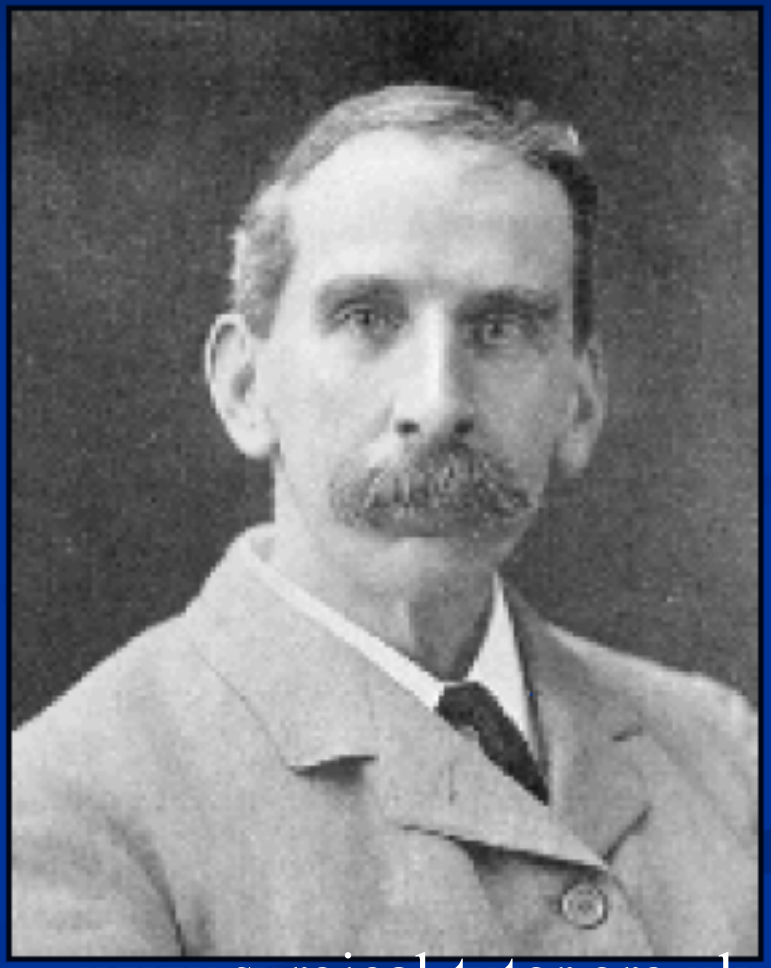
MANAGEMENT OF INTRAMEDULLARY SPINAL CORD TUMORS

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



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History (contd...)

- 1919 - Dandy introduced air-contrast myelography.
- 1940 - Greenwald introduced bipolar coagulation.
- 1964 - Kurze introduced the operating microscope.
- 1967 - Greenwood published a large series detailing successfully removed tumors.
- 1980s-MRI was introduced
- 1990 - McCormick “large surgical series demonstrating excellent long-term outcomes for surgery of ependymomas” and established a clinical grading system.

Spinal Lesions

INTRAMEDULLARY

- Ependymoma
- Astrocytoma
- Hemangioblastoma

INTRADURAL EXTRAMEDULLARY

- Nerve sheath tumor: Schwannoma, neurofibroma
- Meningioma
- Myxopapillary ependymoma
- Lipoma/Dermoid/Epidermoid

EXTRADURAL

- Degenerative: Synovial cyst, disc
- Infection/abscess

Primary spinal tumors - location

Location	*Pediatric*	*Adult*
➢Intramedullary	40%	20%
➢Intradural Extramedullary	10%	60%
➢Extradural	50%	20%

Epidemiology

- Much less common
- 2-4% of all intrinsic CNS tumors
- Astrocytoma and ependymoma - 80-90% of all intramedullary tumors
- Nerve sheath tumors (neurofibroma and schwannoma) and meningiomas - 80% of intradural extramedullary tumors

Ddx of Intramedullary tumors

Pathology	% in adults	Age	Location
Astrocytoma	30-35%	10's – 50's	T>C>LS
Ependymoma	65%	20's – 60's	>50%Conus or Filum
Hemangioblastoma	1-3% (25% have Von Hippel-Lindau)	30's – 60's	T
Lipoma	Rare (unless with dysraphism)		
Epidermoid - Dermoid	Uncommon		
Glioblastoma	1.5%		
Metastases	Rare	50's -	

Others: oligodendroglioma, ganglioglioma, schwannoma, melanoma, teratoma, neuroenteric cyst, cavernous angioma

Symptoms

- Symptoms not specific to spinal cord tumors and may be present in any myelopathic process.
- Because of slow-growth, symptoms precede tumor discovery an average of 2 years.
- Pain often is the earliest symptom, characteristically occurring at night when the patient is supine.
- Pain is typically local at tumor level , but may radiate

Symptoms (contd..)

- Progressive weakness may occur in the arms (Cervical Tx) or legs (C/T OR Conus Tx).
- Patients may have impaired bowel or bladder function.
- Rarely Symptoms of subarachnoid hemorrhage may be present.
- Abrupt deterioration may occur with intratumoral hge.

Examination

- Combination of upper and lower motor neuron signs
- Spine tenderness
- Stiffening of gait
- Trophic changes of extremity
- Sensory loss
- Hyperreflexia
- clonus
- Scoliosis
- torticollis (generally in children).

Imaging

Modality	Features
Plain Film	Increased inter pedicular distance, pedicle thinning (occasional erosion), scalloping of vertebral bodies, scoliosis
Myelogram + CT	Widened spinal cord , dye block
MRI	T1- iso or ↓ signal lesion ± cysts, most gadolinium enhancing, edema, T2 - ↑ signal edema
Angiogram	May be useful in distinguishing hemangioblastoma from AVM

OTHER TESTS :

Electrophysiologic testing

- Not useful in the diagnosis and preoperative management
- More value in monitoring cord function intraop.

