ACUTE TRANSVERSE MYELITIS

Elinore Kaufman MS III
Patient JK: History

- JK is a 46 yo RH woman with a history of diabetes, hypertension and disc disease who presented with a 4d history of neck pain and progressive weakness.

Course of illness:
- 5d PTA: slight sore throat
- 4d PTA: awoke with sore, stiff neck, relieved with ibuprofen.
- 3d PTA: difficulty holding pen to sign name
- 2d PTA: difficulty walking; dragging right foot.
Patient JK: Physical Exam

- General remarkable only for 2cm papular rash on L upper chest.
- Neurological examination:
  - Mental Status: intact
  - Cranial Nerves II-XII: intact
  - Motor: Normal bulk and tone in upper and lower extremities. No tremor or asterixis.
  - Strength:
    |   | Delt | Bic | Tric | WE | WF | FE | FF | IP | Qu | HS | TA | Gas | EHL |
    |---|-----|-----|-----|----|----|----|----|----|----|----|----|-----|-----|
    | L | 5   | 5   | 5   | 5  | 5  | 5  | 5  | 5  | 5  | 4+ | 5  | 5   | 5   |
    | R | 4+  | 4   | 4+  | 4+ | 4+ | 4+ | 4+ | 4+ | 5  | 4+ | 4+ | 5   | 4   |
  - Sensation: Light touch intact throughout. Pinprick diminished in R C5 distribution and L leg.
  - Coordination: No evidence of resting or intention tremor or dysmetria, allowing for weakness.
  - Gait: Using walker with good initiation and right steppage gait
Now we will examine imaging techniques for the spinal cord and spinal anatomy.
Imaging Myelopathy

- Plain films
  - Stability after trauma
  - Osteophytic narrowing of spinal canal
- CT: better assessment in trauma
- Spine MRI: MRI: study of choice for cord and soft tissues, as well as for surgical planning
  - Gadolinium contrast, especially if AVM is in the differential
  - High sensitivity
- CT myelogram if MRI is unavailable or contraindicated
  - Invasive
  - Identifies only large masses
  - Does not distinguish between cystic and solid lesions
- Radionucleide bone scan can be useful in cases of tumor and infection
- Brain MRI
  - Associated demyelinating lesions

(Seidenwurm 2008)
Companion Patient 1: Normal Cervical Spinal MRI

Sagittal and axial views of cervical spinal cord on T2-weighted MRI (PACS BIDMC).
Now we will return to our patient JK to discuss her imaging findings and diagnosis.
Patient JK: Sagittal MRI of Cervical Spine

Sagittal T2-weighted image showing disc bulge and hyperintense lesion within spinal cord. (PACS BIDMC)

Sagittal T1-weighted image after IV gadolinium administration showing ring enhancing lesion. (PACS BIDMC)
Patient JK: Enhancing lesion on axial T1-weighted Image with Contrast

Enhancing lesion seen on axial T1 image, post-contrast (PACS BIDMC)
Work-up of Acute Myelopathy

Adapted from Jacob 2008

Compression: disc, pathological fracture, metastasis, spondylolisthesis

Surgical Assessment

Acute myelopathy

MRI Spine

Normal MRI

Missed compression, e.g. epidural lipomatosis, dynamic compression on movement

Other region of neuromuscular axis

Systemic metabolic, degenerative, vascular or infectious cause

Myelopathy imaged too late or too early

Abnormal MRI, noncompressive changes: Acute Transverse Myelitis (ATM)

