Spinal cord tumors from childhood to maternity

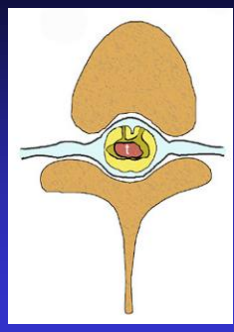
Thierry A.G.M. Huisman, MD, FICIS, EQNR

Professor of Radiology, Pediatrics and Neurology
Director of Pediatric Radiology and Pediatric Neuroradiology
Johns Hopkins Hospital

What will be discussed ?

Neoplasms
- Extra- vs intraspinal
- Extra- vs intradural
- Extra- vs intramedullary

- Spinal metastasis



Epidemiology

- 0.5-1% of all CNS tumors are spinal cord neoplasms (rare lesions)
- 2-4% of all CNS glial tumors are located in the spinal cord
- 35% of all intraspinal tumors are intrinsic spinal cord neoplasms
- 90% are glial tumors
- Most are malignant

Epidemiology

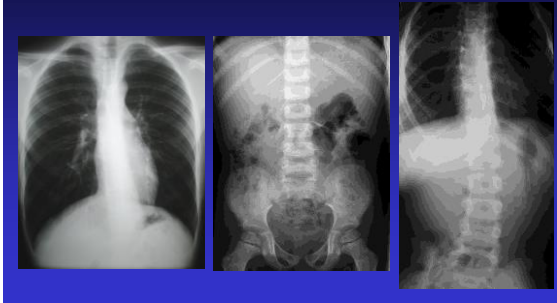
- All age groups are affected
- More frequent toward end of first decade and beginning of second
- No sex predeliction (F=M)
- Spinal neoplasms are less common in children compared to adults

Major problem

- Non-specific clinical symptoms
- Slowly progressive
- Long history of exacerbations and remissions (edema fluctuation)
- Children usually present (very) late

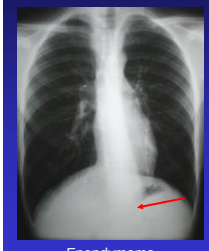
Non-specific clinical symptoms

Which child has a spinal tumor ?

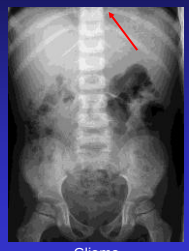


Non-specific clinical symptoms

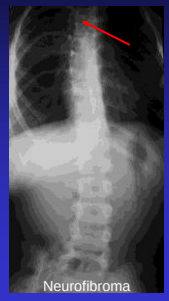
Which child has a spinal tumor ?



Ependymoma



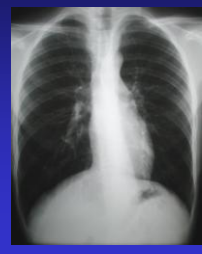
Glioma

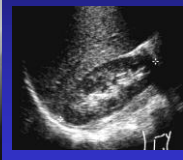


Neurofibroma

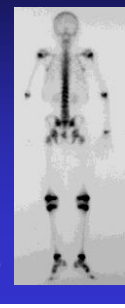
Non-specific clinical symptoms

Typical “radiological history“ of a child with a spinal cord tumor





Incorrect work-up

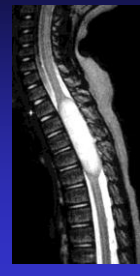


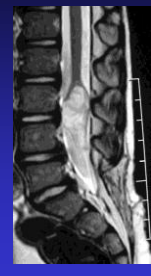
Clinical presentation: Key features

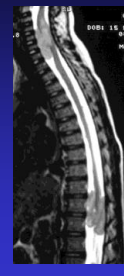
- Progressive motor weakness
- Progressive scoliosis
- Gait disturbance
- Rigidity/paraspinal muscle spasm
- Sensory deficits less common (?)

- 25-30% present with back pain !!!

Every child with persistant back pain should be taken seriously







It could be a spinal cord tumor !!

Back pain

- Spinal pain (70%)
  - Dull and aching pain, localized to bone segments adjacent to tumor
  - Distension of dural sack by enlarged cord ?
- Root pain
  - Mimicks pain caused by nerve root compression
- Tract pain
  - Vague, burning pain with paresthesias
  - Infiltration of spinothalamic tracts ?

