Other Spinal Disorders

STIFF-PERSON SYNDROME (s. STIFF-MAN SYNDROME, MOERSCH-WOLTMAN SYNDROME) .............................................. 1

EPIDURAL LIPOMATOSIS ................................................................................................................................. 1

LUMBAR ADHESIVE ARACHNOIDITIS ........................................................................................................... 1

VENTRAL CORD HERNIATION ......................................................................................................................... 2

ARACHNOID WEB .............................................................................................................................................. 2

Progressive Neuritisizing Myelopathy ........................................................................................................... 2

SUBACUTE COMBINED DEGENERATION (vit. B12 deficiency) → see p. 1309(1) >>

RADIATION MYELOPATHY → see p. Rx11 >>

STIFF-PERSON syndrome (s. STIFF-MAN syndrome, MOERSCH-WOLTMAN syndrome)

EPIDEMIOLOGY
- continuous excessive firing of motor unit (due to disinhibition of descending pathways to Renshaw cells or γ- motor system): 1)
  - idiopathic (rarely autosomal dominant).
  - autoimmune – antibodies against glutamic acid decarboxylase (glutamic acid → GABA).
    * frequently associated with other autoimmune disorders (diabetes mellitus, thyroiditis, myasthenia gravis, pernicious anemia, vitiligo).
  - paraneoplastic – amphiphysin (protein associated with synaptic vesicles) has been implicated.

PATHOLOGIC STUDIES – no abnormalities!

CLINICAL FEATURES
- symptoms develop over several months or years and may either increase slowly or become stable.
- predominantly axial muscles (spread to proximal limbs occurs; cranial involvement in 25% patients).
- vs. neumyastenia (Osas’s syndrome) – distal muscles suffer most!
- examination detects only continuous motor unit activity in patients taking > 40 mg baclofen.
- rigor of diaphragmatic muscles may induce respiratory acidosis.
- stiffness diminishes during: 1) sleep;
  - general / spinal anesthesia.
  - peripheral nerve block.
- spasms are powerful enough to rupture muscles, rip surgical sutures, or fracture bones.
- without treatment, syndrome progresses to total disability (generalized rigidity and secondary musculoskeletal deformities).

DIAGNOSIS
- EMG (essential to confirm diagnosis):
  - continuous motor unit activity (as in normal contraction) at rest that patient cannot voluntarily suppress.
  - reduced motor activity after benzodiazepine administration.
- Serum CK?

TREATMENT
- controlling rigidity:
  1) Diazepam (20-300 mg/day; high doses may be required!); most effective medication!
  2) Baclofen, Tizanidine, Clonazepam, Valproic Acid, Clonidine, Botulinum toxin.

EPIDURAL LIPOMATOSIS
- steroid-induced fat deposition in epidural space.
- occurs in patients taking > 40 mg PREDNISONE for at least 4 months.
- most patients are already cishingood.
- earliest and commonest clinical feature: low back pain typically in thoracic spine (→→→ myelopathy, cauda equina syndrome, radiculopathy).
- diagnosis: MRI (fat is hyperintense on T1; and less intense on T2; vs. inflammatory processes - bright on T2).
- treatment: wide decompressive laminectomy and debulking of adipose tissue (+ weight loss in morbidly obese patient).

LUMBAR ADHESIVE ARACHNOIDITIS

ETIOLOGY
- local tissue injury → inflammatory response within subarachnoid space → fibrotic process:
  1) lumbar operations
  2) oil-based myelography
  3) blood or foreign substances in intrathecal space
  4) chronic spinal infections
  5) epidural steroid therapy.

CLINICAL FEATURES
- multifocal radiculopathy with neck / back pain (due to nerve root adhesions)
- cord involvement occurs less commonly (in severe cases may lead to paraplegia).

DIAGNOSIS
- Imaging (MRI is method of choice): nerve roots clumped together centrally or adhere to dura peripherally, CSF loculations.
- CSF – protein↑; may be mild pleocytosis and glucose↓.

TREATMENT
- lysis of adhesions: opening of subarachnoid cysts, dorsal rhizotomy, dorsal root ganglionectomy, dorsal column stimulation for pain relief.
VENTRAL CORD HERNIATION

Ventral cord herniation (on axial - cord is rotated, anterior CSF collection) – cord plugs dural tear (from disc osteophyte?) H: dural repair after dentate ligament cut.

ARACHNOID WEB

Dural scalpel sign:

PROGRESSIVE NECROTIZING MYELOPATHY

- necrotic areas in cord (esp. thoracic region); in long-standing cases cord is atrophic.
  a) young adults, after infectious illness.
  b) patients with known malignancy (esp. small-cell lung cancer or lymphomas).
  c) FOIX-ALAJONINE syndrome – see p. Vas42 >>

Clinically
  - pain in back or legs, sometimes paresthesias; sensory deficits may be conspicuous.
  - legs become weak →→→ paralyzed.
  - tendon reflexes: lost initially → spasticity and hyperreflexia.
  - sphincter disturbances are usual.
  - progressive course → respiratory disturbances, bulbar signs.

No specific treatment.

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