Brain MRI

Normal

Demyelinating lesions: MS

CSF and serum studies

Identify inflammatory or infectious cause

Idiopathic Acute Transverse Myelitis

Patient JK
# Differential Diagnosis of Acute Myelopathy

## Noninflammatory
- Compression
  - Osteophyte
  - Disc
  - Metastasis
  - Trauma
- Tumor
- Paraneoplastic syndromes

## Inflammatory
- Demyelinating disease
  - Multiple sclerosis (MS)
  - Neuromyelitis optica (NMO)
  - Acute disseminated encephalomyelitis (ADEM)
  - Idiopathic acute transverse myelitis (IATM)
- Infection
  - Viruses: coxsackie, mumps, varicella, CMV
  - Tuberculosis
  - Mycoplasma
- Inflammatory disorders
  - Systemic lupus erythematosus
  - Neurosarcoidosis

Jacob 2008, Seidenwurm 2009
<table>
<thead>
<tr>
<th>Condition</th>
<th>MRI Spinal Cord</th>
<th>MRI Brain</th>
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<tbody>
<tr>
<td>MS</td>
<td>Oval-shaped lesion(s) &lt; 2 spinal cord segments, usually peripheral location. Hyperintense on T2 with homogeneous or ring enhancement.</td>
<td>White matter lesions: periventricular, juxtacortical, infratentorial lesions. Dawson’s Fingers.</td>
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<tr>
<td>NMO</td>
<td>lesion &gt;3 segments; cord swelling and gadolinium enhancement acutely.</td>
<td>Optic neuritis. Cerebral lesions in 60%.</td>
</tr>
<tr>
<td>ADEM</td>
<td>lesion &gt;3 segments; cord swelling and gadolinium enhancement acutely.</td>
<td>Large, confluent white matter lesions.</td>
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<tr>
<td>IATM</td>
<td>Lesion of variable length and shape, &gt;50% of cross-sectional area on axial scan. Hyperintense on T2-weighted images, variable hypointensity on T1, patchy enhancement and swelling.</td>
<td>No brain lesions.</td>
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Idiopathic Acute Transverse Myelitis

- Incidence: 1-4 per million per year
- Diagnosis
  - Myelopathic symptoms
    - 50% lose all leg movements
    - Nearly 100% have bladder dysfunction
    - 80-94% of patients sensory dysfunction
  - MRI evidence of noncompressive myelopathy
  - Inflammation: CSF pleocytosis, elevated CSF IgG index, or gadolinium enhancement
    - if no evidence but strong suspicion, repeat MRI/LP in 2-7 d
  - Exclude: cord radiation, thromboembolic disease, AVF, connective tissue disease, infection, MS/ADEm, malignancy
- Pathology: edema, necrosis, demyelination, white>gray matter changes (Misra 1996)
- Prognosis
  - 1/3 recover without sequelae
  - 1/3 have moderate, permanent disability
  - 1/3 have severe disability (Transverse Myelitis Consortium Working Group 2002)
Radiologic Findings in IATM

- Of patients with ATM and normal brain MRI, none with normal CSF progressed to MS.
  - 53% of those with CSF oligoclonal bands or elevated IgG index developed MS. (Perumal 2008)

- Spinal cord atrophy may be evident on follow-up MRI. (Shen)

- Fractional anisotropy
  - Shows water diffusion in the extracellular space along the axon fibers
  - Higher sensitivity than T2 for white matter damage: can identify lesions in normal-appearing cord
  - Decreased values may demonstrate increased extracellular space due to demyelination, and may predict greater wallerian degeneration and worse prognosis. (Renoux 2006, Lee 2008)
Now we will examine other causes of acute transverse myelitis.
Companion Patient 2: Transverse Myelitis in MS

- Lesions <2 segments
- No cord swelling
- Peripheral lesions
- Differential: 1-3% of lupus patients develop ATM with similar findings to MS.