Epstein F, Epstein N. Pediatric neurosurgery 1982: 529-540

Increased intracranial pressure

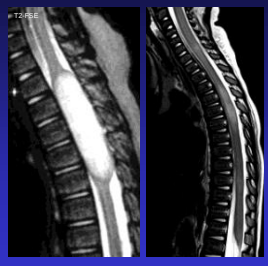
15% of childen with spinal cord neoplasms may present with symptoms of increased intracranial pressure

- Elevated CSF protein ~> impairs CSF resorption
- Blockage of foramen magnum (cervical cord tumor)
- Subarachnoid tumor hemorrhage
- Subarachnoid seeding

Magnetic resonance imaging

1. Cord expansion is a leading criterion for spinal neoplasms

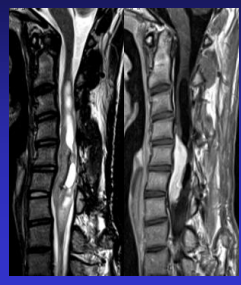
   If absent or minimal, consider non-neoplastic etiologies: MS, ADEM, sarcoidosis, ischemia, ...



Astrocytoma ADEM

Magnetic resonance imaging

2. Vast majority of lesions show enhancement



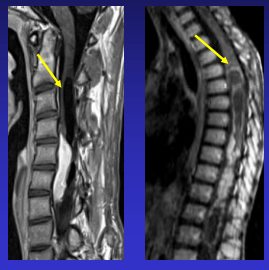
12Y, M, low grade glioma

Magnetic resonance imaging

3. Cysts are frequently seen in spinal tumors

Non tumoral "cysts":
- At poles of solid tumor
- Reactive dilatation of central canal
- Fluid produced by tumor
- No enhancement

Tumoral cysts:
- Within the tumor
- Peripheral enhancement
- Should be resected

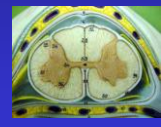


Non tumoral cyst Tumoral cyst

Neoplasms of spinal cord

- Same spinal neoplasms are seen in adults and children, however incidence and presentation differ

- Glial tumors (90-95%)
  - Astrocytoma (60%)
    - Pilocytic and anaplastic
  - Ependymoma (30%)
    - Myxopapillary ependymoma



Neoplasms of spinal cord

- Glial tumors (90-95%)
  - Astrocytoma (60%)
    - Pilocytic and anaplastic
  - Ependymoma (30%)
    - Myxopapillary ependymoma
- Nonglial tumors (5-10%)
  - Hemangioblastoma, Subependymoma, Ganglioglioma, Paraganglioma, Metastasis, Lymphoma, PNET, Neurocytoma, Oligodendroglioma

Spinal astrocytoma

- Most common spinal cord tumor in children
- In children more rostrally compared to adults

- 50% are cervico-(thoracic)
- Small number of segments
- Most frequently pain and motor dysfunction, followed by gait disturbances, torticollis and scoliosis
- Bowel and bladder deficits uncommon

Spinal astrocytoma

- Grade I: pilocytic astrocytoma
- Grade II: fibrillary type
- Grade III: anaplastic astrocytoma
- Grade IV: glioblastoma multiforme

- I,II: 75-80%, III: 20-25%, IV: 0.2-1,5%

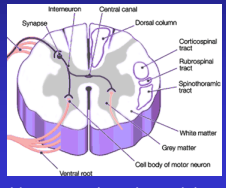
- Slow growth and low recurence rate

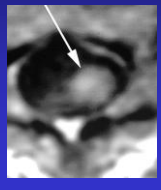
Spinal astrocytoma

- Cleavage plane rarely present between tumor and normal spinal cord
- Infiltrative tumor extending along scaffold of normal astrocytes
- 20-40% of children: tumor cysts (polar and intratumoral) as well as syrinxes (caudal and rostral)
- Rarely hemorrhagic

Spinal astrocytoma

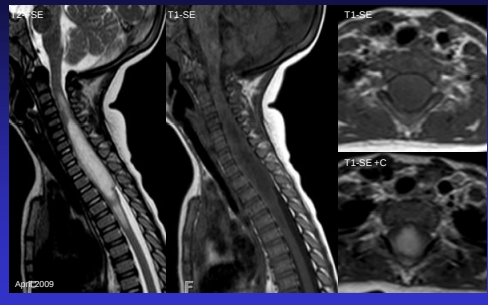
Eccentric location
Asymmetric expansion





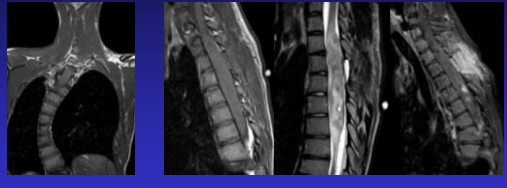
White matter along the periphery!!

