Supra- and infratentorial brain tumors from childhood to maternity

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What to expect?

I am going to show you the characteristic imaging findings of following tumors:

- ✓ Pilomyxoid astrocytoma
- ✓ Juvenile pilocytic astrocytoma
- ✓ Astroblastoma
- ✓ Primitive Neuroectodermal Tumor (PNET)
- ✓ Atypical Teratoid-Rhabdoid Tumor (ATRT)
- ✓ Ganglioglioma
- ✓ Desmoplastic infantile ganglioglioma
- ✓ Oligodendroglioma
- ✓ Ependymoma

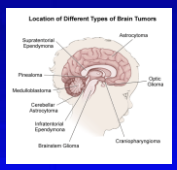
- – Ependymoblastoma
- – Pleomorphic xanthoastrocytoma (PXA)
- – Hemangioblastoma
- – Glioblastoma multiforme (GBM)
- – Choroid plexus carcinoma
- – Pineoblastoma
- – Choroid Plexus papilloma
- – Subependymal Giant cell astrocytoma
- – ……

What to expect?

Please listen carefully and do not distract or interrupt me,

otherwise I cannot get through my 344 characteristic slides in the coming 25 min

- We will "only" discuss
  - the most frequent tumors
  - the most characteristic tumors



http://www.chop.edu/healthinfo/brain-tumors.html

The really important questions:

- Is there a tumor?
- Is the lesion benign or malignant?
- Narrow differential diagnosis

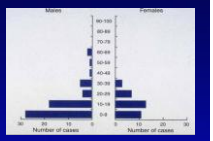
Additional important questions:

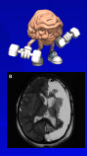
- Localize and characterize the lesion
- Identify threatened structures
- Identify life threatening complications (e.g. herniation)
- Look for additional lesions outside of primary field of view

→ Give clinician the most reliable information for their decision making

“Children are not small adults“

- What is different from adults
  - Clinical symptoms
  - Type and incidence of tumor
  - Imaging characteristica
  - Treatment
  - Prognosis
  - Neuronal/functional plasticity
  - ,.....



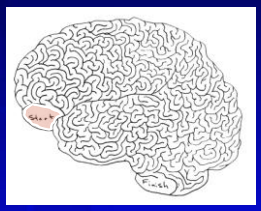


http://www.joey.com

Type and incidence of tumor

- Supratentorial
  - 30% Astrocytoma
  - 15% Craniopharyngeoma
  - 15% Optic pathway glioma

  60%

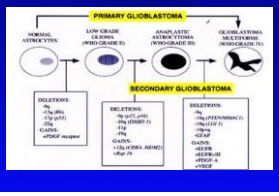


Supratentorial astrocytoma

- 30% of all supratentorial tumors in children
- M=F, all age groups are affected, peak at 7-8Y
- Symptoms depend on location
- Clinics: Seizures, focal neurological defect, signs related to ICP ↑

Supratentorial astrocytoma

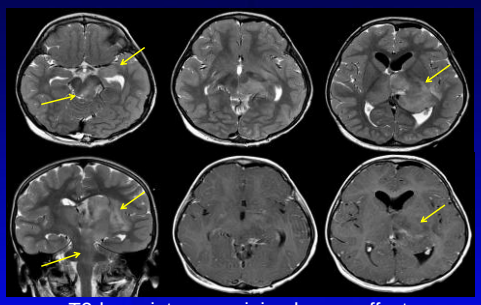
- Most are low grade, GBM does occur
- (J-)PA are less frequent supratentorial than infratentorial
- Spontaneous malignant degeneration may occur



Supratentorial astrocytoma

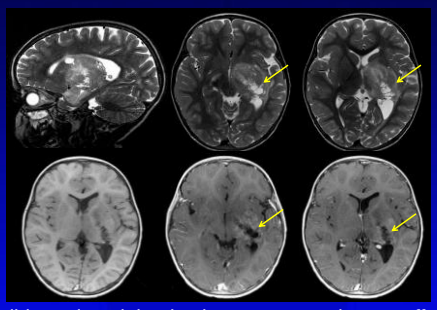
- Variable appearance on imaging
- Solid, solid with necrosis, cystic with mural nodule
- May occur at any location
- Somewhat more frequent in deep location: thalamus, basal ganglia
- Often large at initial presentation
- Symptoms depend on location

