Acute Transverse Myelitis (ATM)

1. Idiopathic (Post-infectious, para-infectious)

2. **Acquired Demyelinating Disease**
   - Multiple sclerosis
   - Neuromyelitis Optica

1. Systemic Disease
Tract specific
Lateral corticospinal tract
MRI clues

- 1-2 vertebral levels: MS
- Partial, asymmetric, periphery (not central) WM tracts: MS
- >3 vertebral levels, LETM (Longitudinally Extensive Transverse Myelitis): NMO
Acute Transverse Myelitis (ATM)

1. Idiopathic (Post-infectious, para-infectious)
2. Acquired Demyelinating Disease (MS, NMO)
3. **Systemic Disease**
   - SLE
   - Sjogren’s
   - Sarcoid
   - Connective tissue diseases
TM associated with auto immune disease

- SLE, systemic lupus erythematosus
- Sjogren’s syndrome
- Sarcoidosis
- Mixed connective tissue disease
- Antiphospholipid antibody syndrome
- Behcets
- cryglobulinemia
Further Work-up

• CSF: cells, protein, IgG
  – Pleocytosis indicates inflammation
  – Confirmatory, supplemental lab tests for systemic disease
    • ANA, SSA, SSB, ACE,

• MRI Brain or more spinal levels

• CBC, CMP
Treatment

• Specific disease
• For TM
  – Methylprednisolone (Solu-Medrol) 1 gram IV daily x 5d
  – No RCT, based on MS,
  – PLEX for not responders.
  – ↓ Edema, ↓ cytokine
  – Cyclophosphamide for SLE, Sjogrens
• Long term immunomodulatory or immunosuppressive Rx for demyelinating disease
Treatment

- DVT prophylaxis
- Analgesics
- Baclofen
- Rx neurogenic bladder and bowel
Pregnant Lawyer

- ER
- Hx Familial Mediterranean Fever
- Brain MRI
- SVD
- MRI T/LS cord
# Moderate Paraparesis

<table>
<thead>
<tr>
<th>Right</th>
<th>Left</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>5</td>
<td>C5 - Elbow flexors (biceps, brachialis)</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>C6 - Wrist extensors (extensor carpi radialis longus and brevis)</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
<td>C7 - Elbow extensors (triceps)</td>
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<tr>
<td>5</td>
<td>5</td>
<td>C8 - Finger flexors (flexor digitorum profundus) to the middle finger</td>
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<tr>
<td>5</td>
<td>5</td>
<td>T1 - Small finger abductors (abductor digiti minimi)</td>
</tr>
<tr>
<td>4</td>
<td>3-</td>
<td>L2 - Hip flexors (iliopsoas)</td>
</tr>
<tr>
<td>4-</td>
<td>4</td>
<td>L3 - Knee extensors (quadriceps)</td>
</tr>
<tr>
<td>3+</td>
<td>3</td>
<td>L4 - Ankle dorsiflexors (tibialis anterior)</td>
</tr>
<tr>
<td>3</td>
<td>3-</td>
<td>L5 - Long toe extensors (extensor halluces longus)</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>S1 - Ankle plantar-flexors (gastrocnemius, soleus)</td>
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## Sensory Examination

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<tr>
<th></th>
<th>Right</th>
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<tbody>
<tr>
<td>T6</td>
<td>2</td>
<td>2</td>
<td>Sixth IS (level of xiphisternum)</td>
</tr>
<tr>
<td>T7</td>
<td>2</td>
<td>2</td>
<td>Seventh IS (midway between T6 and T8)</td>
</tr>
<tr>
<td>T8</td>
<td>2</td>
<td>2</td>
<td>Eighth IS (midway between T6 and T10)</td>
</tr>
<tr>
<td>T9</td>
<td>2</td>
<td>2</td>
<td>Ninth IS (midway between T8 and T10)</td>
</tr>
<tr>
<td>T10</td>
<td>2</td>
<td>2</td>
<td>Tenth IS (umbilicus)</td>
</tr>
<tr>
<td>T11</td>
<td>2</td>
<td>2</td>
<td>Eleventh IS (Midway between T10 and T12)</td>
</tr>
<tr>
<td>T12</td>
<td>2</td>
<td>2</td>
<td>Inguinal ligament at mid-point</td>
</tr>
<tr>
<td>L.1</td>
<td>2</td>
<td>2</td>
<td>Half the distance between T12 and L2</td>
</tr>
<tr>
<td>L.2</td>
<td>2</td>
<td>2</td>
<td>Mid-anterior thigh</td>
</tr>
<tr>
<td>L.3</td>
<td>1→2</td>
<td>2</td>
<td>Medial femoral condyle</td>
</tr>
<tr>
<td>L.4</td>
<td>1→2</td>
<td>2</td>
<td>Medial malleolus</td>
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<tr>
<td>L5</td>
<td>1→1</td>
<td>2</td>
<td>Dorsum of the foot at the third metatarsal phalae</td>
</tr>
<tr>
<td>S1</td>
<td>1→2</td>
<td>2</td>
<td>Lateral heel</td>
</tr>
<tr>
<td>S2</td>
<td>1→2</td>
<td>2</td>
<td>Popliteal fossa in the mid-line</td>
</tr>
<tr>
<td>S3</td>
<td>1→1</td>
<td>1→1</td>
<td>Ischial tuberosity</td>
</tr>
<tr>
<td>S4-S5</td>
<td>0→1</td>
<td>0→1</td>
<td>Perianal area (taken as one level)</td>
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</table>
Workup showed a normal or negative studies including the following