Spinal pilocytic astrocytoma



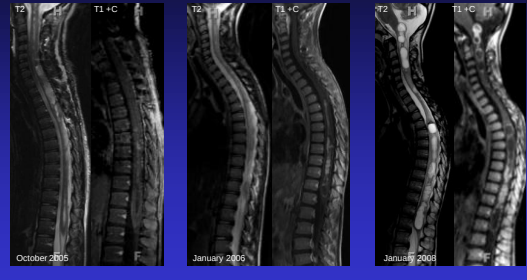
Girl, stopped walking at **14m**, used legs while sitting to "scoot" around. In daycare: curled up around abdomen (suspected abdominal discomfort). Because of progressive weakness of head/upper body ~> MRI at **16m**

Spinal pilocytic astrocytoma



Presented with **scoliosis** and **focal pain**
T2 heterogeneous signal

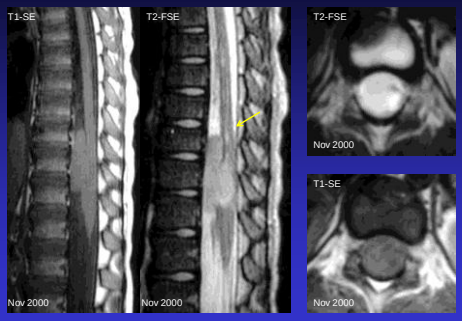
Spinal pilocytic astrocytoma



Recurrent tumor, holocordal affection

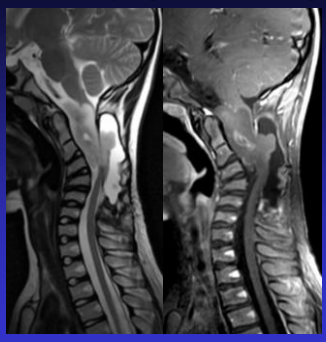
11y M, long history of back pain and left lower extremity weakness.

Spinal exophytic astrocytoma



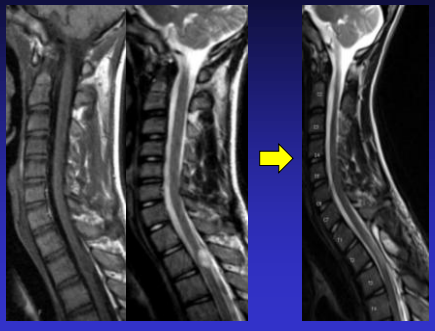
3 Y, M, Thoracolumbar pain. Exophytic growing low grade astrocytoma with discrete hydromyelia

Spinal exophytic astrocytoma



Post partial resection, minimal clinical symptoms!!!

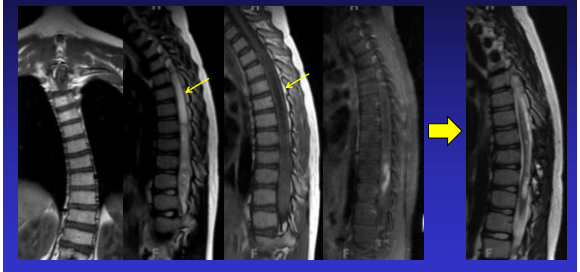
Spinal pilocytic astrocytoma



Excellent postoperative recovery

Fibrillary astrocytoma grade II

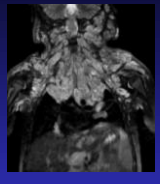
Spinal pilocytic astrocytoma

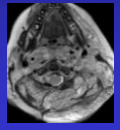


Recovery of the hydromyelia after tumor resection

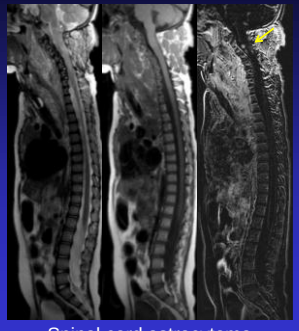
Pilocytic astrocytoma

Spinal astrocytoma





NF1



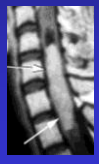
Spinal cord astrocytoma

Spinal ependymoma

- Second most frequent spinal cord tumor
- Most frequently in cervical cord
- Centrally located (ependym)
- Typically back pain and motor weakness
- More frequently sensory symptoms which could be related to central location (spinothalamic tracts !!)

