QUESTION 50

A 43yo woman presents with a 2 day history of mid-thoracic back pain and leg weakness. On 
examination, cranial nerve and upper extremities are normal. Lower extremity examination 
reveals normal tone, with brisk reflexes (left greater than right) and an upgoing left plantar 
response. Power is reduced with grade 4/5 power in hip flexion and ankle dorsiflexion. Sensory 
examination reveals reduced sensation to pin-prick and temperature involving the right leg to 
the level of the umbilicus. Her MRI scans are shown below.

The most likely diagnosis is:

A. Transverse myelitis
B. Spinal cord compression
C. Anterior spinal artery thrombosis
D. Spinal cord arteriovenous malformation
E. Syringomyelia

SPINAL CORD DISEASES

There are a number of causes of spinal cord pathology; these are best divided into acute and 
chronic causes.
Acute:
- Compressive myelopathies
  o Neoplastic
  o Epidural abscess
  o Epidural haematoma
  o Haematomyelia
- Acute transverse myelopathies (non-compressive)
  o Spinal cord infarction
  o Immune-mediated (eg: SLE, sarcoidosis)
Chronic:
- Spondylitic myelopathy
- Vascular malformations
- Retrovirus-associated myelopathy
- Syringomyelia
- MS
- Subacute combined degeneration (B12 deficiency)
- Tabaes Dorsalis (syphilis)
- Familial spastic paraplegia
- Adrenomyeloneuropathy
- Toxins
- Paraneoplastic
- Radiation damage
- Primary lateral sclerosis

This patient has an acute myelopathy so this immediately rules out syringomyelia and AV malformation. The clinical information suggests a predominantly left sided lesion with spastic weakness of the left leg >right leg and loss of pain and temperature of the right leg. The sensory level suggests a lesion around T10.

I would expect spinal artery thrombosis to cause symmetrical symptoms.

The options that remain are transverse myelitis and cord compression. I don’t think you can really differentiate the 2 based on the history provided so need to look at the MRI. It looks to me like there is something pushing on the spinal cord in the thoracic region. My immediate thought was of vertebral mets but not sure if this is actually the case or not. Usually mets look hypointense on T1. This image is T2. The horizontal plane scan looks like there may be destruction of the vertebral body which would fit with mets.

Anyway the correct answer is B spinal cord compression.

Important to differentiate between compressive and non-compressive myelopathies as compression needs immediate intervention. Ultimately need MRI to decide between the 2 but pain prior to development of neurological signs is more suggestive of compression.

**COMPRESSIVE MYELOPATHIES**

**Neoplastic**

- Almost any malignancy can metastasise to the spinal column
- Most common ones are breast, prostate, lung, kidney and lymphoma
- Thoracic cord most commonly involved (but lumbosacral region more common with prostate and ovarian Ca)
- Pain is first symptom – usually precedes compressive symptoms by weeks to months
- Urgent MRI is required when signs of compression present
- Radicular symptoms only means imaging can be delayed for 24-48hrs if necessary
- Pain alone means the scan can wait a few days
- Treatment for mets = high dose steroids, XRT
- Intradural tumours are usually benign and slow growing (meningiomas, neurofibromas most common)
- Symptoms usually start with radicular pain followed by asymmetric progressive spinal cord syndrome
- Treatment = surgery
- Intramedullary tumours are uncommon – present with cord or hemicord syndromes
- Mostly ependymomas, haemangioblastomas or low-grade astrocytomas

**Spinal Epidural Abscess**

- *Clinical triad of pain, fever and rapidly progressive weakness*
- Inflammatory markers raised
- Risk factors include impaired immune function, IVDU, infection of skin or other tissues
- 2/3 result from haematogenous spread of skin, soft tissue or deep visceral infection
- 1/3 result from direct extension of local infection (e.g. vertebral OM, decubitus pressure ulcers, complications of LP/epidural/spinal surgery)
- Most cases due to staph aureus, others include strep, GNB, anaerobes and fungi
- TB important in developing world
- Treatment = decompressive laminectomy with debridement and long-term Abx