Supratentorial astrocytoma



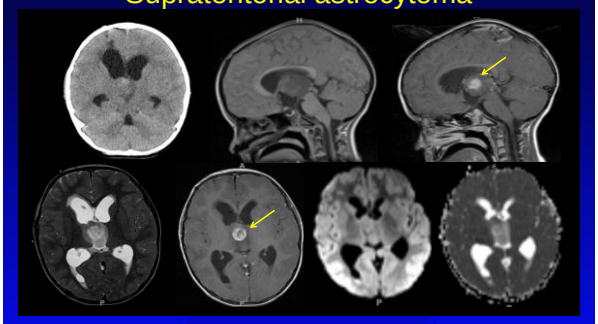
T2-hyperintense, minimal mass effect, infiltration along fiber tracts

Supratentorial astrocytoma



Solid, cystic, minimal enhancement and mass effect

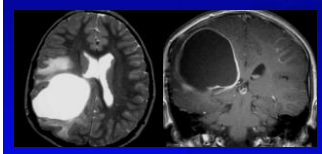
Supratentorial astrocytoma

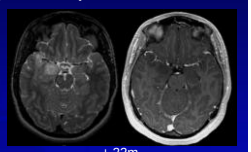


Partial, nodular enhancement

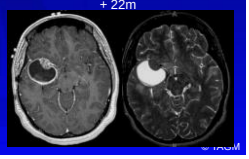
Supratentorial astrocytoma

- Glioblastoma multiforme (grade IV)
- Similar to adults
- Rare in children
- May mimic abscess



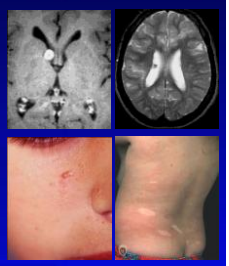


+ 22m

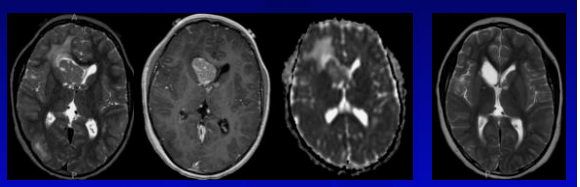


Giant cell astrocytoma

- Near foramen of Monro
- Benign lesion
- 5-15% of TSC patients
- M=F, any age, peak: 5-10Y
- Clinics: Hydrocephalus
- Rarely malignant degeneration
- Arise from subependymal hamartoma?

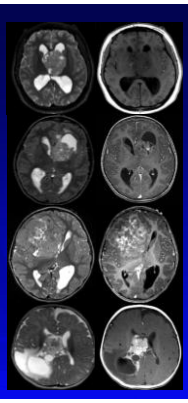


Giant cell astrocytoma



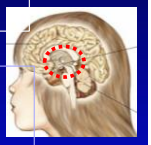
Differential diagnosis

- Many more tumors are known:
  - Ependymoma
  - PNET
  - ATRT
  - Choroid plexus papilloma
  - May look similar to high grade astrocytomas, biopsy may be necessary



Sellar and suprasellar tumors

- Optic pathway glioma/astrocytomas
- Optic Nerve gliomas

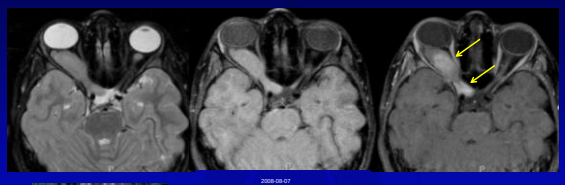


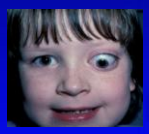
- Craniopharyngeoma
- Hypothalamic hamartoma
- Langerhans' Cell Histiocytosis
- Pituitary tumors
- Suprasellar germ cell tumors

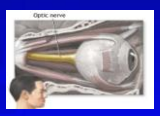
Optic pathway glioma
Optic nerve glioma

- Optic nerve glioma: Starts in intraorbital segment of optic nerve; slow growth; JPA-like histology
- Hypothalamic/chiasmatic tumor: Starts in hypothalamus; more aggressive/invasive, histology similar to hemispheric astrocytomas
- Clinics: Diminished vision, pituitary dysfunction, hydrocephalus, diencephalic syndrome
- 20-50% have NF1