Spinal ependymoma

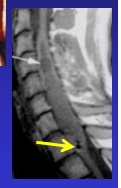
- Slow growth (Grade I,II)
- Tend to compress adjacent spinal cord tissue rather than infiltrate
- Often clear cleavage plane
- Polar cysts common, tumor cysts less common
- Small feeding vessels
- Strong enhancement



Radiographics 2000; 20: 1721-1749

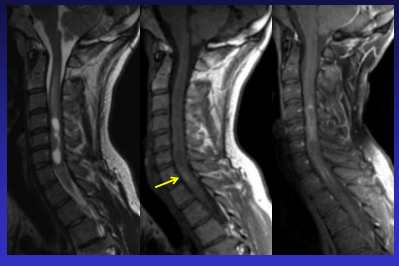
Spinal ependymoma

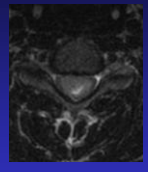
- Small feeding vessels
- Strong enhancement
- High vascularity may result in intratumoral and subarachnoid hemorrhage
- Often "cap sign": Rim of hemosiderin at tumor poles due to hemorrhages



Radiographics 2000; 20: 1721-1749

Spinal ependymoma





Central location
Peripheral white matter compressed

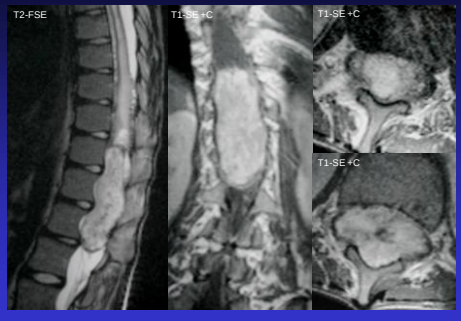
Ependymoma with tumor cyst and intratumoral hemorrhage

Ependymoma

Myxopapillary ependymoma

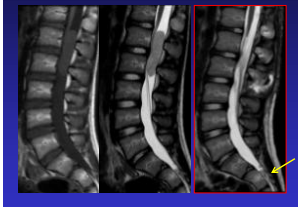
- Variant of ependymoma
- 13% of all spinal ependymomas
- More common in male children
- Predilection for conus medullaris and filum terminale
- Arise from ependymal glia of filum
- Mucin producing, polylobulated soft tumor
- Lower back pain, leg weakness and sphincter dysfunction

Myxopapillary ependymoma

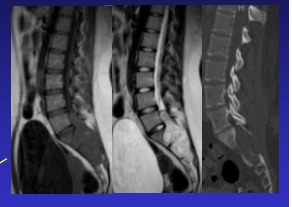


14Y, M. Back pain and bladder dysfunction.

Myxopapillary ependymoma



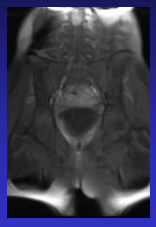
Excellent postoperative result, however small nodule persisted



Significant bony remodelling, large bladder

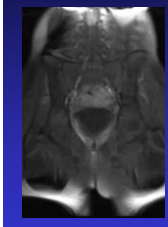
Myxopapillary ependymoma

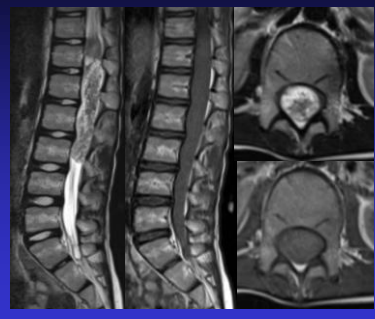
Trouble with the non-specific presentation



Hip pain, incidental finding.

