Interactive Cases: Spinal Tumors and Mimics

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Lecture Outline

- Three unknown cases
  - Intradural, Intramedullary Tumor or Mimic
  - Intradural, Extramedullary Tumor or Mimic
  - Extradural Tumor or Mimic
- Ten predominately intradural spinal tumor or mimic cases → quick case summary → board style questions mixed in
- Revisit unknown cases

Unknown Case #1 – Intramedullary Lesion

Patient: 57 year old male diagnosed with “multiple sclerosis” in 1999, has had progressive left sided weakness and tingling over many years

Unknown Case #2 – Intradural, Extramedullary Lesion

Patient: 31 year old female with history of chronic pain secondary to pelvic chondrosarcoma
**Unknown Case #3**
**EXTRADURAL LESION**
Patient: 25 year old male with sporadic lower back pain

**Case #1**
**INTRAMEDULLARY LESION**
Patient: 34 year old male with 3 months of neck pain, mild upper extremity numbness, found on initial MRI

**Case #1 – Intramedullary Lesion**
**Imaging Findings:** Cord expansion and edema, mass with solid/polar cystic components; foci of T2 hypointense hemorrhage on axial; syringomyelia extending to thoracic spine

**Case #1 – Ependymoma, WHO Grade II**
- Arises from central canal ependymal lining
- Well *circumscribed* mass with symmetric cord expansion
- *Cervical spine* most common, followed by thoracic spine, then conus
- Majority enhance
Case #1 – Ependymoma, WHO Grade II

- **Hemorrhage** – T1 hyperintensity; focal T2 hypointensity corresponding to hemosiderin; T2* GRE hypointensity
- **Cap Sign**: cranial/caudal margin hemosiderin
- **Cysts**: tumoral cysts (necrosis), rostral/caudal polar cysts, syrinx beyond tumor margins
- Rare: intradural, extramedullary

Which of the following statements about spinal ependymomas is NOT true?

- A: Ependymomas are less common than spinal astrocytomas in adults
- B: Associated with neurofibromatosis type II
- C: Most commonly occur between age 35 and 45
- D: Low risk of local and intracranial recurrence after surgical resection

Case #2 – Intramedullary Lesion

**Imaging Findings**: Cord expansion and edema with cystic change and thoracic syrinx

Case #2 – Coccidioidomycosis

- Coccidioides immitis – *fungus*
- Endemic to **soil of southwest USA**, Mexico, Central America, and South America
- **Primary pulmonary infection** from inhaled spores
- **1-5% get disseminated disease**
  - more common in pregnant or immunocompromised
**Case #2 – Coccidioidomycosis**

- Lammering et al. (AJR June 2013) - retrospective imaging review of 23 patients with proven coccidioidal CNS meningitis (CSF and blood antibody titers)
- **23/23 patient had intracranial abnormalities**
  - 91% leptomeningeal enhancement predominately in basal cisterns
  - 78% hydrocephalus (moderate/severe in 72% of those patients, 61% had transependymal flow of CSF)
  - 90% enhancement in MCA cisterns
  - 26% had acute infarcts

- **19/22 (86%) patients had concomitant intraspinal disease**
  - 84% of those patients → leptomeningeal enhancement
  - 63% of those patients → arachnoiditis (nerve root clumping/thickening)
  - 37% of those patients → intramedullary T2 hyperintense signal abnormalities
  - 42% of those patients → focal extramedullary intradural lesions
  - 47% patients who underwent myelogram → complete spinal blocks

**Snowbird in Arizona**

*300k a year estimated in Arizona alone:*

“Retired folk who escape from their cold native habitats to the warm and dry desert of Arizona from November-April every year. While local shopkeepers, innkeepers, and other employees of the service industry may tell them that their dollar is welcome, every native son and daughter of the desert prays for the day the temperature gets above 90 degrees. That is the point that people from the regions known as the “Midwest”, “Canada”, and the “East Coast” board their land barges, manufactured by companies such as Buick, Lincoln, and Winnebago and begin their trek back to the colder lands.”