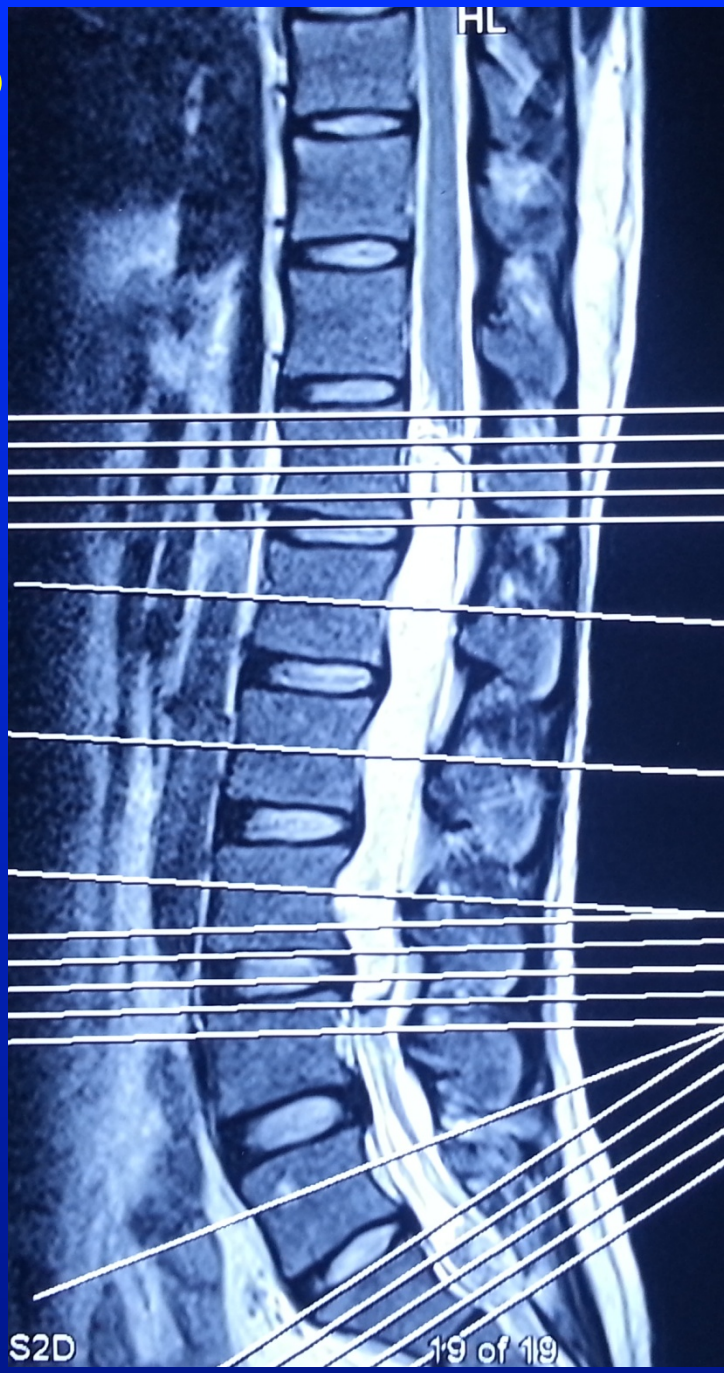
Lumbar puncture

- C/I in complete spinal block by the tx.
- Should not be the first test performed.
- CSF may show extremely elevated protein levels and / xanthochromia

