Intradural Spinal Neoplasms

John K. Birknes, M.D.
Department of Neurosurgery
Thomas Jefferson University Hospital
Spinal Cord Anatomy

- 31 pairs of spinal nerves
  - 8 cervical
  - 12 thoracic
  - 5 lumbar
  - 5 sacral
  - 1 coccygeal
- C & L enlargements
- Conus tapers to ~L1/2
- Filum terminale- attaches to dorsum of 1st coccygeal vertebra
Spinal Cord Anatomy

- Central gray matter
  - Neuronal cell bodies
  - Supporting structures
- Prominent ventral and dorsal components with commissure between halves
- White matter tracts encircle gray matter
Anatomy-Meninges

- **Dura:** closest to VB
  - single layer, contrast with brain
- **Delicate Arachnoid**
- **Pia:** contacts cord
  - attaches cord to dura via dentate ligaments.
Vert a. gives rise to:
- 1 Ant spinal a. &
- 2 Post Spinal a.

Lower 1/3 of C-sp $\rightarrow$ radicular aa. off
- Vert a.
- Ascending cervical a.
- Deep cervical a.
Anatomy-Spinal Cord Vasculature

- Below C-spine continuous anastomoses with radicular arteries
- Aorta → intercostal a. → spinal a. → ant & post radicular a.
- Central branches off ASA alternate sides of cord
Anatomy-Spinal Cord Vasculature

- Artery of Adamkiewicz
- Left T11 (T9-12) radicular art.
- Major blood supply to lower T and L spine
Anatomy-Spinal Cord Vasculature

- **Batson’s plexus:**
  - epidural veins
  - no valves

- **Multiple anastamoses w/**
  - Azygous system
  - IVC
  - Pelvic plexus
  - Prostatic plexus
History

- Sir Victor Horsley (1857-1916)
- 1887: 1st successful resection of intradural spinal neoplasm
  - Meningioma
- 1911: 1st successful resection of intramedullary tumor
  - Charles Elsberg
  - 2 stage procedure → myelotomy, 1wk later remove extruded tumor
Classification: Intradural

- **Extramedullary: ~90%**
  - in subarachnoid space
  - Schwannoma
  - Neurofibroma
  - Meningioma
    - >90% nerve sheath tumor or meningioma
  - Subarachnoid mets (only 4% of spinal mets) or “drop mets”
  - Peds: Dermoid/Epidermoid

- **Intramedullary: ~10%**
  - within spinal cord
  - Ependymoma
  - Astrocytoma
  - Hemangioblastoma
  - Mets (only 2% of spinal mets)
Extradural: Nerve Sheath Tumors

- **Schwannomas**
  - Together ~1/3 of intradural neoplasms
  - Slightly more common
  - Dorsal root
  - Neurofibromatosis (NF-2)
  - Encapsulated
  - Displace nerve
  - Schwann cells
  - Malignancy v. rare

- **Neurofibromas**
  - Together ~1/3 of intradural neoplasms
  - Slightly less common
  - Dorsal root
  - Neurofibromatosis (NF-1)
  - Unencapsulated
  - Entangle nerve- elongate
  - Schwann cells & fibroblasts
  - 5-10% pts w/ NF malignant NST (≤ 1 yr survival)
    - XRT implicated
    - Usually plexiform
Nerve Sheath Tumors

- Extended growth period → osseous remodeling
- Widened neural foramina
- VB scalloping
- Increased intrapedicular distance
- Dumbbell shape (may have extradural component up to 15%)
Nerve Sheath Tumor

- T1WI: iso/hypo-intense
- T2WI: hyperintense → increased water content
- Homogeneous enhancement
- “Target sign”: T2 or T1 with gad
  - hyperintense rim, hypo center
  - Neurofibromas w/ peripheral myxomatous & central fibrocollagenous tissue
- 40% schwannomas cystic
Clinical Presentation: NST

- Middle aged adults (male~female prevalence)
- Uniform distribution in spine
- Symptoms similar to HNP
  - Pain and radiculopathies
  - Paresthesias
  - Weakness
  - Myelopathy
Nerve Sheath Tumor-Schwannoma

- 31 nerve roots sacrificed
  - C5-T1 (n=14)
  - L3-S1 (n=17)
- 23% w/ post-op motor or sensory deficit (7/31)
- 6 cases neurofibroma like characteristics
  - No deficit
- “Spinal roots giving rise to schwannomas are frequently nonfunctional at the time of surgery.”