— Urban dictionary definition

**Case #3 – Intramedullary Lesion**

*Imaging Findings: Cord expansion and edema; T2 hypointense ring shaped capsule; ring enhancement of the capsule*

**Case #3**

**INTRAMEDULLARY LESION**

Patient: 51 year old immunocompromised male with history of streptococcal endocarditis
**Case #3 – Intramedullary Lesion**

*Imaging Findings:* Left frontal lobe ring enhancing lesion with T2 hypointense capsule and surrounding vasogenic edema

**Case #3 – Spinal Cord Abscess**

- Very *rare* entity, commonly delayed diagnosis
- Often present with signs of structural cord lesion as opposed to infection
  - *Idiopathic, iatrogenic,* or *hematogenous seeding*
  - Can be from extension/rupture of epidural abscess

**Case #3 – Spinal Cord Abscess**

- Dysraphism or dermal sinus tract usually present in children with spinal abscess
  - *High mortality/morbidity* → persistent neurologic deficit common
- Additional imaging: *Diffusion +*

**Case #4**

*INTRAMEDULLARY LESION*

Patient: 44 year old male with PMH of head and neck cancer with 12 month history of right leg weakness

**Case #4 – Intramedullary Lesion**

*Imaging Findings:* T2 hyperintense lesion along dorsal cord with serpentine flow voids; long segment cord edema above lesion

**Case #4 – Intramedullary Lesion**

*Imaging Findings:* Homogeneous enhancement associated with lesion along dorsal thoracic cord
**Imaging Findings:** DSA for preoperative embolization; sequential images from right to left demonstrate intense prolonged vascular stain associated with lesion.

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**Case #4 – Intramedullary Lesion**

- Low grade, capillary rich neoplasm (WHO I)
- Sporadic (2/3) or associated with von Hippel-Lindau syndrome (1/3)
  - Mean age: 30 y/o, younger if VHL
- Thoracic spine most common, then cervical
- Sensory > motor deficits
- Image whole axis in VHL, assess for multiple

**Case #4 – Hemangioblastoma WHO Grade I**

- Subpial location, posterior aspect of spinal cord
  - Subpial enhancing nodule
  - Intraspinal cyst common
  - Serpentine vascular flow voids if large
  - Syrinx common, +/- long segment edema
- Small percentage extramedullary

**Which of the following is TRUE regarding spinal hemangioblastomas?**

- A: Rarely undergo malignant transformation
- B: Second most common intramedullary neoplasm
- C: Associated VHL gene is located on chromosome 5
- D: 75% of patients with VHL have a spinal hemangioblastoma

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**Case #5 – Intramedullary Lesion**

- Patient: 36-year-old female, with acute episode of waking up with neck pain and pins and needles sensation in her right arm

**Imaging Findings:** T2 heterogeneous lesion, no significant cord edema, T2 hypointense rim on axial imaging; susceptibility artifact on SWI
**Case #5 – Intramedullary Lesion**

*Imaging Findings:* Lesion demonstrates regions of intrinsic T1 hyperintensity with mild regions of enhancement

**Case #5 – Cavernous Malformation**

- Lobulated vascular channels without neural tissue
- Spinal cord *uncommon* location (<5%), thoracic most common
- T1/T2 heterogeneous from blood locules of varying ages ("popcorn") with fluid-fluid levels
- T2-hypointense rim (hemosiderin)
- Edema if recent hemorrhage
- Minimal or no enhancement
- *Blooming* on T2* GRE

**What is the average risk of bleeding of a spinal cavernous malformation?**

- A: 1.4-4.5% per lesion per year
- B: 5.8-10.9% per lesion per year
- C: 15.4-20.1% per lesion per year
- D: 35.8-40.1% per lesion per year

**Case #6 – Intramedullary Lesion**

*Imaging Findings:* Cord expansion with beaded, cystic dilatation of the cord; cord atrophy above level of cystic dilatation

**Case #6**

**INTRAMEDULLARY LESION**

Patient: 61 year old female with remote history of gun shot wound
**Case #6 – Intramedullary Lesion**

*Imaging Findings: No enhancing mass associated with cystic dilatation*

**Case #6 – Syringomyelia**

- Expansion of the spinal cord with *dilated, sacculated, or beaded* cystic activity
- Etiologies: including post-traumatic, Chiari i/ii, spondylosis, neoplasms, arachnoiditis
- Theories:
  1. CSF driven from abnormal subarachnoid space through perivascular spaces → syrinx enlargement
  2. Cord destruction from primary process → cavitation

**Case #6 – Syringomyelia**

- **Hydromyelia** → cystic central cord canal dilatation (lined by ependyma)
- **Syringomyelia** → paracentral cystic cavity that is not contiguous with central cord canal
  - **Syringobulbia** → syrinx in brainstem
- **“Pre-syrinx state”** – reversible spinal cord edema from CSF flow dynamic alteration

**MRI of a Syrinx: Is Contrast Material Always Necessary?**

- 87 patients with syrinx
  - Complete syrinx imaged on T2 and T1 post-contrast
  - Imaging markers independently determined by two board-certified neuroradiologists blinded to clinical and imaging report information
  - **GOAL: assess for mass underlying syrinx on only T2**

- Pathology:
  - 23 mass lesions
  - 11 Chiari malformations
  - 3 chronic spinal cord contusions
  - 50 cases were idiopathic (no underlying cause determined).

