Syringomyelia

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Syringomyelia: presence of fluid-filled cavity within the spinal cord. The cavity lies outside of the central canal and not lined by ependymal cells.

Communicating syringomyelia: have direct connection with the fourth ventricle.

Noncommunicating syringomyelia: no connection to the fourth ventricle.

Hydromyelia: dilatation of the central canal of the spinal cord (cavity lined by ependymal cells)
Cervical Spinal Cord Anatomy

Motor and descending (efferent) pathways (left, red)
1. Pyramidal Tracts
   1a. Lateral corticospinal tract
   1b. Anterior corticospinal tract
2. Extrapyramidal Tracts
   2a. Rubrospinal tract
   2b. Reticulospinal tract
   2c. Vestibulospinal tract
   2d. Olivospinal tract

Sensory and ascending (afferent) pathways (right, blue)
3. Dorsal Column Medial Lemniscus System
   3a. Gracile fasciculus
   3b. Cuneate fasciculus
4. Spinocephallic Tract
   4a. Posterior spinocerebellar tract
   4b. Anterior spinocerebellar tract
5. Anterolateral System
   5a. Lateral spinothalamic tract
   5b. Anterior spinothalamic tract
6. Spino-dilucy fibers

Sensory Abbreviations:
S: Somatic, T: Thoracic, L: Lumbar, C: Cervical
Clinical Presentation

- Gradual onset of symptoms usually between 25-40 years of age.
- Pain, numbness of the hands, stiffness of the legs, vertigo, neurogenic arthropathy.
- Horner syndrome is common.
Clinical Presentation
Causes

- Familial often associated with Chiari type 1 malformation (basilar impression)
- Secondary to basal/spinal arachnoiditis after meningitis, SAH, TB, trauma, repetitive deceleration in skydivers, CSF leak after brachial plexus avulsion, reaction to contrast, spinal anesthesia.
- Secondary to intra or extramedullary spinal tumors.
- Secondary to cervical spondylosis.
- Secondary to injury to cord from trauma, radiation, infarction, hemorrhage, infection, transverse myelitis.
- Idiopathic
Type 1 Chiari malformation and syringomyelia, T1 MRI
Proposed Pathophysiology

- Multiple proposed mechanisms however not fully understood.
- Initial syrinx formation may be due to infarction of tissue with subsequent cyst formation.
- Hydrodynamic theory - cyst formation due to fluid flowing into the central canal from the forth ventricle secondary respiratory and arterial pressure waves.
- Cyst formation secondary to extracellular fluid edema after spinal cord injury.
- Changes in subarachnoid space compliance, CSF fluid or pressure dynamics from arachnoid adhesions, spinal stenosis, or cord compression could lead to syrinx enlargement.
Diagnosis

- MRI is the imaging modality of choice.
- Often demonstrates that syringes are asymmetrical, loculated, or multiple, and this information can aid operative planning.
- Phase-contrast cine MRI can localize subarachnoid space obstruction and demonstrate normalization of CSF flow following surgery.
Management

- No clear standard approach and area of controversy.
- 17-50% of patients have no progression of syrinx or symptoms over a 10 year follow-up period.
- Spontaneous resolution has been observed in children and adults.
- Surgery reported as effective in Chiari-related syringomyelia with 90% chance of long-term stabilization or improvement.
- Postoperative relief of headaches and neck pain observed in 83% of children with Chiari I malformation.
Management Continued

- Persistent dysesthetic pain can occur despite improvement or collapse of syrinx on post-op MRI.
- Aim of surgery is to reconstruct normal CSF pathways via bony decompression and duraplasty in cases associated with Chiari malformation.
- Shunting procedures can be used in cases not associated with Chiari malformation although the best type of shunting procedure is unknown (syringoperitoneal, syringopleural, syringosubarachnoid).
Management Continued

- With shunt insertion 12-53% of pts will improve, 10-56% will be unchanged, and 12-32% will get worse.
- Shunt failure rate is about 50% in most series.
- Operative complication rates range from 6-18%.
- In cases secondary to trauma or arachnoiditis post-op recurrence of adhesions is the rule.
- Fetal spinal cord stem cells have been transplanted in patients without deterioration however without evidence of growth into surrounding spinal cord.
References

- MedLink Neurology
Questions