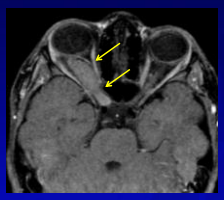
Optic nerve glioma

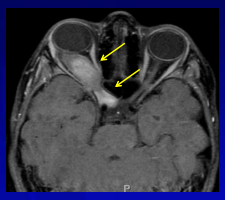






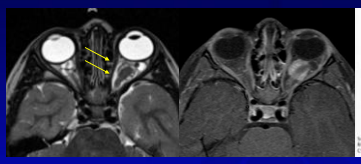
Optic nerve glioma

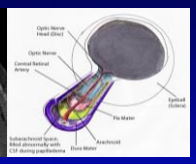




- Frequently stable on follow up
- Look for other (NF1) lesions (UBO, JPA,…)

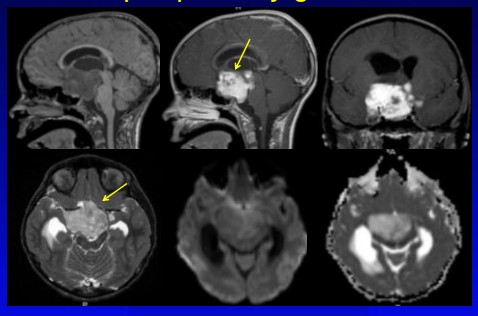
Optic nerve glioma





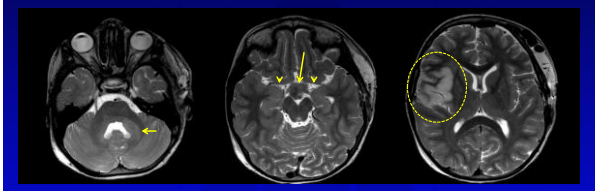
- Subarachnoid optic nerve sheath surrounds glioma
- Differentiation from optic nerve sheath meningeoma (NF2, MISME)

Optic pathway glioma



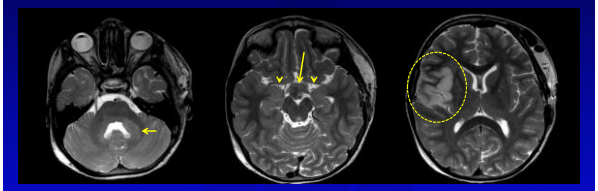
M, 7Y, failure to thrive, diencephalic syndrome, pilomyxoid type juvenile pilocytic astrocytoma

Always look beyond the most obvious findings



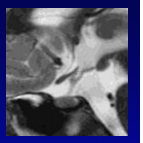
Glioma of the optic chiasm, UBO's, MCA infarction

Always look beyond the most obvious findings

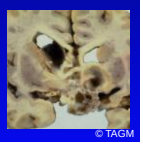


Glioma of the optic chiasm, UBO's, MCA infarction, Moya Moya, post ECA-MCA anastomosis in NF1 patient

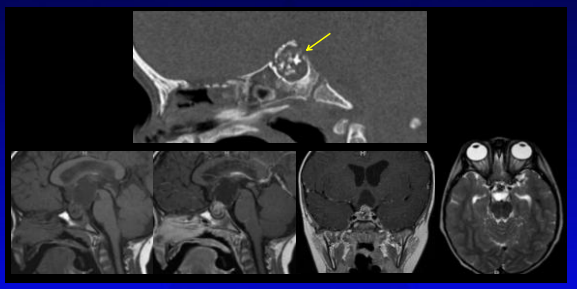
Craniopharyngeoma



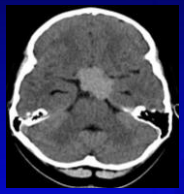
- Along hypothalamic-pituitary axis
- 15% of all supratentorial tumors
- M>F, peak between 10-14 years
- Originate from remnants of pluripotents cells
- Clinics: Visual field defects, pituitary or hypothalamic dysfunction, hydrocephalus
- Imaging: Solid w/wo cysts, calcifications, vary greatly in size
- May infiltrate adjacent brain