**Syrinx diameter**

- Mean associated with mass: 8.1 mm
- Mean associated without mass: 4.1 mm
- **Association with > syrinx size and spinal cord mass**
  - Size threshold of 5 mm
  - No neoplastic lesion present when axial diameter < 5 mm (except cav. mal. & AVM)
**Case #7 – Intramedullary Lesion**

**Imaging Findings:** Patchy long-segment cervical and thoracic T2/STIR cord signal abnormality

**Case #7 – Intramedullary Lesion**

**Imaging Findings:** Regions of abnormal T2 cord signal associated with enhancement; no intrinsic T1 hyperintensity

**Case #7 – Neuromyelitis Optica**

- **Autoimmune inflammatory disorder** → affects myelin of spinal cord and optic nerves → possible targeting of transmembrane channel protein for water transport (aquaorin-4)

- Acute transverse myelopathy spectrum
  - inflammatory (acute transverse myelitis)
  - radiation, paraneoplastic, vascular disease, or idiopathic causes of cord dysfunction

- **Longitudinal, enhancing T2 hyperintensity** which usually involves entire cord cross section + optic nerve enhancement

**Case #7 – Neuromyelitis Optica**

- **Revised diagnostic neurology criteria:** Optic neuritis + myelitis and 2 of 3 supportive criteria (99% sensitive, 90% specific)
  1. Cord lesion → 3 or more segments
  2. NMO-IgG seropositivity
  3. Initial brain MRI non-diagnostic for MS

- **Annual relapse rate of 1.3%**
  - Increased relapse risk during pregnancy

- **IV steroids** first line of treatment
NMO vs. MS vs. ADEM Nuggets

**NMO**
- Longitudinal, cross section
- Outcome worse than MS
- Older age of onset than MS
- Oligoclonal bands in 20-30%
- Cannot be distinguished from ADEM on first attack

**Multiple Sclerosis**
- Typical periventricular white matter lesions
- <1 vertebral segment (more focal), multiple lesions
- Cord periphery
- Oligoclonal bands in 85%
- Relapsing as opposed to ADEM

**ADEM**
- Para/Post-infectious
- Associated encephalopathy
- Monophasic (2-4 weeks)
- Thalamic involvement
- Cranial nerve involvement indicates ADEM over MS

What is the sensitivity and specificity of NMO-IgG seropositivity for NMO?

- A: Sensitivity: 25%, Specificity: 80%
- B: Sensitivity: 57%, Specificity: 75%
- C: Sensitivity: 76%, Specificity: 94%
- D: Sensitivity: 99%, Specificity: 99%

Case #8 – Intramedullary Lesion

**Imaging Findings:** T2/STIR hyperintense expansile mass lesion involving nearly the complete cord cross section, no syrinx

Case #8 – Astrocytoma

- Slow onset myelopathy most commonly
- Most common intramedullary tumor in young adults and children
  - Overall ependymomas are more common accounting for all ages
  - Fusiform cord expansion which commonly spans less than 4 segments
  - Eccentricity in the cord more common
  - Can be multisegmental or holocord

Case #8

**INTRAMEDULLARY LESION**

Patient: 40 year old male who noticed mild left lower extremity weakness, having difficulty raising his left leg to put his socks and ski boots on

**Case #8 – Intramedullary Lesion**

**Imaging Findings:** Very mild enhancement associated with this lesion
Case #8 – Astrocytoma
- Majority of lesions enhance and are T2 hyperintense
- Partial and **mild enhancement** more common
- **Cervical** spine most common
- +/- cyst, less likely to hemorrhage
- 80-90% **low grade** (WHO I or II)

Which of the following is NOT more common in spinal astrocytomas compared to ependymomas?
- A: Astrocytomas are often longer and more eccentric within the cord
- B: Bony remodeling and scoliosis
- C: Ill defined margins
- D: Most common primary spinal neoplasm in kids

Case #9 – Cauda Equina Lesion

Imaging Findings: Symmetric enlargement and enhancement of nerve roots

Imaging Findings: Cranial nerve enhancement in the internal auditory canals and cisternal trigeminal nerve segments

Case #9 – Cauda Equina Lesion

Patient: 47 year old female with history of multiple sclerosis and developing worsening lower extremity weakness
**Case #9 – Chronic inflammatory demyelinating polyneuropathy (CIDP)**