- MRA of the brain, MRV of the brain, MRI C-spine, CT angiogram in the abdomen and pelvis, Echo
- Negative HIV, RPR, lupus, anticoagulant screening, anticardiolipin antibodies, beta-2 glycoproteins, homocysteine, ANA, double-stranded DNA, SSA, SSB, C4 level. Protein S is normal.
- She has had a total cholesterol of 216, LDL of 103, C3 slightly elevated, anti thrombin III slightly elevated, Factor II activity slightly elevated and protein C minimally elevated. Pending tests include ANCA, rheumatoid factor, Factor V Leiden, hepatitis C.
- CSF protein was also normal.
- Pending FMF genetic study. (MEFV gene)
Treatment

- Prednisone and ASA
- heparin for DVT prophylaxis
- colchicine.
- She is also on iron, prenatal vitamins and vitamin C.
Infarction of cord

- Venous hypertension, localized leading to venous infarction, reduced perfusion
- Arterial
  - Thoracic cord
  - Aortic manipulation
  - Hypotension
  - Artery of Adamkiewicz
  - AVMs, AV Fistulas
Differential Diagnosis Mimics

- venous or arterial infarction
- GBS
Differential diagnosis: Mimics GBS

- Guillain-Barré Syndrome or acute demyelinating inflammatory polyradiculoneuropathy (AIDP)
- Transverse myelitis must be differentiated acutely from the most common paralytic acquired illness in the United States:
Differential Diagnosis Mimics

- venous or arterial infarction
- GBS
- Infectious myelitis
  - Treponema pallidum
  - HIV vacuolar myelopathy
  - Polio, West Nile virus
  - VZV, HSV, CMV
  - bacterial
Differential Diagnosis Mimics

- venous or arterial infarction
- GBS
- Infectious
- Hypokalemia
- B12, nitrous oxide,
- Copper deficiency (Zinc)
- Neoplasia
- hereditary
Other Myelopathies, usually subacute to chronic

• Neoplasia
  – ependymoma, astrocytoma, hemangioblastoma
  – Paraneoplastic, SP Radiation

• Hereditary
  – Spastin gene (HSP), Frataxin gene (Friederichs ataxia)
Gliosarcoma brain and spine
Other Myelopathies, usually subacute to chronic

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• Hereditary
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Table 1. Diagnostic Criteria for Transverse Myelitis.*

<table>
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<td>Bilateral (not necessarily symmetric) sensorimotor and autonomic spinal</td>
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<td>cord dysfunction</td>
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<td>Clearly defined sensory level</td>
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<td>Progression to nadir of clinical deficits between 4 hours and 21 days</td>
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<tr>
<td>after symptom onset</td>
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<td>Demonstration of spinal cord inflammation: cerebrospinal fluid pleocytosis</td>
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<td>or elevated IgG index,† or MRI revealing a gadolinium-enhancing cord</td>
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<tr>
<td>lesion</td>
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<td>Exclusion of compressive, postradiation, neoplastic, and vascular causes</td>
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* Clinical events that are consistent with transverse myelitis but that are not associated with cerebrospinal fluid abnormalities or abnormalities detected on MRI and that have no identifiable underlying cause are categorized as possible idiopathic transverse myelitis.

† The IgG index is a measure of intrathecal synthesis of immunoglobulin and is calculated with the use of the following formula: (CSF IgG ÷ serum IgG) ÷ (CSF albumin ÷ serum albumin), where CSF denotes cerebrospinal fluid.
Abnormal MRI of cord

• Axial anatomy: complete/central or partial with specific tract involvement
• Sagittal longitudinal anatomy: 1 vertebral segment, 3 or more vertebral segments
• Gadolinium enhancement? Inflammation
• Normal Cord? Confirm suspected level, consider early involvement, consider alternatives: GBS
Transverse Myelitis

• Inflammatory
  – MS, NMO
  – Systemic
  – Post or para-infectious
  – Idiopathic
  – infection

• Non inflammatory
  – infarction
  – Radiation myelitis
Summary

• Acute myelopathy: recognize it
• MRI a key study
• Acute transverse myelitis: many causes, Big 3: primary demyelinating, systemic, idiopathic/post-infectious.
• Rx steroids:
Questions and Comments

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