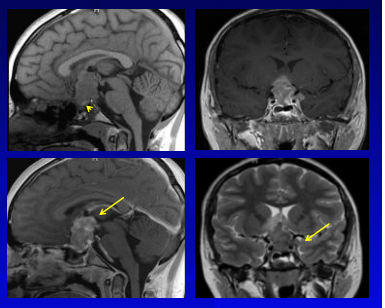


Craniopharyngeoma



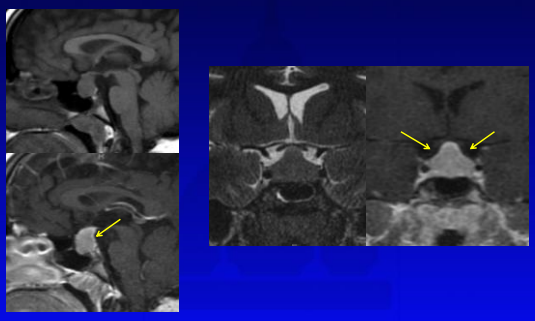
Germinoma





Vision loss, 11Y Girl, confirmed by biopsy

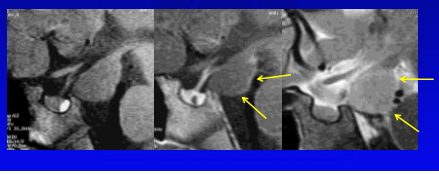
Pituitary macroadenoma



11y female endocrine disorder

Hypothalamic hamartoma

- Tuber cinereum hamartoma
- Rare congenital malformations
- Normal neuronal tissue
- In region of mamillary bodies/tuber cinereum
- Precocious puberty, gelastic seizures



precocious puberty

Infratentorial tumors in children

➢Take advantage of statistics

➢Cerebellar astrocytoma	30-35%
Medulloblastoma	20-25%
Brainstem glioma	20-25%
Ependymoma	10-15%
Total	80-90%

➢Meningiomas, schwannomas, metastasis are rare in children!!!

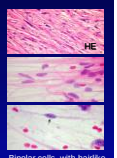
➢"Prognosticators"

Cerebellar astrocytoma

- Most frequently encountered posterior fossa tumor (30-35%)
- Peak incidence 5-13 years
- 1:1.45
- Low grade (75-80%), anaplastic (15-25%)
- Usually benign course, slow growth, expansive
- Located within cerebellar hemispheres or vermis
- Compression of IV ventricle ~> hydrocephalus
- Headache, nausea, vomiting, ataxia, gait disturbance

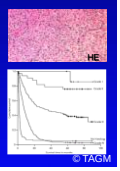
Cerebellar astrocytoma

- Pilocytic astrocytoma (WHO 1):
  - Usually macrocystic with solid tumor nodule
  - "Hairy" tumor
  - Leptomeningeal metastases rare on initial presentation (5%)
  - Good long term prognosis if treated (90%,10y)

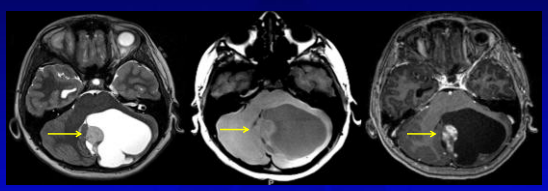


Bipolar cells, with hairlike processes

- Anaplastic astrocytoma (WHO III-IV)
  - Usually in older children
  - More solid, small cells
  - More aggressive/infiltrative, poor prognosis
  - More frequent leptomeningeal metastases

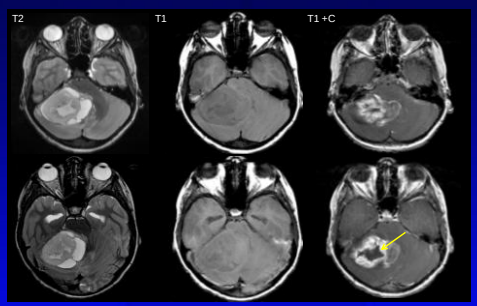


Cerebellar pilocytic astrocytoma



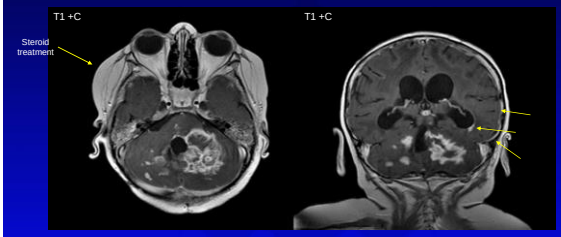
Tumor nodule enhances, cyst does not enhance

