Definition

Intramedullary accumulation of cerebrospinal fluid resulting from dilation of the central canal (hydromyelia) or paracentral cavitation (syringomyelia); these conditions are often simultaneously present and indistinguishable. **Syringobulbia**: Spread into the medulla oblongata. **Communicating syringomyelia**: Occurs in 14% of cases. Involvement of the fourth ventricle, associated with hydrocephalus.
- Congenital malformations: Chiari I malformation, encephalocele, Dandy-Walker syndrome, cysts.
- Meningitis.
- Intraspinal hemorrhage. **Noncommunicating syringomyelia**: 65% of cases. No communication with the fourth ventricle.
- Chiari I malformation.
- Spinal arachnoiditis.
- Tethered cord.
- Cystic degenerative tumors (ependymomas, astrocytomas).
- Atrophic syringomyelia (ex vacuo syringomyelia).
- Loss of parenchyma from spinal trauma or infarction.
- Equally common in both sexes. Peak age: 20–40 years.

Imaging Signs

- **Modality of choice**
  MRI.
- **Radiographic findings**
  Associated bony malformations of the spine: Atlantooccipital fusion, spina bifida, Klippel-Feil deformity. **Secondary bony changes**: Scoliosis, dilation of the spinal canal, degeneration, and/or destruction of the facet joints.
- **CT findings**
  Detailed visualization of bony changes. **CT myelography**: Where MRI is contraindicated. Widening of the spinal cord. Syrinx fills on late images.
- **MRI findings**
  General:
  - Sagittal and axial T1- and T2-weighted images.
  - CSF flow study (at the respective level or in the other CSF spaces).
  - Contrast studies are indicated in uncertain cases to exclude tumor.
  - Complete spinal cord examination: From the cervical spine including the foramen magnum (to exclude a Chiari malformation) to the lumbar spine.
  - Supplementary cerebral MRI to exclude a cerebral malformation.
T1:
Tubular to lobulated focal signal alteration in spinal cord, usually isointense to CSF, possibly partially septate.
- Signal is occasionally hyperintense to CSF due to increased protein content.
  Spinal cord may be distended.

T2:
- Focal signal alteration in the spinal cord, usually isointense to CSF.
- The pulsatile motion of the syrinx fluid produces flow voids, a finding of prognostic importance as these lesions usually respond well to creation of a shunt.

Clinical Aspects

- Typical presentation
  Dissociated sensory deficits (often loss of sensitivity to pain and temperature)
  Radicular pain
  Weakness in the extremities, sensation of stiffness in the legs
  Muscle atrophy in the upper extremities
  Neurogenic arthropathy, progressive scoliosis
  Brainstem symptoms where the lesion extends into the hindbrain.

Fig. 2.21 a, b  Spasticity, more pronounced on the left side, beginning several years after previous spinal trauma. MR image of cervicothoracic region (sagittal, T2). Intramedullary cavitation from T2 to T6, widest at T5 (detail, b).
Fig. 2.22  MR image of T5 (axial, T2). Eccentric cavitation on the left side at the level of T5. A fine linear structure extends from the spinal cord to the dura: post-traumatic tethering.

Fig. 2.23  MR image of the cervical spine (sagittal, T1) in an asymptomatic 27-year-old man. Hypointense longitudinal defect at C7.

Fig. 2.24  Same patient as in Fig. 2.23 (sagittal, T2). Hyperintense signal alteration.

Fig. 2.25  Same patient as in Fig. 2.23 (sagittal, T1 with contrast). No abnormal enhancement. No Chiari I malformation. Final diagnosis: typical syrinx.
Therapeutic options

Treatment is indicated only for symptomatic syrinx • Decompressive laminectomy and syringotomy (posterolateral myelotomy to drain the syrinx into the subarachnoid space) • Syrinx shunting • Percutaneous needle aspiration (Caution: Syrinx may refill) • Terminal ventriculostomy: With a lumbar syrinx, the filum terminale is opened to create communication between the terminal ventricle and subarachnoid space • Patients with posttraumatic syrinx respond especially well to surgical treatment.

Prognosis

This depends on the underlying cause and the severity of the neurologic deficit.

Differential Diagnosis

Cystic tumor or tumor component

Selected References