Sagittal T2-weighted image from MS patient (Scotti 2001)

Axial T2-weighted images from MS patient (Jacob 2008)
Risk of Developing MS after ATM

- **MR Brain**
  - $\geq 2$ lesions: 88% develop MS. (Jacob 2008)
  - No brain lesions: 10-33% develop MS.
- **Mean time to MS:** 16m, no new cases $>24m$.
- **Next evidence of MS** is more likely to be clinical than MRI based (serial MRI does not speed diagnosis).
- **African American ATM patients** are more likely to develop MS than Caucasian patients: (35% vs. 26%).
  - African Americans develop MS more quickly, in 9.6 months vs. 20.3 for Caucasians.
  - African Americans experience greater disability from this disease despite earlier use of disease-modifying therapy. (Perumal 2008).
Companion Patient 3: Dural arteriovenous fistula on MRI

Longitudinal hyperintense lesion on T2; contrast enhancement of dilated blood vessels: AVF (Jacob 2008)

- AVMs and dural arteriovenous fistulae can either bleed or cause increased venous pressure and progressive myelopathy; rarely acute onset.
- Anterior spinal artery occlusion: atherosclerosis or complication of aortic surgery: peak within 4h (Jacob 2008)
- Vasculitis can cause cord swelling followed by cord atrophy. (Scotti 2001)
Companion Patients 4 & 5: Spinal Cord Tumor on MRI

Sagittal T2 image of lesion initially thought to be tumor, ultimately determined to have an inflammatory cause. (Brinar 2006)

Sagittal T2 image of lesion initially thought to be inflammatory, ultimately determined to be a tumor. (Jacob 2006)
Companion Patient RB: History and Physical Exam

- RB is a 32yo LH woman who presented with severe headache and worsening neurological symptoms:

  **Course of illness:**
  - 3 wks of severe R-sided retro-orbital headache
  - 2 wk of numbness in both legs up to the navel
  - 1 wk of R eye blurry vision
  - 3d of urinary retention
  - 1d of bowel incontinence

- Physical exam confirmed severe vision loss, decreased pinprick sensation to the T8 level, and upper motor neuron weakness in the legs.
Companion Patient RB: Spinal lesions and leptomeningeal enhancement on MRI

Post-contrast T1-weighted image showing three enhancing lesions and linear leptomeningeal enhancement. (PACS BIDMC)
Companion Patient RB: Clinical Course

- She was treated with acyclovir (possible CNS HSV), ciprofloxacin (E. Coli UTI) and methylprednisolone.

- Her vision improved, and she was discharged.

- 1 wk later, she returned with fever and severe L-sided retro-orbital headache. Her other symptoms were stable, but her vision was now extremely blurry in her left eye.

- She was readmitted and received plasmapheresis with slight improvement, but her symptoms worsened again after discharge.
Companion Patient RB: Diagnostic Work-Up

- Brain MRI showed only the optic neuritis.
- CSF showed pleocytosis and elevated protein but no oligoclonal bands.
- Extensive CSF and serum studies showed no evidence of CNS infection, collagen vascular disease or malignancy.
- Preliminary diagnosis: Neuromyelitis Optica
  - Atypical features
    - Spinal cord lesion length <3 segments
    - Presence of leptomeningeal enhancement more typically associated with lymphoma, sarcoidosis or Lyme disease
    - Absence of NMO antibody in serum and CSF.
- Chest CT showed no evidence of sarcoidosis or lymphoma
- PET scan was performed to rule out lymphoma and showed no lymphadenopathy, bone lesions, or other abnormal uptake.
- Multiple sclerosis
Summary

- Acute myelopathy is most often compressive but otherwise may be infectious, inflammatory or idiopathic.
- MRI is the imaging modality of choice for the spinal cord, although CT myelography may be used if needed.
- Imaging locates the lesion and can provide clues to the diagnosis but often leaves a wide differential.
  - MS: other CNS lesions
  - NMO: lesion >3 segments
  - Idiopathic ATM: diagnosis of exclusion
Acknowledgements

Many thanks to:

- Dr. Daniel Cohen, Dr. Jed Barasch, Dr. Jennifer Avallone and Dr. Courtney McIlduff of the BIDMC Neurology service
- Maria Levantakis
- Dr. Gillian Leiberman