Cerebellar pilocytic astrocytoma



Tumor nodule may show central necrosis

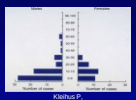
Cerebellar anaplastic astrocytoma



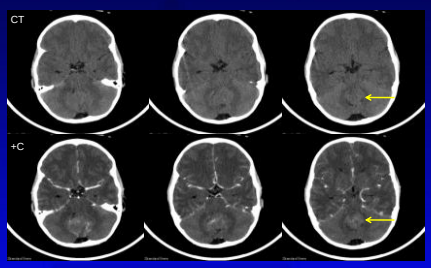
Follow up after treatment,
leptomeningeal and local dissemination

Medulloblastoma

- 2nd/3rd most frequent tumor, 20-25%
- First decade, peak at 7yrs
- 1:3
- 75-90% in cerebellar vermis
- 10-15% in cerebellar hemispheres: lateral medulloblastomas (older children)
- IV ventricle compression with obstructive hydrocephalus
- Overall 5 year survival 60%, depending on histology and risk factors higher survival rates (90%)



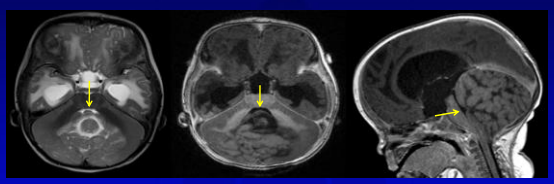
Medulloblastoma



High density

male 7y, headache, vomiting, ataxia

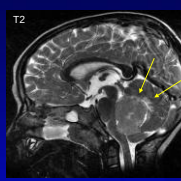
Medulloblastoma



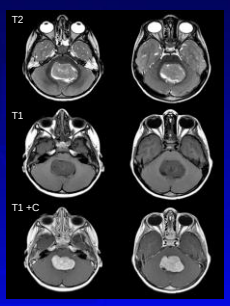
Posterior to fourth ventricle

Desmoplastic medulloblastoma

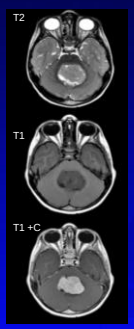
Medulloblastoma

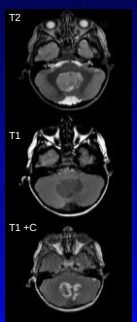


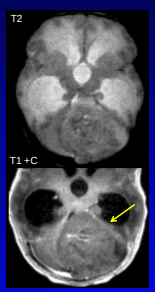
Unsharp posterior margins



Medulloblastoma

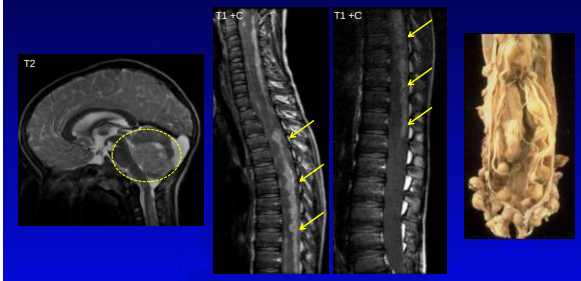






Variable enhancement

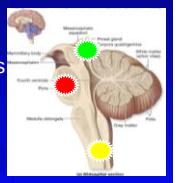
Medulloblastoma



CSF-metastases in 30-50% of children on initial presentation

Brainstem glioma

- 2nd/3rd most frequent tumor, 20-25%
- First decade, peak at 7-9yrs
- 1:1
- Many classification systems
  - Most frequently according to primary location & neuroimaging characteristics
    - Diffuse intrinsic brainstem glioma (80%)
    - Posterior exophytic glioma of cervicomedullary junction (15%)
    - Focal tectal glioma (5%)