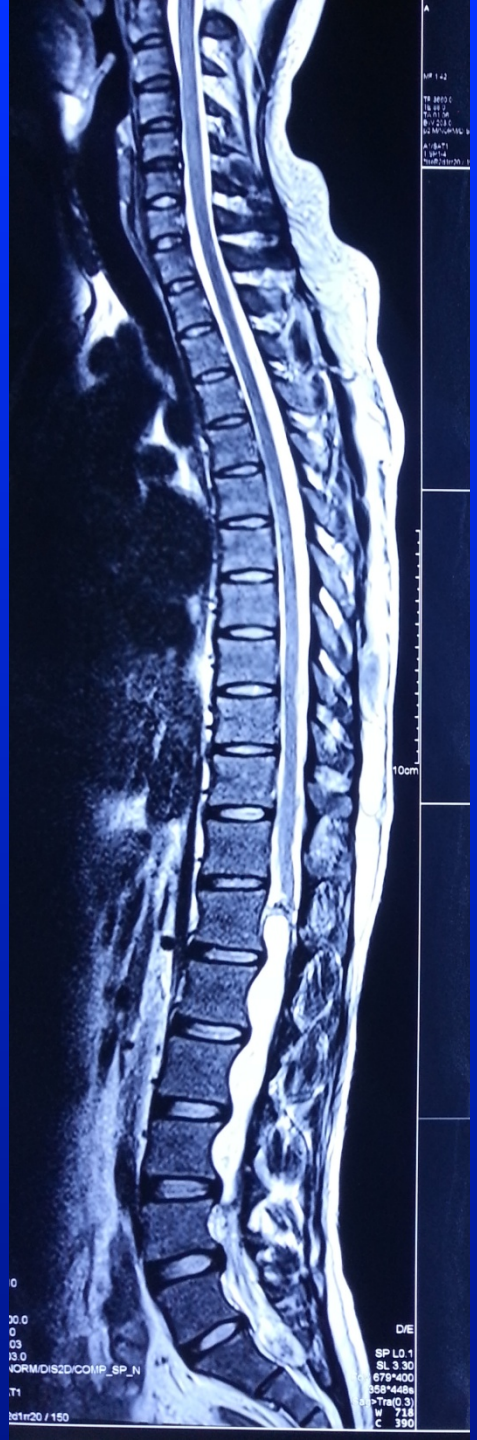
Tx specific characteristics

Ependymoma

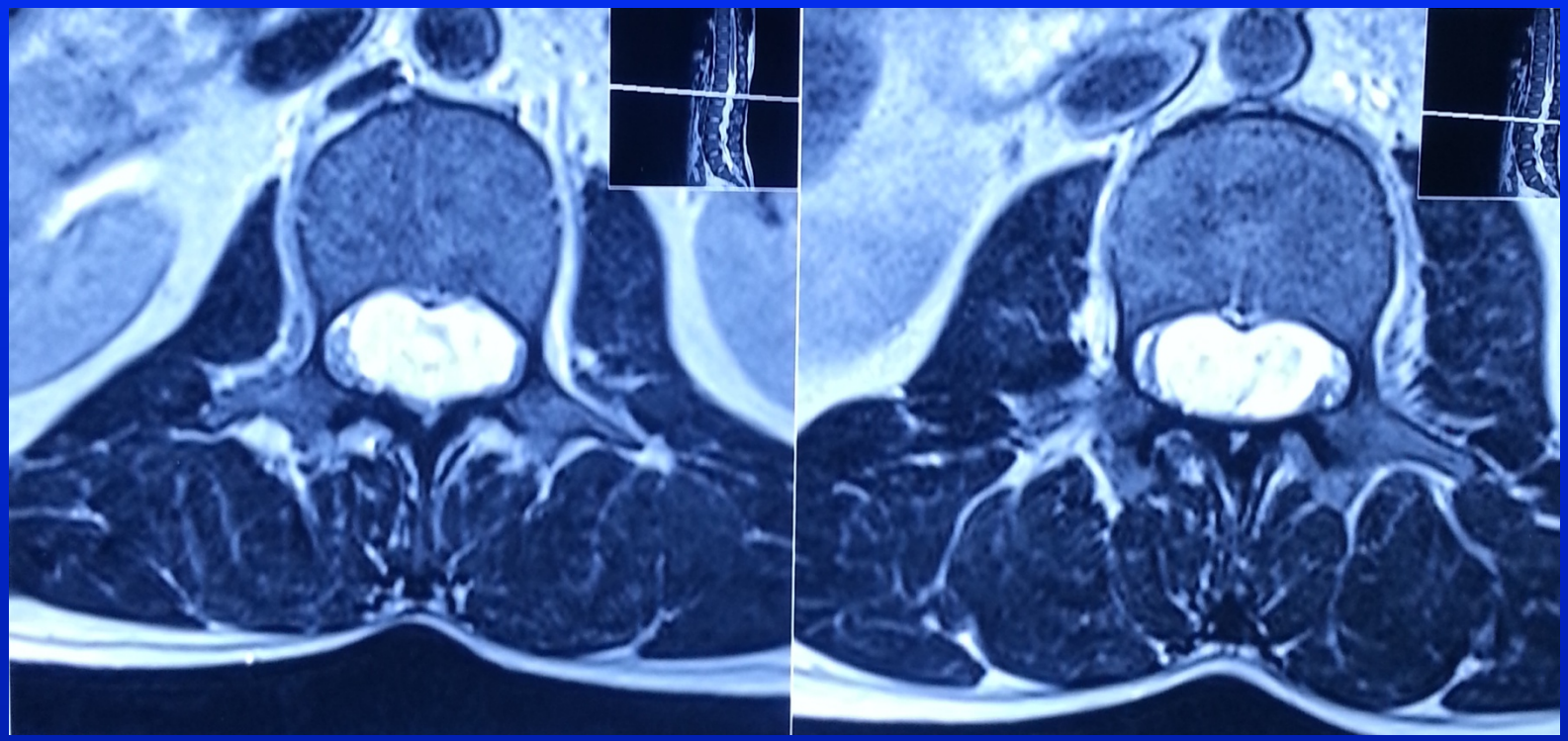
- 40-60% in adults, 30% in children
- most common intrinsic SCTx
- male predilection
- mean age - 35-40 years
- occur anywhere, m/c in conus medullaris
- rarely change growth characteristics and metastasize.
- hypovascular, well circumscribed, and noninfiltrative



- Arises from ependymal cells of central canal
- Most WHO grade II
- Slow growing
- Compress rather than infiltrate cord
- tend to expand the cord symmetrically and focally, astrocytomas more diffuse and eccentrically located