Kim et al., J. Neurosurgery, 1989
Spinal Meningioma

- 25-30% of spinal tumors
- 1:8 spinal to intracranial
- Most dorsal or lateral to cord
- Solitary (only 1-2% multiple)
- $\leq 5\%$ extradural or both
Spinal Meningioma

- 80% in T-spine (15% C-spine)
- Rare bone remodeling
- Isointense to cord T1 & T2, bright homogeneous enhancement- “dural tail”
Clinical Presentation: Spinal Meningioma

- Middle-aged women (80% women)
- Motor deficit: 90%
- Sensory deficit: 60%
- **Pain:** 50-70% (diffuse localized over region or radicular)
- Sphincter dysfunction~50%
Spinal Meningioma

- N=174 (143 women, 31 men)
- 96.5% Gross total resection
- Surgical mortality $\rightarrow$ 1%
- Recurrence rate $\rightarrow$ 6%
- 92% good-excellent post-op neuro status
  - long-term follow-up (avg. 15 yrs)
  - 70% pre-op
- Even anteriorly positioned tumors were resected via posterior approach (sectioning dentate lig.)

Solero et al., Neurosurgery, 1989
Intramedullary Neoplasms

- 2% of adult & 10% of pediatric CNS neoplasms
- Adults → 50-70% Ependymomas
- Peds → 55-65% Astrocytomas
- Hemangioblastomas → 5%
- Miscellaneous → 5%
  - (gangliogliomas, oligodendroglialomas, paragangliomas)
- Mets → v. rare (2% of spinal mets)
Intramedullary Glial Neoplasms

- **Ependymoma**
  - Cellular (C-sp or anywhere;)
  - Myxopapillary (conus; slight ♀)
  - Mean age: 43 y/o
  - Cystic degeneration (>50%) w/ hemorrhage at margins
  - Diffuse cord enlargement multiple levels
  - Sharp deliniation from cord good plane
  - Homogeneous enhancement

- **Astrocytoma**
  - Low-grade: fibrillary
  - AA & GBM (10% peds & 20% adults)
  - Mean age: 21 y/o
  - Cystic as well, less likely to hemorrhage
  - Diffuse cord enlargement multiple levels
  - More infiltrative often poor plane
  - Heterogeneous enhancement
19-year-old presented with numbness and finger clumsiness.

a. Coronal T1WI demonstrates a rostral cyst & expansile cervical tumor.

b. Sagittal T1WI demonstrates the enhancing tumor from C2–C5.

c. The axial T1WI w/ gad: characteristic central location of this tumor type
Intramedullary Glial Neoplasms: Clinical Presentation

- Pain present over extended time
  - Often localized to spinal segment
  - Worse @ night/awakening ➔ hypercarbia venous engorgement
- Gait abnormalities (spastic paresis or ataxia)
- Sensory changes
- High-grade astrocytomas sx for mean of 4-7 mos vs low-grade mean sx duration 41 mos.
Intramedullary Glial Neoplasms

- >90% 5-yr survival
- Goal: bx & prevent further neuro deficit
- N=239 low-grade spinal neoplasms
- Neurologic outcome
  - 40% improved
  - 50% unchanged
  - 10% worsened

Brotchi et al., Contemp Neurosurg., 1999
Intramedullary Glial Neoplasms

- N=69 (intramedullary spinal cord tumors)
- Neuro outcome (mean f/u of 54 mos.)
  - 20% improved
  - 50% unchanged
  - 30% worsened
  - Improvement @ 6-18 mos (dosal columns longest)
  - Preop neuro fxn best prognostic indicator outcome
- 5/6 high grade astrocytomas died by 9-16 mos
  - 1 alive @ 10 mos but with progression

Cristante & Hermann, Neurosurg, 1994
Intramedullary Ependymoma

- N=23, intramedullary ependymoma
  - 8 reoperation; only 4 conus, 0 filum
  - GTR in all cases
- Mean f/u of 62 mos (6 mos.-13yrs)
  - No pts lost to f/u
  - No recurrence
  - 8 pts improved
  - 12 pts unchanged
  - 3 pts deteriorated
- With GTR no role for adjuvant Tx

McCormick et al., J of Neurosurg, 1990
Intramedullary Astrocytoma

- N=25 intramedullary astrocytomas
  - 6 pts with high-grade → 5 died (4-23 mos post-op)
  - 2 pts with advanced neuro disability preop died from medical complications
- 17 pts w/ mean f/u of 50 mos. (16-89 mos)
  - Fxn: 3 pts improved, 12 unchged, 2 worse
  - 15 pts: no tumor recurrence
  - 2 pts: small residual neoplasm without progression
- Surgery beneficial in low-grade but not AA

