a disease itself. Treatment of the primary cause often leads to regression of the syrinx.

Definition of Syringomyelia

Introduced by Ollivier D’Angers in 1827, the term “syringomyelia” epitomizes cystic cavitations of the spinal cord. However, not every cystic formation of the spinal cord should be addressed as a syrinx. Syringomyelia describes a progressive accumulation of fluid inside the spinal cord. This fluid may be localized inside the parenchyma or the central canal. Few other seemingly synonymous distinct terms need clarification:

- **Hydromyelia** - a syrinx confined to the central canal
- **Holo-syrinx** - fluid filled cavity extending through the entire spinal cord
- **Syringobulbia** - signifies 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 patho-physiological theory unequivocally explains syringomyelia. The following few facts are, however, universally accepted:

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

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

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<tr>
<th>Theory</th>
<th>Mechanism of syrinx formation</th>
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</thead>
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<tr>
<td>Chiari et al.</td>
<td>Persistence of embryological state</td>
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<tr>
<td>Gardner et al.</td>
<td>Hydrodynamic theory-arterial pulsations lead to CSF accumulation</td>
</tr>
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<td>Williams et al.</td>
<td>Valsalva maneuvers lead to abnormal CSF accumulation</td>
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<tr>
<td>Oldfied 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>
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</tbody>
</table>

Classification of Syringomyelia

Syringomyelia is a pathologic response of the spinal cord. It can be primary/congenital and secondary/acquired. The most prevalent example of primary congenital idiopathic syringomyelia is Arnold Chiari Malformation (ACM). There are 4 types of ACM:
ACM Type I: Syringomyelia with tonsillar herniation more than 5 mm

ACM Type II: Syringomyelia with neural tube defects with hydrocephalus

ACM Type III: Occipital encephalocele

ACM Type IV: Cerebellar hypoplasia.

Secondary/acquired syringomyelia is secondary to a known primary cause. Thus, the different causes of secondary syringomyelia are:

- Post-traumatic (commonest)
- Post-infective (post-tubercular, post-arachnoiditis)
- Secondary to a tumor in the spinal cord or less likely due to intracranial tumor
- Spinal cord tethering and scarring
- Irritation by old contrast agents like metrizamide used in myelography
- Degenerative disc disease (rare)
- After any spinal intervention (secondary to fibrosis and tethering).

Symptoms of Syringomyelia

It is rather a disillusion that many patients are asymptomatic or minimally symptomatic. The exact cause of the same is unknown.

For the symptomatic ones, the main features of syringomyelia are:

- Dissociated anesthesia due to 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 sensation are the most common presenting features.
- **Suspended anesthesia** is preferential involvement of upper limbs in a cape like distribution due to medially located cervical fibers.
- Early involvement of bowel/bladder.
- Local destruction of anterior horn cells at the site of the syrinx leads to lower motor neuron type of weakness in the involved segments. It is characterized by presence of wasting, weakness, and diminished reflexes.
- Syrinx leads to disruption of the local microvascular system leading to **ischemia** at the watershed levels of the spinal cord. This is the C8-T1 region with borderline blood supply and it commonly manifests as gross wasting of small muscles of the hand.
- Further expansion of the syrinx can cause involvement of corticospinal tracts, descending sympathetics, and lastly the posterior column.
- Patchy dysesthesias because of asymmetric expansion of the syrinx is classically described.
- **Charcot joints (neuropathic arthropathy)** can occur in the shoulder joints due to loss of sensory fibers to the joint.
- Syringomyelia is associated with **scoliosis**.
- Abnormal abdominal reflexes are seen in children with syringomyelia.

The major tracts involved and the following consequent manifestations are tabulated below for easy memorisation and recall:

<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 predominant cape like 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 hand</td>
</tr>
</tbody>
</table>

**Progression**

If untreated, syringomyelia can relentlessly expand. The 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 most specific investigations needed to reach the diagnosis.

**Imaging studies** are often the preliminary investigations performed.

**MRI of the spine** (plain with contrast) is the most common and useful initial investigation. It helps in ruling out **cystic lesions** of the spinal cord and any associated
spinal tumors. Lepto-meningeal enhancement can signify infection. Arachnoid scarring can sometimes be appreciated.

Cardiac gated Cine-MRI flow study is the talk of the day. It is the most sensitive tool available to document CSF flow disturbances, CSF flow around foramen magnum and around potential obstructions. It is also instrumental in comparing 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 fundamental fact that each syrinx is a manifestation of underlying pathology has significant repercussions. Treatment of the primary disease is the gold standard treatment for the syrinx. Presence of syrinx should lead to a detailed and meticulous work-up to find the primary pathology. Not every syrinx requires treatment. It is rather a sign, not a disease by itself. 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. The next in line are causes for perturbed CSF flow, the commonest being cranio-cervical junction disorders, Chiari malformations, and arachnoid scarring. The treatment is then tailored as per the cause.

The role of surgery can be summarized in the following manner:

- In patients with intramedullary tumors, the tumor should be removed.
- In patients with a Chiari I malformation, a 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 has to be decompressed.
- In patients with a tethered cord, the tethering has to be released.
- In few patients, **progressive syringomyelia** or **re-tethering secondary to multiple spinal surgeries** demand direct treatment of the syrinx. The **co-peritoneal shunt** is valuable in these patients. Deliberate caution is exercised in these patients as “**Once a shunt, always a shunt.**” The shunt surgery is notorious for its **high complication rate**. Programmable shunts might alleviate this problem in the future.

Other shunt surgeries like **syringo-peritoneal shunt** and **syringo-subarachnoid shunt** stay in the gray zone. The benefits do not clearly outweigh the risks and failures are involved. Almost every material has been used to shunt fluid but none outshines the other in a conclusive manner. The search for the best shunt type, material and valve still continues.

**Complications of Syringomyelia**

The progressive nature of the disease necessitates treatment. The complication rate for surgery is about 21%.

The most common complication is **CSF fistula formation**. Other potential complications are:

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

Summary

Syringomyelia is not a disease in its own right, but rather a manifestation of something more sinister. Symptoms are patchy dissociated suspended anesthesia and early bowel/bladder involvement with late motor affection and relative preservation of posterior column sensations. The inciting event needs treatment and the right treatment of the same can potentially lead to regression of the syrinx.

References


DeJong’s Textbook of Neurology

Youman’s Textbook of Neurological Surgery

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