Syringomyelia

Synonyms: syringomyelia, hydromyelia, syringohydromyelia, myelosyringosis, Morvan's disease

Epidemiology

Pathology

Etiology

Pathogenesis

Clinical features

Differential diagnosis

Treatment

Posttraumatic syrinx → see p. T255

Epidemiology

- exact incidence unknown but it is rare.
- more frequent in men.

Pathology

- lateral (tabular) cavitation in central spinal cord.
- syrinx is lined by dense, glistening tissue and surrounded by dense glial fibrol wall.
- syrinx may communicate with central canal (fluid within syrinx is similar to, if not identical with CSF).
- most common starting location - base of posterior horn in cervical cord.
- transverse diameter varies from segment to segment (usually maximal in cervical and lumbosacral enlargements).
- syrinx slowly enlarges (due to hydrodynamic forces) to involve much of both gray and white matter (at times, only narrow rim of parenchyma is left).
- if syrinx is long, it may have septations but usually (but not always) all cavities communicate.

Etiology

1. Congenital
- familial cases have been described.
- rarely occurs in isolation; usually associated with other anomalies:
  1. Arnold-Chiari malformation (60-90%)
  2. platybasia
  3. atresia of Magendie foramen
  4. Dandy-Walker syndrome
  5. vascular malformations.

2. Syringomyelia may also be late consequence of spinal trauma.
   - ex vacuo after absorption of intramedullary hematoma
   - flexion-extension injury
   - cord ischemia due to hypotension
   - birth trauma

3. Tumors - syrinx may cap rostral or caudal pole of intramedullary (rarely, extramedullary) tumor (e.g. low grade astrocytomas).


Pathogenesis

Communicating* syringomyelia (s. hydromyelia) – most cases of syringomyelia!

A) hydrodynamic Gardner's theory: occluded exit foramina of fourth ventricle (e.g. by developmental defect in rhombic roof) + CSF pulsations directed downward → dilatation of central canal ± hydrocephalus.

B) Williams theory: bulk-swell effect of cerebellar tonsils in Arnold-Chiari malformation (central spinal constriction at level of foramen magnum) → during Valsalva maneuver CSF may pass caudad through narrowed canal → syrinx formation; foramen magnum decompression usually results in syrinx resolution.

Noncommunicating syringomyelia

A) extension of CSF under pressure along Virchow-Robin spaces
B) cystic degeneration of intramedullary tumor
C) ischemia in anterior spinal artery insufficiency
D) resorption of intramedullary hematoma (hematomyelia)
E) cord contusion / compression → microcystic cavitation.
F) obliterated original communication with ventricular system.

Clinical features

Manifests in 3-5th decade (mean age at onset ≈ 30 years) - chronic, slowly progressive central cord syndrome
- usually located in cervical or upper thoracic segments (rarely, syrinx develops in lumbar cord either in association with or independent of cervical syrinx).

Damage to different spinal cord myelins signs (anesthesia with thalamoanesthesia):
- bilateral, frequently asymmetrical.
- over shoulders (“cape” distribution) or across shoulders and upper torso, front and back in shawl-like distribution (en cuirasse).
- deep and aching chronic pain (≈ 30%) in impaired segments.
- complications - painless hail ulcers, burns, and whitlows.

Damage to ANS deferens: segmental LMN signs (areflexia, weakness, atrophy, fasciculations).
**SYRINGOMYELIA**

- Intrinsic hand muscles are often affected first → striking early hand atrophy - claw-hand deformity (main en griffe).
- Ascends to forearms, and ultimately affects shoulder girdle.
- Complications:
  1. Thoracic scoliosis (thoracic LMN innervating paraspinal musculature).
  2. Neurogenic arthropathies (Charcot joints) in shoulder, elbow, wrist.

Classic vignette: cervical syrinx → Charcot shoulder

As syrinx size increases, other spinal structures can be involved. See p. Spin1 ->

1. Dorsal columns
2. Corticospinal pathways → spastic leg weakness.
3. Intermediolateral columns: e.g. hands may develop remarkable subcutaneous edema and hyperhidrosis (main succulente).