Intramedullary Ependymoma

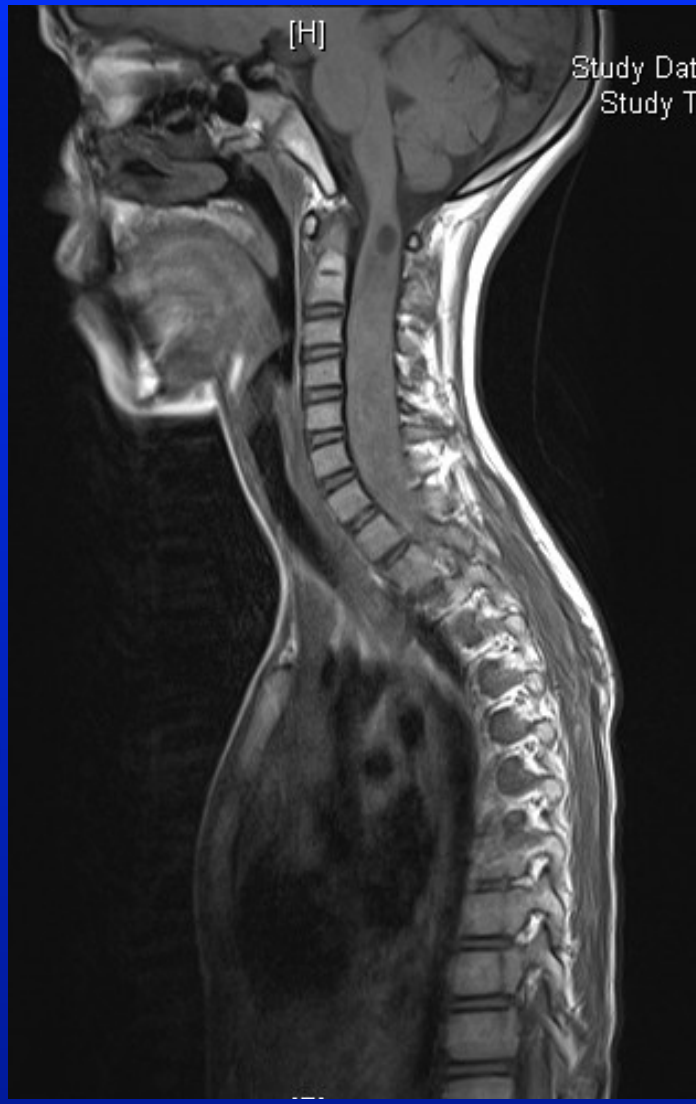


Usually enhance more homogeneously than astrocytomas with sharply defined poles and are capped superiorly by a cyst, inferior cyst is less common

Astrocytoma

- **1/3 of spinal cord gliomas**
- **m/c IMSCTx in children**
- **Holocord involvement common in children**
- **WHO: Grade I: 75%**

  Grade IV: Uncommon (0.2-1.5%)
- Only 10-20% are high grade (3-4)
- Usually grey tumor, more infiltrative , often poor plane