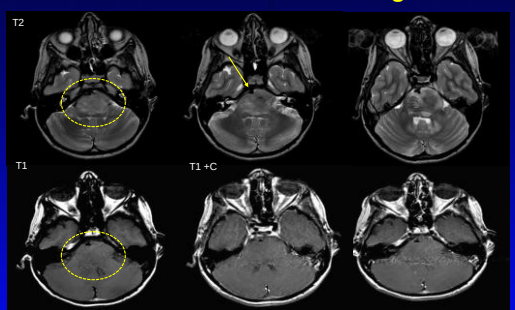
Brainstem glioma

- Depending on localization, prognosis and treatment vary significantly
  - Tectal glioma excellent prognosis compared to diffuse intrinsic brainstem glioma
  - Exophytic glioma may be operated, diffuse brainstem glioma cannot be operated
- Clinical presentation depends on primary location and involved neurofunctional structures

Diffuse intrinsic brainstem glioma

- Most frequently centered within pons
- Involve > 50% of cross-sectional area
- Triad: ataxia, long tract signs, multiple cranial nerve deficits
- Mood change and irritability
- Fibrillary astrocytoma WHO III-IV
- Poor prognosis, most children die < 2 yrs
- No effective treatment
- Radiotherapy may relieve symptoms temporarily
- Neuroimaging is specific, no biopsy necessary

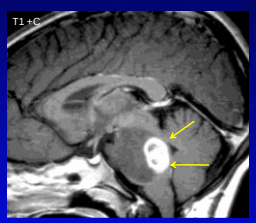
Diffuse intrinsic brainstem glioma

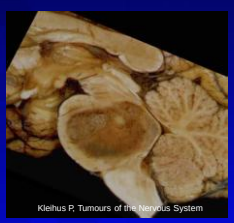


Basliary artery embracement, preserved fiber tracts, minimal enhancement

Boy, 9yrs

Diffuse intrinsic brainstem glioma



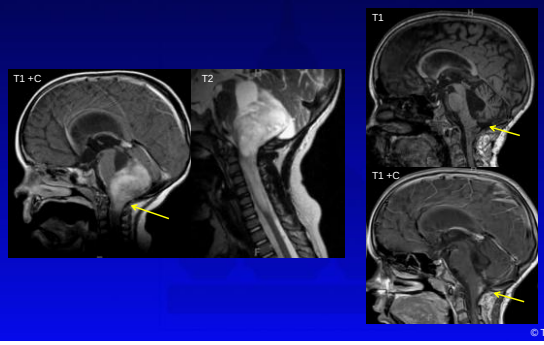


Variable enhancement on follow up, dedifferentiation (WHO III ~> IV)

Posterior exophytic glioma

- Located at cervicomedullary junction
- More favourable prognosis, median survival > 5yrs
- Most frequently, pilocytic astrocytoma
- Extend into IV ventricle
- Almost no infiltrative components
- Long history of non-specific headache and vomiting. Lower cranial nerve deficits, impaired speech and swallowing. Torticollis due to tonsillar herniation
- At least partial surgical resection is possible

Posterior exophytic glioma

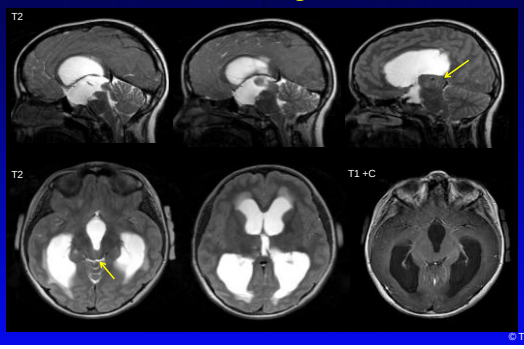


Focal tectal glioma

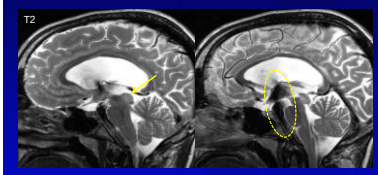
- Well demarcated low grade glioma
- Tectal plate
- Good prognosis, median survival > 7 yrs

