Intramedullary Spinal Tumors

PATHOLOGY

- 15-20% of intracranial tumors
- Spinal tumors:
  - extramedullary - 55%
  - intramedullary - 40%
  - intramedullary - 5-10%
  - in children, 50% intradural lesions are extramedullary, 50% - intramedullary.
Intramedullary tumors:
  - a) Stills of all spinal tumors
  - b) 2-4% of brain tumors (6-10% of pediatric brain tumors)
  - intramedullary tumors are more common in children, extramedullary tumors - in adults.
  - 5% neurofibromatosis patients develop multiple spinal cord tumors.
CNS tumors:

- < 15-20% are malignant, > 90% are benign - subject to potential resection.
- extend over many spinal cord segments* - signs and symptoms are more variable (than those of extramedullary tumors).
- 70% are associated with cysts (may produce own symptoms of spinal dysfunction):
  - a) intratumoral cysts (wall consists of tumor)
  - b) peritumoral or capping cysts - cone-shaped glial-lined cavities extend above and below tumor for limited number of spinal segments.
- syringomyelia (most frequent with hemangioblastoma) - indistinguishable from other forms of syringomyelia.
- leptomeningeal dissemination (drop metastases) occurs in 58% of high-grade (malignant) tumors; uncommon in low-grade tumors.

LOCATION

- anywhere from cervicomedullary junction to filum terminale.
- 50% in thoracic cord (because of relative length of this area); 30% in lumbosacral cord.

ETOLOGY

Strikingly different from brain tumors!

1. Ependymoma (56-70% in adults; only 30% in children)
2. Astrocytoma (29%; in children 40-70%; 90% at age < 1 yo); Pilocytic astrocytoma, other low-grade astrocytomas, anaplastic astrocytoma, glioblastoma
3. Hemangioblastoma (3-5%)
4. Oligodendroglioma (3%)
5. Developmental tumors (3%):
  1) dermoid
INTRAMEDULLARY SPINAL TUMORS

2) epidermoid
3) teratoma

6. Lipoma (2%)  

7. Others (4%): 
1) subependymoma 
2) ganglioglioma 
3) intramedullary schwannoma 
4) neurofibroma 
5) metastases (uncommon, < 2%) – most commonly from small cell lung carcinoma

- ASTROCYTOMAS and EPENDYMOMAS are more common in patients with neurofibromatosis type 2.

Conus tumors: 
1) myxopapillary ependymoma 
2) ganglioglioma

CLINICAL FEATURES

Progressive myelopathy (mimics syringomyelia) – Central Cord Syndrome

In most instances, clinical presentation does not indicate if tumor is EXTRADURAL or INTRADURAL

- slow-growing nature - symptoms precede diagnosis by = 2 years (vs. extramedullary tumors – shorter period).
- neurologic manifestations commonly begin unilaterally (full-blown Brown-Sequard syndrome is rare), becoming bilateral when tumor is quite large.
- dull, aching neck/back pain (from level of lesion; local or radiating) often is earliest symptom! – characteristically at night when patient is supine (related to venous outflow disturbance and/or decrease of endogenous glucocorticoids, may be increased by Valsalva (coughing or sneezing). – pain is usually less prominent than of extramedullary tumor.

- myelopathy with progressive paraparesis predominates early (LMN* → UMN); N.B. kids may manifest as DEXTROscoliosis or TORICOLLIS.

- dissociated sensory loss with sacral sparing, sphincter dysfunction, trophic changes.
- hydrocephalus (15%, esp. in malignant tumors) – due to increased CSF viscosity from elevated protein content.

DIAGNOSIS

IMAGING

Some tumors occur in multiple areas - image entire neuraxis (e.g. HEMANGIOBLASTOMA).

Plain X-rays - insensitive and nonspecific:
1) spinal canal widening (around slowly expanding tumor) 
2) posterior scalloping of vertebral bodies (on lateral radiographs) 
3) medial erosion of pedicles → widening of interpedicular distance (on AP radiographs) 
4) kyphoscoliosis, dextroscoliosis (in children)

Contrast-enhanced MRI - very sensitive for tumors!
- fusiform enlargement of spinal cord over several levels (vs. inflammatory lesions - normal or minimal increase in cord size).
- most tumors are isointense or slightly hypointense. 
- great majority of gliomas enhance at least partially (vs. brain gliomas).
- tumor-associated syrinx may be seen.

CT myelography - used when MRI is not available:
1) multisegmental smooth spinal cord widening (± exophytic outgrowth) → narrowed subarachnoid space on both sides of cord. 
2) block of contrast flow (50-90%) 
3) enlarged vessels on cord surface (80% HEMANGIOBLASTOMAS, 10% EPENDYMOMAS).