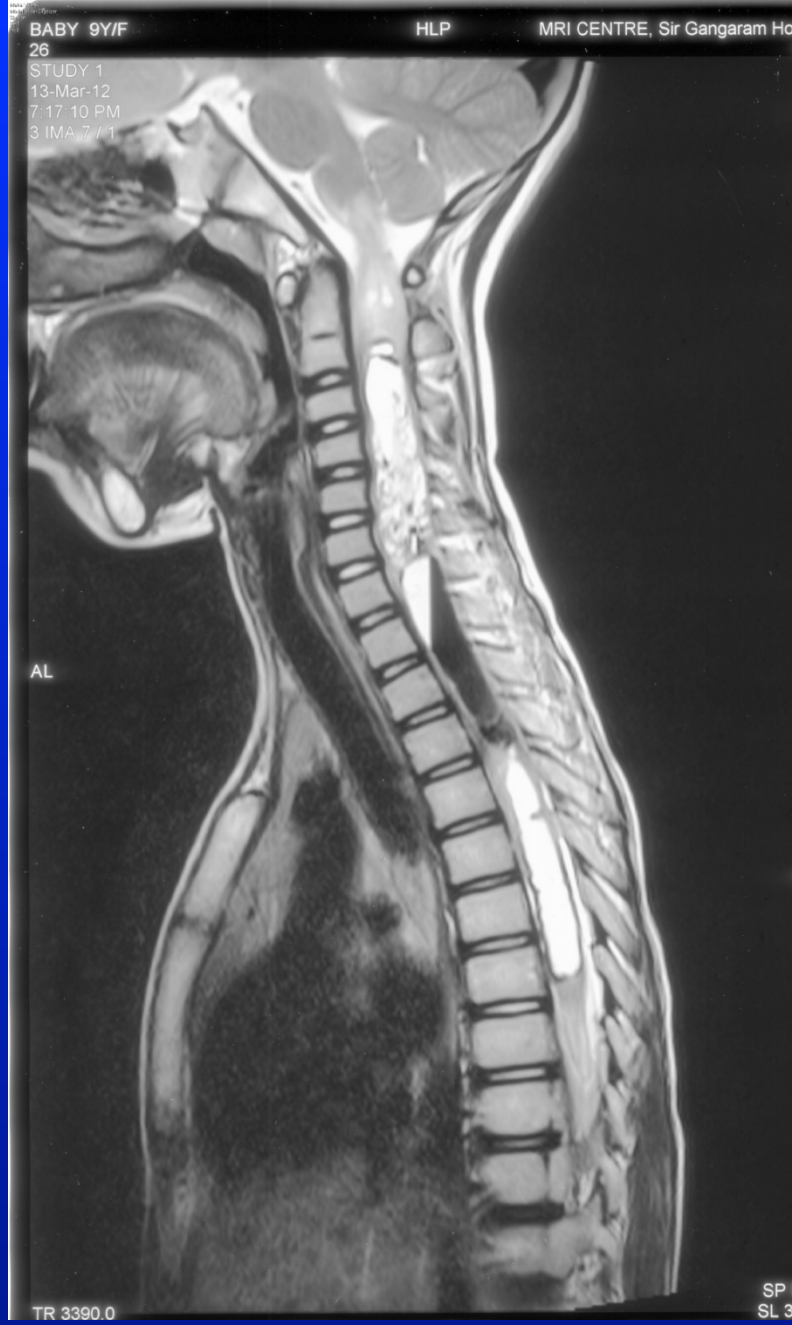


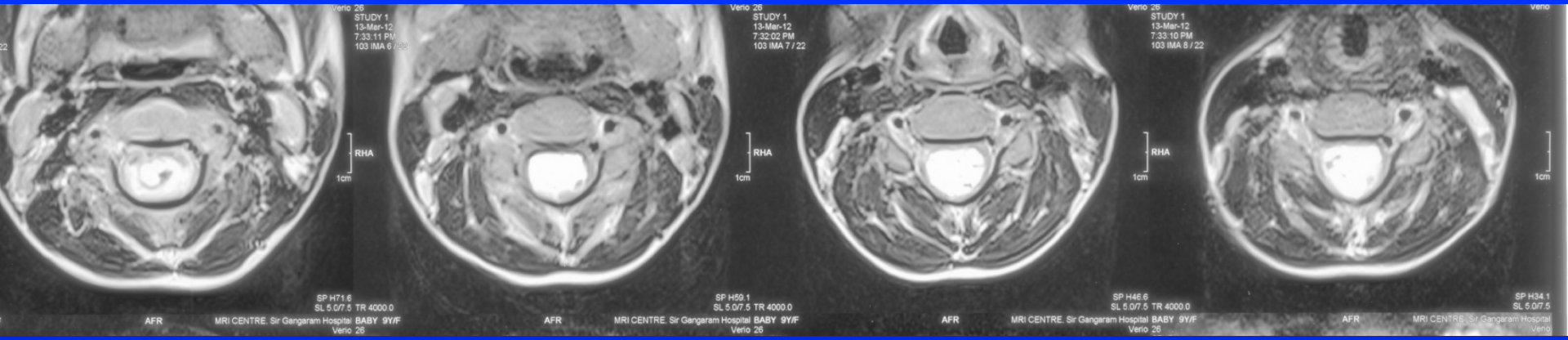
Imaging:

CT- Canal widening & VB scalloping

MRI -

- Poorly defined margins
- T1: Iso- to hypointense
- T2: Hyperintense
- Inhomogenous enhancement
- Cysts common
- Average length of involvement: 7 vertebral segments





- 2 histologies – diffuse fibrillary and pilocytic (low grade)

- On MRI diffuse fibrillary usually seen as a non-enhancing diffuse cord thickening while pilocytic are usually focal, intensely enhancing and associated with a large cyst which can span many spinal levels

Ependymoma vs Astrocytoma

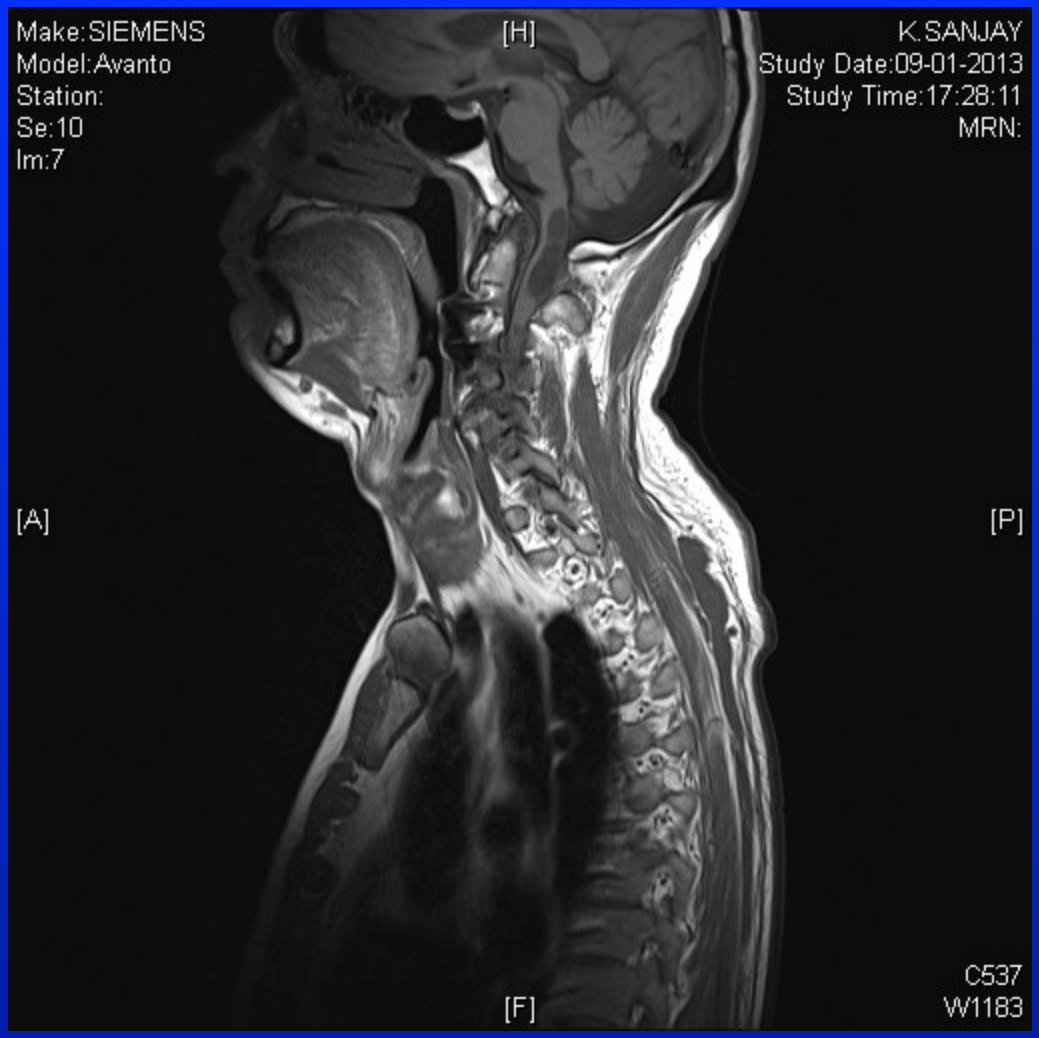
	EPENDYMOMA	ASTROCYTOMA
POPULATION	ADULTS	CHILDREN
LOCATION	CENTRAL	ECCENTRIC
MORPHOLOGY	WELL – CIRCUMSCRIBED	ILL-DEFINED
HEMORRHAGE	COMMON	UNCOMMON
ENHANCEMENT	FOCAL, INTENSE, HOMOGENOUS	PATCHY, IRREGULAR
CONUS/FILUM	YES	NO