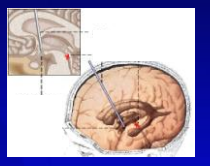
- Present with increased ICP due to obstructive hydrocephalus
- Internuclear ophtalmoplegia, Parinaud's syndrome
- Hydrocephalus usually treated with 3rd ventriculostomy

Focal tectal glioma



Focal tectal glioma

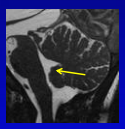




Treatment: 3rd Ventriculostomy

Ependymoma

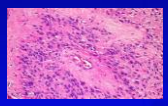
- 4th most common posterior fossa tumor (10-15%)
- Peak incidence 3-5 yrs, up to 18 yrs.
- 1:1.5
- Arise from ependymal lining of IV ventricle (esp. velum medullare posterior)
- Typically respect ventricular system
- Tumor extension along ventricles and their outlets (Magendie/Luschka)



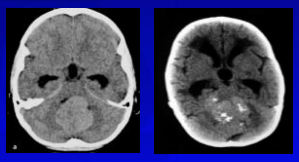
- Present with signs of increased intracranial pressure (obstructive hydrocephalus)
- Ataxia and cranial nerve palsy

Ependymoma

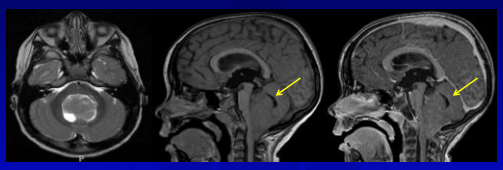
- CSF-seeding to spinal canal may occur
- If CSF-seeding is seen, anaplastic ependymoma should be suspected
- High cellularity ~> hyperdense on CT



Perivascular pseudorosets

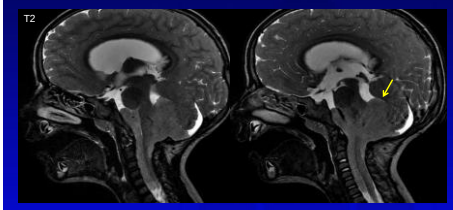


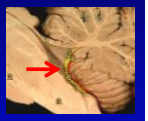
Ependymoma



Epicenter in fourth ventricle

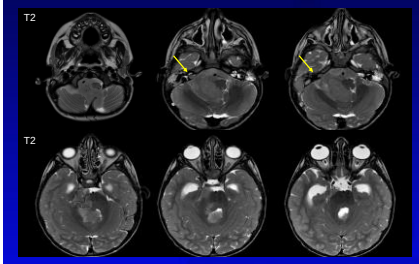
Ependymoma

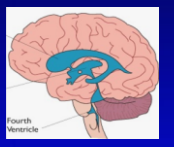




Ependymoma frequently originates from velum medullare posterior

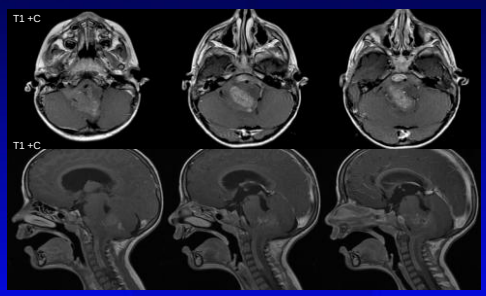
Ependymoma





Extension along ventricular system (even into internal auditory canal)

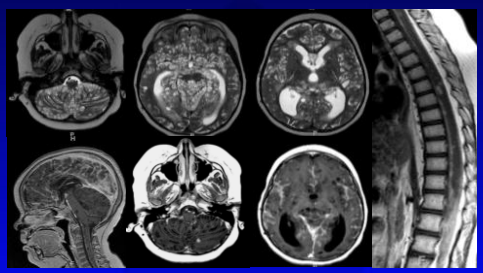
Ependymoma



Moderate, patchy contrast enhancement

Finally

- Metastases ~> primary brain tumors



M, 4Y, malignant neuroectodermal tumor

Summary

- Is it really a tumor?
- What is the most likely diagnosis?
- Is surgical resection an option?
- Use your statistics
- Be prepared for the unexpected
- Consider non-neoplastic etiologies
- Do not forget the spine
- Get the best anatomical and functional image quality

Rene Margrit