**Epidural Haematoma**

- Haemorrhage into epidural or subdural space
- Rare complication of LP/epidural
- Acute onset of pain and variable signs of cord compression
- Treatment = surgery, reversal of clotting defects if present

**Haematomyelia**

- Haemorrhage into spinal cord itself
- Due to trauma, vasculitis, vascular malformation, bleeding disorders or spinal cord neoplasm
- Therapy is supportive

**ACUTE TRANSVERSE MYELOPATHIES**

- Acute transverse myelopathies are rapidly progressive spinal cord syndromes
- Associated with leg weakness, incontinence and bilateral sensory loss not due to cord compression
- Time from onset to maximum symptoms is often hours to days but can occur more slowly
- 5 general causes of acute transverse myelopathies:
  - Spinal cord infarction
  - Systemic disorders including SLE and sarcoidosis
  - Infectious
  - Demyelinating diseases
  - Idiopathic transverse myelitis
Spinal Cord Infarction

- There is a single anterior spinal artery and paired posterior spinal arteries which are the main blood supply to the spinal cord
- Anterior spinal artery supplies the anterior 2/3 of the spinal cord
- Spinal cord ischaemia can occur at any level but is most common in the *upper-thoracic segments* (watershed area)
- Infarction results in *spastic paralysis, dissociated sensory loss affecting pain and temperature but sparing vibration and position sense, and loss of sphincter control*
- Onset can be sudden but is more often *progressive over a few minutes to hours*
- Back pain at the level of infarction is common
- Areflexia due to spinal shock is often present early but spasticity appears with time
- Infarction in the territory of the posterior spinal arteries can also occur leading to loss of vibration and position sense
- Spinal cord infarction is associated with aortic atherosclerosis, aortic dissection and hypotension
- Other possible causes include cardiogenic emboli, vasculitis and surgical interruption of the aorta
- In many cases no cause is found
- MRI may show infarction
- Treatment aimed at predisposing condition

Immune-Mediated

- Acute transverse myelopathies occur in ~1% of pts with *SLE* and can be presenting symptom
- CSF normal or mildly elevated lymphocytes, antiphospholipid antibodies present in 2/3 of cases
- May respond to steroids or cyclophosphamide
- Other immune-mediated causes include Sjogren’s syndrome, mixed connective tissue disease, Behcet’s syndrome and vasculitis with pANCA antibodies
- *Sarcoid* myelopathy can mimic a tumour with large oedematous swelling
- CSF can show elevated lymphocytes and oligoclonal bands
- Look for other evidence of sarcoid – CXR/CT, slit lamp examination for uveitis, serum ACE
- Treat with steroids +/- immunosuppressants

Infectious

- Many *viruses* associated with acute myelitis caused by direct infection of the spinal cord
- Herpes zoster most common, others include HSV 1 + 2, EBV, CMV, rabies
- Treat with antivirals
- Bacterial causes less common but almost any species can be responsible including mycoplasma
- Schistosomiasis is important parasitic cause in endemic areas
- Consider toxoplasmosis particularly in patients with AIDS
- Postinfectious myelitis or postvaccinial myelitis can occur with many agents
Demyelination
- MS can present as acute transverse myelopathy but rare in Caucasians
- Neuromyelitis optica is a syndrome of ATM plus optic neuritis that is associated with MS as well as other immune-mediated diseases such as SLE
- MRI for diagnosis, including MRI brain to assess for other lesions (and thus likelihood of progressing to MS)
- Treat with steroids

Idiopathic Transverse Myelitis
- No cause found in about ¼ of cases of ATM
- In cases associated with inflammation (eg: contrast enhancement on MRI or CSF leukocytosis) but no evidence of infection, glucocorticoids are 1st treatment choice, followed by plasma exchange

CHRONIC MYELOPATHIES

Spondylitic Myelopathy
- Common in elderly
- Bony impinging and overgrowth of soft tissue causes compression of nerve roots
- Neck and shoulder pain are early symptoms, followed by radicular arm pain, most often C5-C6 distribution with muscle wasting in hands
- Compression of cervical cord produces a slowly progressive spastic paraparesis, parasthesia especially vibration sense
- Urgency or incontinence in advanced cases
- Diagnosis with MRI
- Treatment = surgery, cervical collar can help in milder cases