Spontaneous arrests for several years are not uncommon.

Violent cough or sneeze may produce hemorrhage into syrinx.

**SYRINGOBULBIA**

Syrinx may expand into brainstem - into medulla oblongata or even into pons:

- CN9-12 may be involved - harbar palsy, usually asymmetrically.
- Nuclear involvement of CN5 → facial pain and thermal hypesthesia in onion-skin pattern.
- Nystagmus is not rare.
- Rarely, syrinx extends into centrum semiovale (syringocephalus).

DIAGNOSIS

**CSF** - few abnormalities (CSF pressure may be elevated, cell count rarely > 10/mm³, mild elevation of CSF protein).

**MRI** - diagnostic procedure of choice! (syringomyelia, and its cause, are shown well)

- Signal intensity of cyst = CSF.
- Cyst margins often irregular (may demonstrate periodic folds or septations - result from turbulent flow within cavity).
- Further MRI evaluation:
  1. Brain hydrocephalus?
  2. Craniovertebral junction (Arnold-Chiari malformation?).

N.B. If syringomyelia occurs without Arnold-Chiari malformation or prior spinal cord injury → complete spinal MRI with gadolinium (to rule out intramedullary tumor).

**MRI-T1**: large syrinx + associated Arnold-Chiari malformation (cerebellar tonsillar herniation below foramen magnum).

**MRI-T2**: syringobulbia due to cervicothoracic pilocytic astrocytoma in 4 yo female.

**MRI**: dilated central spinal canal + pointed cerebellar tonsils (Chiari I malformation).
Syringomyelia: MRI (syringomyelia + Chiari malformation):
A. MRI-T1: descent of cerebellar tonsils and vermis below level of foramen magnum (black arrows); CSF collection dilates central canal (white arrows).
B. MRI-T2: high-signal-intensity syringomyelia (white arrows) expanding cervical cord with signal intensity equivalent to CSF.

T1-MRI (syringohydromyelia + Chiari I malformation): cerebellar tonsils (upper thick black arrow) lie below neural arch of C1, and distended syrinx (white and black arrows) extends from C2 to T2.

Myelography: followed by delayed CT.
- Spinal cord is:
  - enlarged (fusiform expansion) in 80%;
  - normal in 10%;
  - small (flattened in sagittal plane) in 10%.
- Rotating patient from prone to supine position → considerable changes in cord size and shape.
- Contrast medium enters all intramedullary cavities (most likely 6–12 h after myelography).

Patient prone (A) and supine (B) - fluctuation in cyst size (low radiodensity) with posture.
(C) patient supine 8 h after myelogram - contrast penetration and retention in syrinx cavity (arrow).
DIFFERENTIAL DIAGNOSIS
- all easily excluded with MRI:
  1) intramedullary tumors.
  2) ALS
  3) MS
  4) cervical spondylisis
  5) anomalies of craniovertebral junction

TREATMENT
Incidental syrinx (i.e. asymptomatic and no neurologic deficit) with no identified etiology → observation with MRI at 2-3 year intervals (if the size remains stable).

Symptomatic / enlarging syrinx → surgery.

A. First choice - correction of abnormal CSF dynamics:
   • tumor: mass removal → syrinx resolution.
   • Arnold-Chiari malformation:
     a) hydrocephalus is present → ventriculoperitoneal shunting.
     b) no hydrocephalus (or ineffective ventriculoperitoneal shunt) → posterior fossa decompression, ± simultaneous syrinx shunting.

B. Second choice - drainage of syrinx cavity when etiology cannot be easily addressed (e.g. in arachnoiditis, post-traumatic syrinx): see p. Op230:*
   a) temporary – percutaneous needle aspiration (can be repeated) or open syringotomy.
   b) permanent – various shunts (syringopleural, syringoperitoneal, syringo-subarachnoid).

BIBLIOGRAPHY for ch. “Spinal Disorders” → follow this LINK >>