Spinal angiography - only if HEMANGIOBLASTOMAS is suggested; see below

- Rapid decline in leg function 
- MRI not very definitive for tumor
- GET THE ANGIO

CTM or AP cervical myelogram - diffuse widening of cervical cord, bilateral attachment of centrodural fluid space (arrowshead).

Teratomas of spinal cord - well delineated on T1 MRI without contrast - high intensity component of tumor (arrows) is fat.

LUMBAR PUNCTURE

- not indicated (unless patient is being evaluated for leptomeningeal spread)
CSF in spinal block - protein ↑↑↑ (Froin syndrome), xanthochromia (due to high protein content).

TREATMENT

Remains controversial.

- in selected situations, watchful waiting can be considered (e.g. high surgical risk and/or mild neurologic dysfunction).
- high-dose steroid (Dexamethasone, 50 mg IV → 10 mg q6h) may improve neurologic function transiently.

SURGERY

Surgical extirpation is treatment of choice for benign tumors! (cures have been reported only after complete surgical resections); no aggressive surgery for high-grade tumors!

- Total removal with preservation of neurologic function?

Neurological deficits preop correlate with poor outcome postop – do not delay surgery!

PROCEDURE

- see p. Op200+55

POSTOPERATIVE

- ICU for 24–48 hours.
- flat for 1-3 days (esp. lower thoracic – lumbar tumors).
- cervical tumors → continued mechanical ventilation in immediate postoperative period.
- prophylaxis for deep vein thrombosis.
- pain – majority of patients have increased deficit during immediate postoperative period (edema from surgical manipulation, blood flow alteration) – typically transient and must return to baseline within 3-6 months.
- short course of tapering steroids may be used to help offset any cord injury → rapid steroid tapering (steroids inhibit wound healing - predispose to CSF leakage).
- hematomas is recognized by immediate progressive deterioration of nervous function → MRI/CT confirmation → urgent reexploration

- typically, temporary sensory disturbances due to posterior column retraction.
- ambulation is recommended after 1-3 days of bedrest in flat.
- CSF leakage* should be treated aggressively – suction closure, collodion, lumbar drainage, reoperation for closure.
- frequently as poor healing of incision
- new-onset urinary retention may require prolonged bladder catheterization.
- bowel stimulation regimen may be necessary for new abnormalities.
- early physical / occupational therapy.
- MRI day after surgery (completeness of resection); progradal tumor:
  a) repeat resection (for ependymoma)
  b) radiotherapy (for astrocytoma)
  c) watchful waiting (e.g. developmental tumors, lipomas - prolonged survival despite residual tumor).

Postoperative pain:

- Somatic (acute) pain - results from manipulation of nerve roots (e.g. ligation of dorsal nerve root due to bleeding from radicular vessel; better approach - sharp incision of nerve roots with focal cauterization of any bleeding); H: steroids are very helpful.
- Central (chronic) pain - results from resection of intramedullary tumors: gnawing, sometimes burning, persistent pain; can be triggered by light touch and may extend well beyond area of stimulation; does not respond well to drugs or stimulators.

FOLLOW-UP

- (serial neurologic examinations and MRI):
  - consider, in select cases, maintaining the patient in prone position to avoid pial dural scarring.
  - tumor recurrence → image entire neuraxis (even benign ependymomas may change their growth characteristics and produce seeding – recurrent tumor after surgery is first choice!)
  - poor efficacy – Ependymoma (surgically excised ependymomas need not undergo subsequent radiotherapy!)
  - doxorubicin 50 Gy in daily 1.5-2 Gy fractions - this dose is not curative (some report doses > 50 Gy reduce local failure rates):
    - higher doses can be used for lesions involving only causa equina or if irreversible complete transverse myelopathies already has occurred.
  - margin 2-3 cm or two vertebral bodies above and below lesion.
  - most important adverse effects:
    1) acute and delayed myelopathy
    2) diminished skeletal growth in young children
    3) increased difficulty with subsequent surgical tumor removal (important if radiotherapy does not control growth of lesion).
  - SRS may have role (esp. for malignant tumors); consider laser ablation – disconnection procedure – disconnects cord from tumor so radiation becomes possible.
  - although similar somatosensory control may be achieved over short term when compared with surgical resection, recurrence and malignant tumor transformation have been observed after radiotherapy!
  - advent of proton beam.

RADIONERTHERAPY

- primary treatment for:
  1) malignant lesions (e.g. ANAPLASTIC ASTROCYTOMAS, GLOBALGAMAS – surgical tumor removal has no value - survival is < 2 years).
  2) inoperable tumor

- may be useful for:
  1) residual tumor after surgery (e.g. most ASTROCYTOMAS)
  2) recurrent tumor (repeat surgery is first choice!)
  3) poor efficacy – Ependymoma (surgically excised ependymomas need not undergo subsequent radiotherapy!)