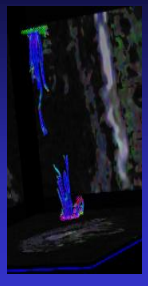
Myxopapillary ependymoma

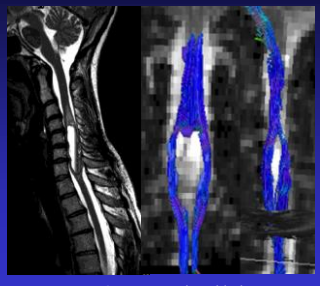




Hip pain, incidental finding. Myxopapillary ependymoma

Differentiation by DTI



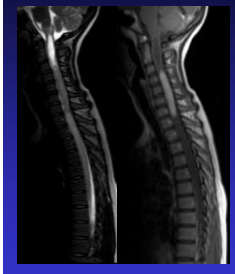


Courtesy Karl Lövblad

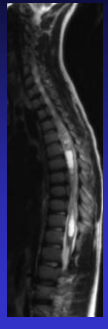
Neoplasms of spinal cord

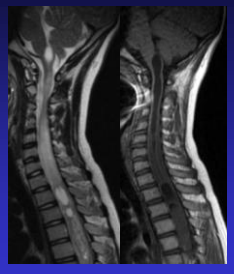
- Glial tumors (90-95%)
  - Astrocytoma (60%)
    - Pilocytic and anaplastic
  - Ependymoma (30%)
    - Myxopapillary ependymoma
- Nonglial tumors
  - Hemangioblastoma, Subependymoma, Ganglioglioma, Paraganglioma, Metastasis, Lymphoma, PNET, Neurocytoma, Oligodendroglioma

Nonglial tumors



Neurocytoma



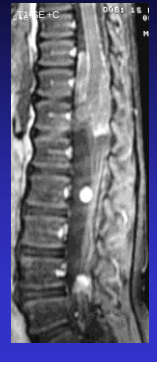


Ganglioglioma grade I

Many may look similar

Various nonglial tumors

- Subependymoma
- Ganglioglioma
- Paraganglioma
- Metastasis
- Lymphoma
- PNET
- Neurocytoma
- Oligodendroglioma

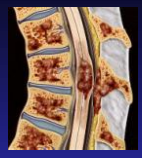


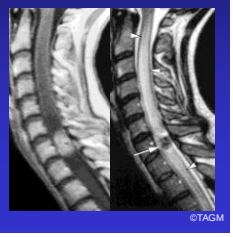
Metastasis

- Intramedullary metastasis are rare
  - Hematogenous (arteries) spread
  - Direct extension from leptomeninges
- Extramedullary, intradural more frequent
  - CSF-seeding (intracranial neoplasms)
  - Medulloblastomas, ependymomas, anaplastic astrocytomas, germinomas, choroid plexus tumors, pineal gland tumors,…..

Metastasis

- Intramedullary metastasis
  - Rapid progession of symptoms
  - Poor prognosis
  - Extensive edema



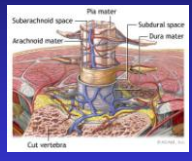


Metastasis

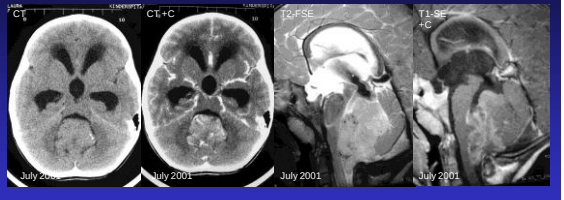
- Intramedullary metastasis
  - Rapid progession of symptoms
  - Poor prognosis
  - Extensive edema
- CSF seeding
  - Nodular and irregular thickening of thecal sac
  - Coating of surface of cord
  - Nerve roots thickening

Metastasis

- CSF-seeding
  - Lumbosacral region most commonly affected
  - Contrast-enhanced sequences are mandatory
  - Vessels on the cord surface may mimic metastasis
  - Staging has to be performed preoperatively!!