- **Clinical diagnosis**: Sensory/motor neuropathy, usually symmetric, developing over greater than 8 weeks
  - Monophasic, progressive, or relapsing
  - Responsive to steroid therapy
  - EMG abnormal
  - Nerve biopsy if unclear

- **Enlarged, T2 hyperintense nerve roots**
  - (extraforaminal > intradural), peripheral nerves, or plexi → enhancement

- **Lumbar > thoracic/cervical > cranial nerves**

- **Guillain-Barré (AIDP)**: similar nerve root enlargement/enhancement → more acute ascending paralysis onset → more sensory preservation

**Non-imaging findings described with CIDP include the following, EXCEPT?**

- A: “Onion bulb” formation on nerve biopsy
- B: Decrease in F-wave conduction velocities on EMG
- C: Increase in CSF total protein
- D: Mixed sensorimotor neuropathy
- E: All of the above are actually true

**Case #10 – Intradural, Extramedullary Lesion**

Patient: 30 year old male with slowly increasing lower extremity sensory loss

**Imaging Findings**: Ovoid, T1/T2 hyperintense mass with saturation on STIR sequence, no enhancement was noted

**Case #10 – Intradural Lipoma**

- **Presentation**: Slow ascending mono- or paraparesis, spasticity, sensory loss.
  - Symptomatic patients unlikely to improve without intervention.

- May grow dramatically during infancy. Can shrink with weight loss.

- **Intradural location**:
  - **Thoracic (30%) > cervicothoracic (24%) > cervical > lumbosacral**
  - **Dorsal (73%) > lateral/anterolateral > anterior**
**Case #10 – Intradural Lipoma**

- T1/T2 hyperintense, fat suppression, no enhancement
- +/- dysraphism
- Terminal lipoma $\rightarrow$ associated with distal cord/foramen insertion $\rightarrow$ may extend into subq fat through defect $\rightarrow$ cord tethering and syrinx

**Unknown Case #1 – Intramedullary Lesion**

*Imaging Findings:* Cord signal abnormality at T1 and T2 levels with associated contour abnormality of the dorsal aspect of the cord

**Unknown Case – Arachnoid Web**

- Arachnoid webs: *intradural, extramedullary transverse bands* of tissue that can extend to spinal cord pial surface
- Presence of dorsal subarachnoid space web $\rightarrow$ *alteration of CSF flow dynamics* $\rightarrow$ pre-syrinx state versus syringomyelia

**Unknown Case – Arachnoid Web**

- Reardon et al. (AJNR May 2013)
- 14 patients (ages 31-67 years) described with “scalpel sign” on MRI or myelogram
  - 10/13 patients who underwent MRI showed T2 cord hyperintensity
  - 7/13 patients demonstrated syringomyelia
  - 7 patients underwent surgery $\rightarrow$ 5 demonstrated arachnoid web as cause of dorsal indentation on pre-operative imaging $\rightarrow$ all patients symptoms improved post-operatively

**Unknown Case #2**

*INTRADURAL, EXTRAMEDULLARY LESION*

Patient: 31 year old female with history of chronic pain secondary to pelvic chondrosarcoma
**Unknown Case #2 – Intradural, Extramedullary Lesion**

*Intrathecal catheter tip granuloma*: T2 hypointense enhancing focus at the tip of the intrathecal catheter; inflammatory granulomas can cause pump-catheter dysfunction (increased pain/withdrawal).

**Unknown Case #3 – Extradural Lesion**

Patient: 25 year old male with sporadic lower back pain

**Unknown Case #3 – Osteoid Osteoma**

- Benign osteoid-producing tumor
  - < 1.5 cm in size (larger lesion = osteoblastoma)
- 10% occur in spine, neural arch; lumbar > cervical > thoracic
- Can cause focal scoliosis with concavity towards the side of the tumor
- *Night pain* → treatment *aspirin* or NSAIDS

**Iatrogenic Companion Case**

*History is key*

Presented to outside facility with post-lumbar puncture headache

- Received epidural blood patch
- T2 hypointense, T1 hyperintense nonenhancing intrathecal mass

Presumably an iatrogenic thrombus → risk of arachnoiditis

**Unknown Case #3 – Osteoid Osteoma**

- **Central nidus:**
  - Usually lucent on CT with variable ossification
  - MRI: low T1 signal, variable T2 signal, enhancement
- **Reactive zone:**
  - Dense *sclerosis and edema* surrounding the nidus
  - High STIR/T2 signal, low T1 signal, enhances
  - May have soft tissue mass or pleural thickening/effusion if near the lung
THANK YOU