Other Spinal Disorders

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**Subacute Combined Degeneration** **(vit. B12 deficiency)** → see [p. 1570 (3) >>](http://www.neurosurgeryresident.net/USMLE%202\Hematology%20(1501-1649)\1570%20(3).jpg)

**Radiation myelopathy** → see [p. Rx11 >>](http://www.neurosurgeryresident.net/Rx.%20Treatment%20Modalities\Rx11.%20Radiotherapy.pdf#Complications)

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:
3. sleep
4. general / spinal anesthesia.
5. 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.

Arachnoscopy – see [“Arachnoscopy: A Special Application of Spinal Intradural Endoscopy” >>](http://www.medscape.com/viewarticle/740455?src=mp&spon=26)

Progressive Necrotizing Myelopathy

- necrotic areas in cord (esp. thoracic region); in long-standing cases cord is atrophic.

1. young adults, ***after infectious illness***.
2. patients with known ***malignancy*** (esp. small-cell lung cancer or lymphomas).
3. **Foix-Alajouanine syndrome** – see [p. Vas42 >>](http://www.neurosurgeryresident.net/Vas.%20Vascular\Vas42.%20Spinal%20Hemorrhages,%20Spinal%20Vascular%20Malformations.pdf)

##### 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 >>](http://www.neurosurgeryresident.net/Spin.%20Spinal%20Disorders\Spin.%20Bibliography.pdf)

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