Hemangioblastoma

- 1-7% of spinal cord neoplasms
- Cell of origin – unknown
- WHO grade I
- Most solitary
- Multiple – think VHL (approximately 1/3)
- Associated syrinx common
- Most intramedullary
- 10-15% along nerve roots
- Occasionally exophytic
- Diffuse cord expansion
- Highly vascular
- Rarely may be a source of hematomyelia or SAH

Hemangioblastoma

- Slow growing
- On imaging may be solid mass or nodule & cyst
- Tortuous arteries and varicosities often seen emerging from solid component
- Bleeds profusely if entered : not internally debulked before removal but circumferentially removed.
- Complete excision usually feasible
- A-gram and embo possible prior to surgery



Cervical intramedullary hemangioblastoma

MRI -

- T1: Variable, most common isointense
- T2: Hyperintense
- May see flow voids
- Intense enhancement
- May have surrounding edema
- Cyst formation common

Ddx on MRI

- Sarcoidosis
- Tuberculoma
- Dural AV fistula with edema
- Demyelination
- Transverse myelitis
- Syrinx
- Multiple sclerosis - May show multiple lesions of neuraxis
- Cord infarction
- Abscess
- Hematoma
- Arteriovenous malformation
- Amyloid angiopathy

- Determining whether an abnormal MRI definitively indicates the presence of a tumor can be problematic
- Cord appears enlarged when tumor is present, while inflammatory lesions result in normal or minimal increase in cord size
- In cases with syrinx - search for Chiari malformation or abnormal contrast enhancement

Treatment - Surgery

Indication for Surgical Removal -three reasons:

- ✓ ascertain the histological diagnosis
- ✓ apply the most effective oncologic treatment,
- ✓ prevent long-term neurologic dysfunction.

Standard microsurgical techniques with suction and bipolar cautery are used together with specialized instruments that aid in minimizing surgical trauma to normal spinal cord tissue

Ultrasonic Aspirator (CUSA)

- uses high-frequency sound waves to fragment tx tissue
- allows removal of bulk of tx tissue easy and quick.
- However, experience with intraop. monitoring has shown that partial injury to the motor pathways occurred not infrequently.
- Thus modified in that it is safe to remove already partially detached tumor bulk, but less safe to 'dig' into tumor tissue which is still largely in situ.

Laser (Nd:YAG Contact Laser™ System)

- excellent surgical tool for spinal cord surgery
- particularly useful for myelotomy, and to demarcate the glial-tumor interface.
- laser-suction combination is very safe to remove tumor in a piecemeal fashion.
- Contrary to bipolar electric coagulation , intraoperative monitoring can continue unimpaired
- Esply. useful for firm txs.
- For the rare spinal cord lipoma the microsurgical laser is also the instrument of choice for vaporization of fat and internal debulking.

Intraoperative Neurophysiological Monitoring

First used by Spinal orthopedic surgeons

Somatosensory evoked potentials (SEPs)-

- technology slow, prone to artifact, and often difficult to interpret
- SEPs reflect the functional integrity of sensory pathways, information for the more relevant motor pathways , only indirect
- Intramedullary surgery carries clear risk for selective damage to the motor tract, not necessarily reflected by changes in SEPs
- deterioration or loss of SEPs during the myelotomy at intramedullary operations is common and also does not correlate to the motor outcome
- due to signal averaging, a time delay occurs and the identification of injury can lag behind the progress of the surgery

Motor evoked potential (MEPs)-

D-wave

Muscle MEPs

D – wave -

- Based on understanding of motor system developed since 1950s, after Merton and Morton's , description of transcranial electrical motor cortex stimulation in man, gives rise to a recordable travelling wave, the D-wave.
- High clinical correlation, does not require averaging , near real-time feedback, and a pattern of reversibility that allows corrective action
- D-waves are recorded as travelling waves directly from the spinal cord with an electrode inserted into spinal epidural space by the surgeon after laminectomy
- D-wave parameter monitored is the peak-to-peak amplitude.
- A decrease >50% of the baseline value – a/w a long-term motor deficit

Muscle MEPs-

- elicited with *'multipulse technique'*
- CMAPs recorded with needle electrodes from target muscles in all four extremities (thenar, anterior tibialis, and abductor hallucis)
- Real-time feedback is possible here as well.
- Muscle MEPs are recorded in an alternating fashion with D-waves
- parameter monitored is the presence or absence of muscle MEPs (*all-or-none concept*)
- motor deficit occur only when the muscle response is lost

Anesthesia

- GA is performed using TIVA, which entails a combination of IV opioids (fentanyl) and a continuous administration of propofol
- Halogenated volatile anesthetics are avoided - interfere with SSEPs
- Short-acting muscle relaxants are given only for intubation
- Low levels of muscle relaxants are used to minimize spontaneous muscle activity but permit MEP and detect elicited EMG activity
- The spinal cord is sensitive to decreased perfusion, and an arterial line is needed to ensure that dips in blood pressure are detected and corrected as quickly as possible