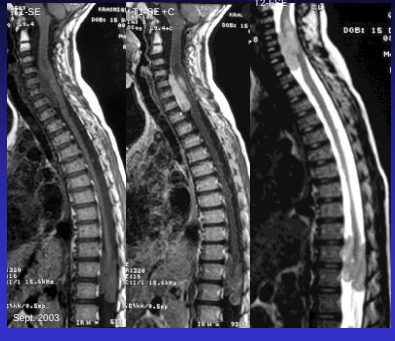


Metastasis



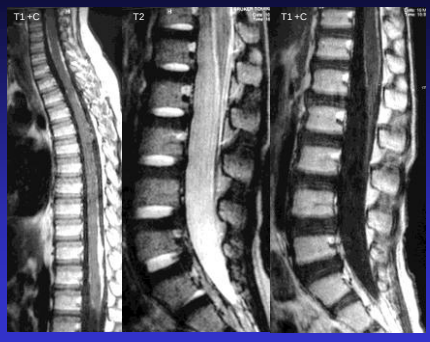
4Y, F. Anaplastic ependymoma, Surgery July 2001, Chemo and Radiotherapy until august 2002

Metastasis



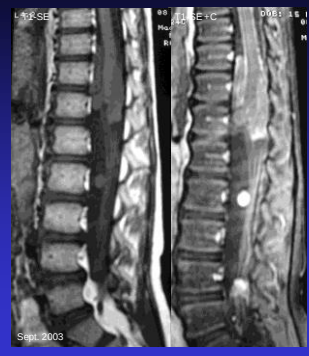
4Y, F. Anaplastic ependymoma, Surgery July 2001, Chemo and Radiotherapy until august 2002

Metastasis



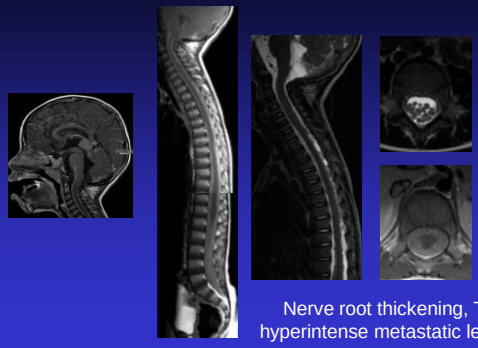
PNET. Diffuse CSF-seeding with "sugar coating"

Metastasis



4Y, F. Anaplastic ependymoma, Surgery July 2001, Chemo and Radiotherapy until August 2002

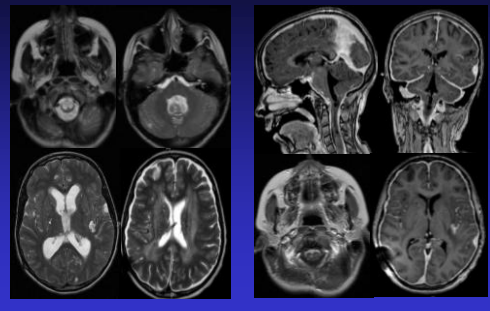
Metastasis



Nerve root thickening, T2-hyperintense metastatic lesions

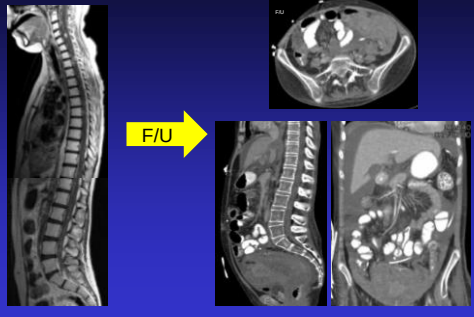
Metastatic pineoblastoma

Metastasis



Pilomyxoid astrocytoma

Metastasis



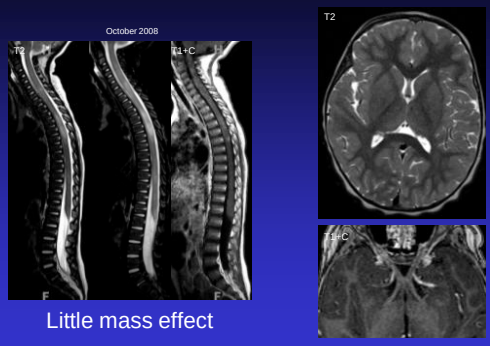
Shunt!!!