- margin 2-3 cm or two vertebral bodies above and below lesion.

CHEMOTHERAPY
Motor and autonomic functions continue to improved up to 24 mos postop (sensory function plateaus

8. 7. 6. 5. 4. 2.

Prognostic factors:

1. Histology - aggressive tumors have poor prognosis despite treatment - radical surgery can lead to severe neurologic impairment.

Tumor histology is the most important predictor of neurological outcome because it predicts resectability and recurrence.


Gross total resection was achieved in:

- ependymoma - 90.9%
- hemangioblastoma - 91.7%
- astrocytoma - 14.3% (all those were pilocytic astrocytomas; none of the grade II, III, or IV astrocytic tumors had GTR).

At mean follow-up of 41.8 months, recurrences were observed:

- ependymoma - 7.3% cases
- hemangioblastoma - no recurrences
- astrocytoma - 47.6% cases

At time of last follow-up, neurological status was:

- ependymoma - 20% patients improved, 69% remained the same, 10.9% worsened.
- hemangioblastoma - 8.3% improved, 91.7% remained the same. No patient worsened!
- astrocytoma - 4.8% improved, 47.6% remained the same, 47.6% worsened.

2. Preoperative deficit - those with advanced neurologic compromise generally have no worthwhile improvement (need for early intervention and close follow-up).

3. Completeness of resection

Historically, intracavitary tumor resection has been based on whether plane dissection can be identified, which is often dependent on tumor histology. Ependymomas typically demonstrate clear tumor and spinal cord interface, whereas astrocytomas exhibit more infiltrative pathology.

4. Age > 60 yr is negative prognostic factor.

5. Lesion location (higher morbidity is associated with surgery of upper thoracic and conus lesions).

6. Size of lesion - tumors spanning several levels may produce cord compression (rather than infiltration; requires extensive dissection of spinal cord in order to expose tumor).

7. Arachnoid scarring, cord atrophy - negative prognostic factors for ependymomas.

There is a potential for late scarring of pia to dura with a tension injury to the spinal cord and loss of function.

8. Syrinx - suggests noninfiltrative lesion (better prognosis).

SPECIFIC TUMOR TYPES

EPENDYMOAMA

- arise from ependymal cells lining central canal; see p. Onc 14

50% in conus medullaris (myxopapillary ependymoma - Acrilan blue stain for myelin).

characteristically hypovascular, cystic degeneration with hemorrhage at margins (“hemosiderin cap” on MRI), well circumscribed, noninfiltrative (cord compression rather than infiltration; complete resection → prolonged survival).

mean age at presentation - 43 years (myxopapillary variant - 21 yrs but reported in 3 month-old to 86 year-old).

pregnancy or trauma may precipitate FISCHER'S syndrome (acute subarachnoid hemorrhage with sciatica).

slow growth - likely to result in bony remodeling.

- clear cleavage plane - complete excision is possible! - chemotherapy has no role.

radiotherapy has role:

- radical surgery after chemotherapy can delay progression.

myxopapillary ependymoma was associated with improved progression-free survival and local control.

myxopapillary ependymoma in grade I but big tumors may seed CSF space - try to resect en bloc + adjuvant panpapillary radiation (current techniques help to spare bone marrow; chemotherapy has no established role).
Ependymoma of distal spinal cord (A – T2; B – contrast T1) - large, fusiform, intramedullary enhancing tumor associated with extensive proximal syrinx.

Contrast T1-MRI - ependymoma with small capping cyst (arrow):

Myxopapillary ependymoma (MRI) - lobulated mass extending down from L4 level.

A. Operative photograph - myelotomy exposes dorsal surface of tumor; note clear demarcation of tumor from surrounding spinal cord.

B. Tumor specimen that has been completely removed.
INTRAMEDULLARY SPINAL TUMORS

Myxopapillary ependymoma - cells around papillations that have myxoid connective tissue core:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD)

Myxopapillary ependymoma - streaming vessels with arrangements of tumor cells around them; cytoplasmic round vacuoles are filled with mucinous contents:

**Intramedullary Spinal Tumors**

**Astrocytoma**
- More common in children (most common intramedullary tumor in pediatric age group!)
- Average length - 7 vertebral-body segments.
- Sometimes associated with microcysts or syrinxes.
- Less hemosiderin, more peritumoral edema, more heterogeneous enhancement (cf. ependymoma).
- Pilocytic astrocytoma is well differentiated with definable surgical plane – possible to remove surgically.
- Other low-grade astrocytomas - infiltrative and impossible to remove grossly (but residual tumor often has indolent course).
- Anaplastic astrocytoma, glioblastoma are rare (< 10-20%), may seed CSF; surgery does not improve course! – death within 2 years.

Currently, no satisfactory modality is available for malignant astrocytomas!