Combined Interpretation of D-Wave and Muscle MEPs

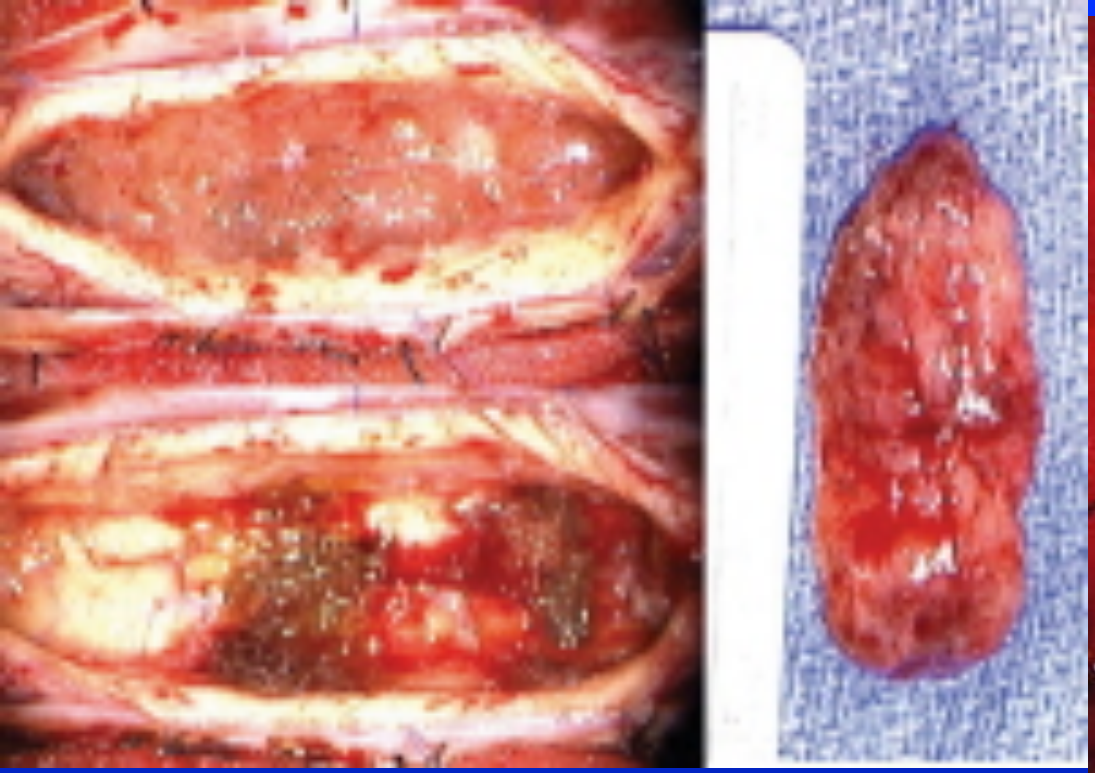
- D-wave amplitude reflects no. of fast-conducting fibers in the corticospinal tract. If 50% of these fibers are damaged by the procedure, the amplitude will decrease to 50% of its baseline value.
- In general ,D-wave amplitude decrease is a/w loss of some muscle MEPs
- In any event, preservation of D-wave >50% cutoff value is predictive of long-term preservation (or recovery) of voluntary motor control in the lower extremities.
- With loss of muscle MEPs and preserved D-wave amplitude, a temporary motor deficit is expected postop. In this situation it is still safe to complete resection, or to pause and wait for recordings to improve again, which they often do.
- This situation is the window of reversible change, which allows for a change in surgical strategy before irreversible injury has occurred

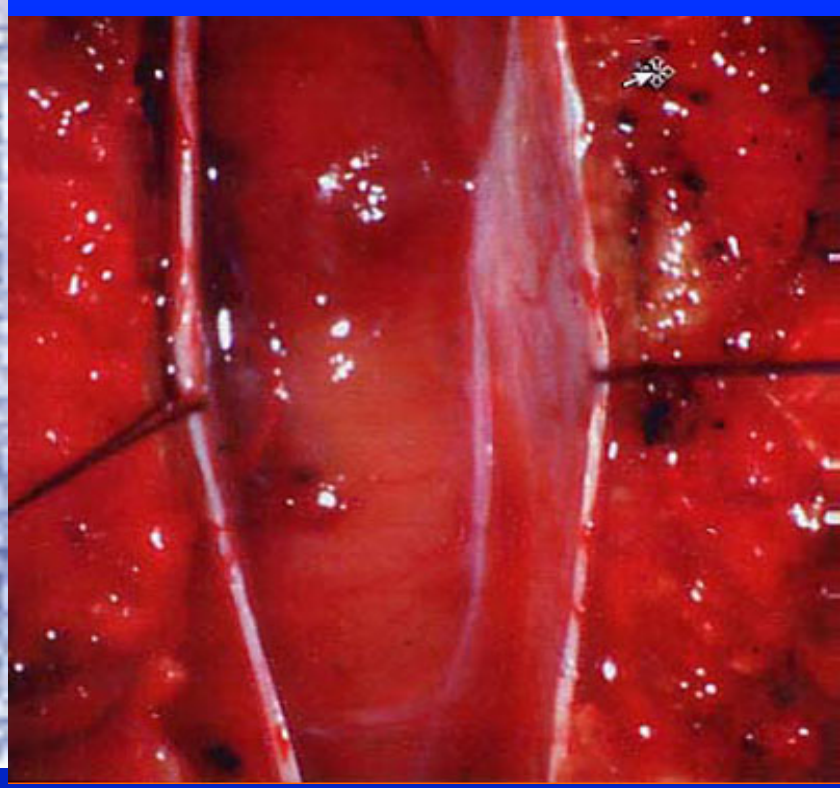
Incorporation of Neurophysiologic Information into Surgical Techniques

- Usually MEP changes occur towards the end of the resection
- Often muscle MEPs disappear first, preceded by an increase in threshold
- S/t pausing the resection and irrigating the cavity with warm saline results in reappearance of the response
- Similarly, some D-wave amplitude decrease may also be reversible by pausing and irrigating
- Sometimes dissection in a particular location results in MEP changes, and the resection can proceed at a different area in the meantime
- Sudden decrease in D-wave amplitude, often coinciding with sudden loss of muscle MEPs, is considered a result of vascular mechanism rather than direct physical tissue manipulation
- Temporary moderate elevation of mean blood pressure has been a successful means to improve the MEPs, with a satisfactory clinical result postoperatively

Treatment - Surgery

- Laminectomy approach
- Midline durotomy separate from arachnoid to avoid precipitous CSF egress and decompression of epidural veins and bleeding
- Arachnoid opening
- Midline myelotomy to tumor
- Intraoperative U/S useful for delineating poles, localizing myelotomy, defining cysts (which are generally not excised)





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View of a cervical intramedullary ependymoma in situ after midline myelotomy and initial dissection . The tumor was removed en bloc , and the postsurgical cavity in the spinal cord is shown .