Epstein et al., J. Neurosurgery, 1992
Adjuvant Therapy

- **Ependymoma**
  - Follow w/ serial MRI if GTR
  - Local XRT ~50 Gy if subtotal resexn or disseminated dz
  - No role for chemo

- **Astrocytoma**
  - Follow w/ serial MRI if GTR, low-grade & well-circumscribed
  - If high-grade of diffuse:
    - 50 Gy local XRT in 30 fractions
    - Chemo: Temozolomide or PCV (procarbozine/CCNU/vincristine)

Stereotactic spinal radiosurgery yet to be defined
Intramedullary Hemangioblastoma

- ~1/3 of pts with VHL
- 80% symptomatic by 5th decade
- Presentation similar to glial neoplasm
  - Rarely present w/ sudden deficit from hemorrhage
Intramedullary Hemangioblastoma

- Bright homogeneous enhancement
- No more than 1 VB in length
- 80% w/ cystic tumor nodule; serpiginous vessels
- A-gram & embo possible prior to surgery
Thank You
MRI in a 19-year-old male who presented with numbness and finger clumsiness. Histological diagnosis was an ependymoma.

**a.** Coronal T1-weighted MRI demonstrates a rostral cyst and expansile cervical tumor.

**b.** Sagittal T1-weighted MRI demonstrates the enhancing tumor from C2–C5.

**c.** The axial T1-weighted images with contrast demonstrate the characteristic central location of this tumor type.
Figure 1A and B. (A) MRI of the cervical spine, performed first, shows an area of irregular enhancement within the cord at C2-3, with an associated multiloculated syrinx extending in both cranial and caudal directions. The very intense enhancement of the lesion marks it as a hemangioblastoma and prompted a spinal survey. (B) MRI of the thoracic spine shows a second lesion at T9-10, containing a flow void and also showing bright enhancement. Cystic change within the cord extends all the way from the cervical lesion to the thoracic tumor.
Hemangioblastoma

- *Figure 1C.* The intraoperative appearance of the cervical cord gives a classic picture of hemangioblastoma *in situ*, with engorged, numerous arteries and draining veins leading to and from a well-circumscribed and highly vascular tumor visible at the pial surface. This tumor, as well as its thoracic counterpart, was excised completely and its suspected identity confirmed. She remains well (with asymptomatic pancreatic cysts) six years after diagnosis.
FIGURE 1. Schematic drawing (A) and representative T2-weighted sagittal magnetic resonance image (B) demonstrate a strictly intradural tumor (Group 1 tumor).

FIGURE 2. Schematic drawing (A) and representative gadolinium-enhanced T1-weighted coronal magnetic resonance image (B) demonstrate a tumor with both intradural and extradural components within the spinal canal (Group 2 tumor).
FIGURE 3. Schematic drawing (A) and representative gadolinium-enhanced T1-weighted coronal magnetic resonance image (B) demonstrate a strictly extradural tumor within the spinal canal (Group 3 tumor).

FIGURE 4. Schematic drawing (A) and representative gadolinium-enhanced T1-weighted axial (B) and coronal (C) magnetic resonance images demonstrate a strictly extradural tumor extending through the intervertebral foramen (Group 4 tumor).
FIGURE 5. Schematic drawing (A) and representative gadolinium-enhanced T1-weighted coronal magnetic resonance image (B) demonstrate a tumor with both intradural and extradural components extending through the intervertebral foramen (Group 5 tumor). Arrow indicates an intradural component.
Bar graph showing classification of spinal nerve sheath tumors at the various spinal levels.
Vert a. gives rise to 1 Ant spinal a. & 2 Post Spinal a. (see JSH anatomy talk)

Blood from verts supply cervical spine, but below is continuous anastomoses with radicular arteries
Intradural-Extramedullary

- **Nerve Sheath Tumors:**
  - Schwannomas slightly more common than Neurofibromas
  - Dorsal root (sensory)
  - 35-45% have Neurofibromatosis: Neurofibromas w/ NF-1 & Schwannomas w/ NF-2 (p676 Wilkins)
  - Schwann cells vs. Schwann cells & fibroblasts
  - Displace nerve vs. Entangle nerve fascicles
  - Malignant nerve sheath tumor degeneration: increased incidence with NF