Vascular Malformations
- AVMs most often located posteriorly at or below a mid-thoracic level
- Typically present in middle age with progressive myelopathy
- Acute deterioration can occur with haemorrhage
- At presentation most patients have sensory, motor and bladder disturbances
- Can be mixture of UMN and LMN signs
- Diagnosis by MRI (but may miss small AVMs) and spinal angiography
- Treatment = embolisation

Retrovirus-Associated
- Slowly progressive spastic paraparesis with variable sensory and bladder disturbance is common presentation associated with HTLV-I
- Usually thoracic level
- Insidious onset, may be asymmetric signs, hyperreflexia only sign in arms
Syringomyelia

- Developmental, slowly enlarging cavity in cervical cord
- Insidious onset in teens or early 20s
- Irregular progression, may be spontaneous arrest for several years
- Pathophysiology controversial
- Central cord syndrome with dissociated sensory loss and areflexic weakness of ULs
- Cape like distribution of pain/temperature loss
- As lesion enlarges, spastic weakness of LLs and Horner’s syndrome may occur
- Associated with Chiari type 1 malformation (protrusion of cerebellar tonsils through foramen magnum)
- Treatment = ?surgery – generally unsatisfactory

MS

- Chronic progressive myelopathy is common
- Typically asymmetrical

Subacute Combined Degeneration

- Due to B12 deficiency
- Predominant involvement of posterior and lateral tracts
- Presents with paraesthesia in hands and feet, early loss of vibration and position sense, progressive spastic and ataxic weakness
- Reflexes may be absent due to superimposed peripheral neuropathy

Tabes Dorsalis

- Rare
- Characteristic fleeting but repetitive lancinating pains in legs>arms, face, back
- Primarily affects dorsal columns
- Loss of position sense → sensory ataxia in 50%
- Cardinal signs = loss of reflexes in legs, impaired position and vibratory sense, Romberg’s sign and bilateral Argyll Robertson pupils
- Can get paraesthesia and bladder disturbances also

Familial Spastic Paraplegia

- AD, AR and X-linked forms
- Present with progressive spasticity and weakness in legs (corticospinal tracts predominantly affected)
- Usually symmetrical
- Sensory signs generally absent
- Variable age of onset
**Adrenomyeloneuropathy**

- X-linked, variant of adrenoleukodystrophy
- Presents as progressive spastic paraparesis in early adulthood
- Adrenal insufficiency

**Primary Lateral Sclerosis**

- Degenerative disorder resembling ALS but without LMN involvement
- Spasticity and weakness +/- dysphonia and dysarthria

**Lesion of the Cervical Cord**

- Upper cervical cord lesions produce spastic quadriplegia and diaphragm weakness – breathing by accessory muscle only
- Lesions at C5 result in LMN weakness of biceps, deltoid, brachioradialis and rhomboids with loss of BJ and inversion of brachioradialis jerk (UMN signs in rest of UL and LLs)
- Lesions at C8 produce wasting of intrinsic muscles of hand (UMN signs in LL)

**Lesions of Thoracic Cord**

- Sensory level on trunk (nipples T4, umbilicus T10)
- UMN weakness of LLs
- T7-8 causes loss of upper abdominal reflex
- T10-11 causes loss of lower abdominal reflex and upwards displacement of umbilicus

**Lesions of Lumbar Cord**

- L1 – loss of cremasteric reflex
- L4 – LMN weakness and wasting of quadriceps, loss of KJ
- L5-S1 – LMN weakness of knee flexion and hip extension (S1) and abduction (L5) plus ankle and foot movements, KJ present, AJ lost, no plantar response, anal reflex present

**Lesions of Sacral Cord/Conus Medullaris**

- Saddle anaesthesia (S3-S5)
- Prominent bladder and bowel dysfunction, impotence
- Loss of anal reflex
- Muscle strength largely preserved

**Cauda Equina**

- Low back or radicular pain
- Asymmetric leg weakness
- Asymmetric sensory loss
- Variable areflexia
- Relative sparing of bladder and bowel function