Treatment - Surgery

- 30-80% incidence of instability in children with multilevel laminectomy for intramedullary tumor
- In adults stability largely dependent on integrity of facet joints
- In children dorsal ligaments provide significant tension band preventing forward flexion and instability
- Hence osteoplastic laminotomy preferred

Operative details

- Spinal cord is sensitive to decreased perfusion, and hypotension should be avoided..
- Perform myelotomy at the thinnest area between the tumor and spinal cord
- Myelotomy should be made in a linear fashion to spare vertically running white matter tracts
- Incision generally made in midline, although occasionally eccentric lesions may be approached through the dorsal root entry zone.

- Exophytic component initial area of approach
- Immediate biopsy taken for histological examination
- Resection initiated at midportion rather than poles
- Poles are least voluminous and manipulation most dangerous
- Debulk exophytic part prior to parenchymal tumor
- Monitoring spinal cord function using intraoperative electrophysiology useful

Astrocytoma

- Controversy regarding extent of resection
- Several studies have shown no correlation between extent of resection and incidence of recurrence or progression free survival
- 2 studies have shown ↑ survival with radical resection vs partial resection or Bx Epstein et al J Neurosurg, 1992. **77**(3): p. 355-9
- Recommendation- as radical a resection as dictated by tumor planes and changes in electrophysiology in benign lesions

Outcome for astrocytomas

- ***Neurological outcome***
  - Gross total (>95%) or subtotal resection (80-95%) does not significantly affect the long-term outcome
  - Partial resection (<80%) fared significantly worse
- ***Oncological outcome***
  - even with gross total resections, some residual microscopic fragments always remain in the resection bed.
  - this residual tissue may remain dormant or involute over time
  - a resection that exceeds 80-90% removal is as good as 98-'100'% removal in terms of long-term progression-free survival

Astrocytoma

- Grade is most important predictor of long term survival:

Grade	*Median survival*
WHO I	98 mth
WHO II	68 mth
WHO III	15 mth

- 5 yr survival:

All astrocytomas	50-60%
Low grade	80%
High grade	0-15%

Ependymomas

- Overall 5 year survival 70-90%
- 10 year survival 70%

Surgery	*10 yr survival*
Total resection	85-90%
Subtotal	80%
Biopsy	25%

- Tumor grade only independent variable predictive of outcome – well differentiated 97% 5 yr survival, poor or intermediately differentiated 71%

Developmental tumors (3%)

- Dermoid, epidermoid, and teratoma are slow-growing neoplasms
- Thoracolumbar predominance
- These may have a dense capsule, precluding complete removal
- Residual compatible with prolonged symptom-free survival
- Debris produced by the tumor may cause an early recurrence of symptoms

Lipoma (2%)

- Fibrous adhesions to cord make total removal difficult
- Carbon dioxide laser particularly useful for resection

Others (4%)

- Unusual lesions include subependymoma, ganglioglioma, intramedullary schwannoma, and neurofibroma
- Management of low-grade lesions parallels other indolent lesions.

Outcome after surgery

- Dysesthetic pain common especially if present preoperatively
- Loss of proprioception because of dorsal myelotomy, deviations from midline and excessive retraction contribute to this – usually improves wks – months
- Generally stable post op – few significantly improve or deteriorate
- Tumor grade, tumor type, surgery (for ependymoma but not all astrocytoma) and severity of preop deficits are prognostic indicators for neurologic outcome and survival

Radiation

- Astrocytoma –
  - minimally resected or biopsied benign lesions
  - high grade tumors
  - rapid recurrence or progression
- 50 Gy locally over 25 doses
- Ependymoma – offered to incomplete resections

Radiation

- Hemangioblastoma – surgical excision almost always & curative – no role
- Delayed effects include:
  - Radiation damage to spinal cord
  - Spinal deformity
  - Radiation induced tumors (in children – 10 & 20 yr 2nd malignancy rate – 4 & 13%

Residual/recurrent tumor

- Residual tumor can be considered for repeat resection, radiation therapy, or observation
- In recurrence , imaging of entire neuraxis warranted as even benign ependymomas may change their growth characteristics and produce seeding.

Current Management of Intramedullary Neoplasms in Children and Young Adults

- Majority of neoplasms are histologically benign being astrocytomas or gangliogliomas
- Most common location- cervical region
- Plain radiographs mandatory in presentation with scoliosis
- Serve as baseline for future management of spinal deformity
- Two-thirds develop spinal deformity following laminectomy
- one-third require stabilization procedure

Factors associated with progressive deformity

- cystic tumors
- prior radiotherapy
- age less than 7 years
- cervical region

Mean time to stabilization procedure 3.4 years

Extradural ependymomas

- Occur in four perisacral locations
  - extradural spinal canal in association with dural part of terminal filum
  - bone substance of sacrum
  - pelvic cavity anterior to sacrum
  - subcutaneous tissues dorsal to sacrum
- Posterior subcutaneous location most common
- Histologically, all reported cases of extradural ependymomas -myxopapillary

- Intraspinal extradural ependymomas arise from ependymal cell remnants in extradural dural part of the terminal filum
- Others arise from ependymal rests present at birth
- Present with symptoms caused by local mass effect
- local pain caused by erosion into the sacrum most common
- No dissemination within the CNS
- Significant risk of metastasis to other organ systems, such as lymph, bone, lung, and liver.

THANKYOU