

Other Spinal Disorders

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

ETIOPATHOPHYSIOLOGY

- **continuous excessive firing of motor unit** (due to **disinhibition of descending pathways** to *Renshaw cells* or *γ-motor system*):

- 1) **idiopathic** (rarely autosomal dominant).
- 2) **autoimmune** – antibodies against **glutamic acid decarboxylase** (glutamic acid → GABA).
 - frequently associated with other autoimmune disorders (diabetes mellitus, thyroiditis, myasthenia gravis, pernicious anemia, vitiligo).
- 3) **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. neuromyotonia (Isaacs' syndrome) – distal muscles suffer most!
- examination detects only **muscle hypertrophy and stiffness**; passive muscle stretch provokes exaggerated reflex contraction that lasts several seconds.
- stress / exertional activity provokes progressive **painful muscle cramps** that may last for hours (resemble tetanus!):
 - 1) **severe lumbar lordosis** (chronic spasm of paraspinal muscles).
 - 2) slow and laborious movements, "**tin soldier**" gait.
- rigidity of diaphragmatic muscles may induce *respiratory acidosis*.
- stiffness diminishes during:
 - 1) sleep
 - 2) general / spinal anesthesia.
 - 3) 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):

- 1) **continuous motor unit activity** (as in normal contraction) at rest that patient cannot voluntarily suppress.
- 2) 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 cushingoid.
- 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 - brighter 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

- 1) **imaging** (MRI is method of choice): nerve roots **clumped together** centrally or **adhere to dura** peripherally, CSF loculations.
- 2) **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.

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) *thrombophlebitis* of spinal veins (**FOIX-ALAJOUANINE syndrome**).

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.

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