Pilomyxoid astrocytoma

Differential diagnosis

- Non-neoplastic diseases
  - Transverse myelitis
  - Multiple sclerosis
  - ADEM
  - Devic Disease

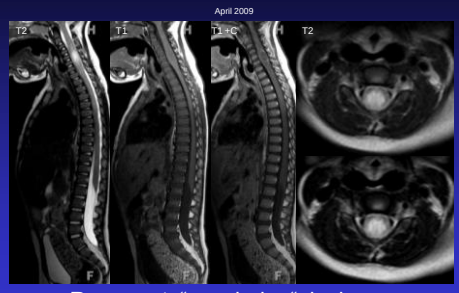
ADEM



Little mass effect

33m M: 2w history of urinary incontinence and rapidly progressing 1d history of lower extremity hemiparesis progressing into hemiplegia. Diagnosis: ADEM, recovered fully within one week with prednison

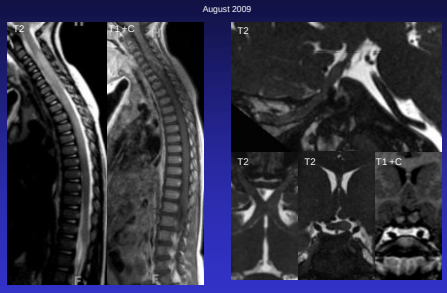
ADEM



Recurrent, “wandering“ lesions

Using his R hand less, R arm pain, limping R leg, bowel and urinary incontinence since 3d after URI 3w ago

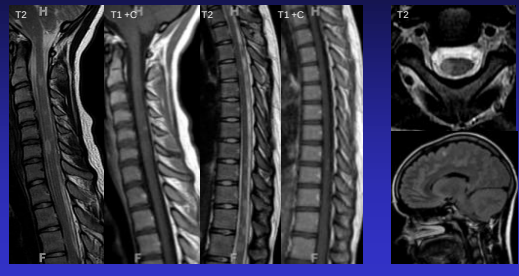
ADEM ~> Devic



Multifocality

3rd episode with black sheet over both eyes. Spinal cord + optic nerves ~ > Devic disease, Neuromyelitis optica

Multiple Sclerosis

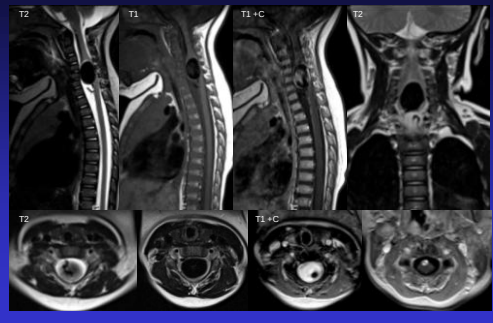


Multifocality, eccentric location, check the brain

17Y F Final Diagnosis: MS

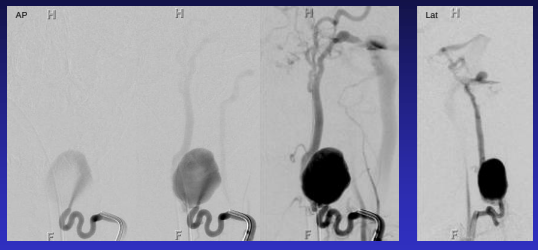
Various confusing “tumors“

AV-Fistula



7M, F. type III perimedullary fistula

AV-Fistula

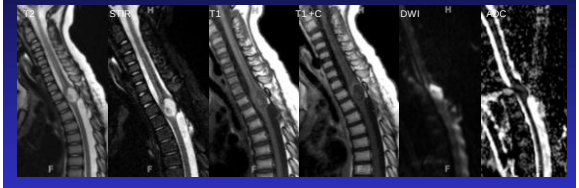


Large venous varix

7M, F. Hypertrophic branch from the left subclavian artery supplying type III perimedullary fistula and resultant venous outflow aneurysm

Various confusing “tumors“

Spinal dysraphia

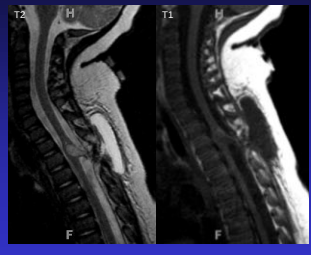


Check clinical history and look for all details

September 2008

3Y, F.

Spinal dysraphia



April 2005

3Y, F. Post repair thoracic dermal sinus with intradural lipoma

Summary

- Spinal cord tumors are rare lesions
- 90-95% astrocytomas and ependymomas
- 5-10% various nonglial tumors
- Unspecific clinical symptoms
- Back pain often leading complaint
- Triplanar MRI with contrast necessary
- Preoperative spine imaging in cases of cerebral tumors with potential CSF seeding

Thank you