A. T1-MRI - expansion of upper cervical cord (arrows) by mass lesion in cervicomedullary junction.
B. T2-MRI - high-signal-intensity intramedullary mass expanding upper cervical cord (arrows).
C. Contrast T1-MRI - irregular peripheral enhancement (arrows).

A. T1-MRI - large cyst in lower cervical cord and smaller cyst extending up into medulla; intervening spinal cord is slightly enlarged but demonstrates no signal abnormality.
B. Contrast T1-MRI - enhancing tumor at C2-3 level.
Intramedullary Spinal Tumors

High-grade astrocytoma (contrast T1-MRI) - complex solid and cystic tumor of distal spinal cord with areas of intense enhancement; slight expansion of bony spinal canal.

Glioblastoma (T1-MRI) - marked cord expansion by irregular mixed signal mass containing areas of recent hemorrhage (arrow).

T1-MRI - anaplastic astrocytoma of upper thoracic cord (arrow); note cystic change.

Pilocytic astrocytoma (contrast T1-MRI).
**INTRAMEDULLARY SPINAL TUMORS**

GBM – rare appearance (looks more benign on MRI):

OLIGODENDROGLIOMA

**DEVELOPMENTAL TUMORS [DERMOID, EPIDERMOID, TERATOMA]**

(3%) - slow-growing neoplasms with lumbar predominance (esp. conus medullaris).

- can be associated with spinal dysraphism and dermal sinus tract
- may also be acquired – due to lumbar puncture with needle without stylet.
- dense capsule may preclude complete removal (tumor debris may cause early recurrence).
- avoid operative spilling of irritating (epi)dermoid content (→ inflammation, arachnoiditis, adhesions).

- 20% may occur in multiple locations!
- SAH is classic presentation!
- mean age at presentation – 4th decade.
- associated with von Hippel-Lindau disease in 30-80% cases.
- cyst with tumor nodule (50-70%).
- 20% may occur in multiple locations!
- enhances strongly with MRI contrast.

**TUMORS**


**HEMANGIOBLASTOMA**

also for general features see p. Onc24 >>

- mean age at presentation – 4th decade.
- associated with von Hippel-Lindau disease in 30-80% cases.
- cyst with tumor nodule (50-70%).
- 20% may occur in multiple locations!
- nearly always involve POSTERIOR COLUMNS – simplified surgical approach.
**INTRAMEDULLARY SPINAL TUMORS**

- **angiography** often provides definitive diagnosis (but usually is not necessary preliminary to operative treatment):  
  1) homogeneous, well-circumscribed dense capillary blush  
  2) one or two supplying arteries are slightly enlarged  
  3) enlarged (or normal sized) draining veins opacify only little earlier than normal.

**Treatment**

- can be cured by **surgical excision**
  - surgical principles similar to those used in treating AVMs - feeding arteries are coagulated, and tumor is dissected and **removed en bloc** (do not remove in piecemeal fashion - significant bleeding may ensue!).
  - neuroradiology has low value - surgery should be guided by tissue plane and tumor has to come out.
- **LINAC radiation therapy** has also been proposed as a treatment modality, with a great deal of success.
- **BEVACIZUMAB** - case report described its use in a patient with a surgically unresectable cervical cord hemangioblastoma, showing significant tumor regression and clinical improvement.

**LIPOMA**

- not true neoplasm!  
  - often associated with **spinal dysraphism** and cutaneous abnormalities (nevi, dimples, hyperpigmentation, hypertrichosis, capillary angiomas, midline hairy patches, subcutaneous lipomas)  
  - presents in first 3 decades of life (when fat is being deposited).  
  - T1 – very hyperintense signal, T2 – hypointense (?) signal.  
  - loss of total body fat may be necessary to reduce tumor mass.  
  - fibrous adhesions to cord, no distinct cleavage plane make total removal difficult.  
  - **N.B.** removal is not goal of surgery (CO2 laser is particularly useful).

**SUBEPENDYMOMA**

**GANGLIOGLIOMA**

**INTRAMEDULLARY SCHWANNOMA**

**NEUROFIBROMA**

Dilated neuroforamen:

**METASTASES**

(unusual)  
- 61% have multiple CNS metastases.  
- myelogram may be normal (42%).  
- most common sources - lung cancer, breast cancer.  
- surgery is recommended for solitary metastasis and limited cancer (can be completely resected through definitive cleavage plane).

**MELANOMA**

Metastatic melanoma (cues [to differentiate from nerve sheath tumor] – hyperintense on T1, nondilated neuroforamen):
Intramedullary Spinal Tumors

BIBLIOGRAPHY for ch. “Neuro-Oncology” — follow this LINK >>

Viktor’s Notes™ for the Neurosurgery Resident
Please visit website at www.NeurosurgeryResident.net