

Posterior fossa tumours in Children:

1. Medulloblastoma:

- The most common paediatric brain tumour (25% of paediatric brain tumours and 1% of tumours in adults).
- Most commonly arises from the vermis, however in 18% can arise from cerebellar hemisphere. Solid usually soft fills the fourth ventricle. In 15% desmoplastic, in 15% there is invasion of the floor of the fourth ventricle. Histology as above.
- Presentations: Hydrocephalus, neck pain and tilt and occasionally back pain and limb weakness secondary to spinal metastases and long tract signs secondary to brain stem invasion
- Management:
 - A. CT (hyperdense), MRI (T1-iso or hypointense, T2-hyperintense, -enhancing tumour filling the fourth ventricle, usually midline but can be hemispheric). MRI spine to exclude metastases.
 - B. Management of hydrocephalus. If the child comes with hydrocephalus but still conscious- start on dexamethasone, get MRI scan brain and spine and do craniotomy and excision of the tumour on the next available list. Insert EVD before the craniotomy. If the patient comes with decreased level of consciousness, start steroids, insert external drain, and avoid overdrainage (upward herniation). Definitive surgery on the first available list. Leave the drain few days after the operation. Only 30% of children require permanent shunt. Some surgeons still insert a shunt before the operation (risk of upward herniation, shunt complications, dissemination into peritoneal cavity (rare). Always send CSF for cytology.
 - C. Insert EVD, craniotomy, craniectomy in prone or lateral position, ultrasonic aspirator, EMG facial nerve and abducens nerve if there is brain stem invasion. Aim at total macroscopic removal. Leave tumour that is attached to the floor of 4th ventricle. Water tight closure of the dura. Leave EVD in for a few days. If patient is dependent on the drain for more than 10 days consider shunt (only 30% require shunt).
 - D. Postoperative radiotherapy in patients >5 years (**doubles survival**), Chemotherapy in younger children (**variable results**)
- Collin's law: if patient survives time equals the age at presentation + 9 months =cure (not always)
- Patients with medulloblastoma are classified into **average and high risk** based on age, extent of resection and the presence and absence of brain metastases:
 1. **Average risk: age >3 years, residual tumour on postoperative MRI < 1.5 cm² and absence of metastasis.** 5 year progression free survival 90%+-6%. (Packer et al 1994). **CCG-70% 5 year survival.**
 2. **High risk: age <3 years, residual tumour > 1.5 cm² and M1-4** (M1=positive CSF for cytology before surgery or >10 days after, M2= nodules in cranial SA space, M3= nodules in spinal SA space and M4=distant metastasis). **5-year progression free survival 67%+-15 %.** (Packer et al 1994). Modified Chang classification (TM). Original classification included TNM. **CCG- 57% 5year survival for M1 and 40% for M2.**

- Molecular genetics (**expression of ErbB 2 oncogene in 50% of cells** is associated with poor survival, **PDGFR** is associated with high incidence of metastasis, **TRKC is associated with better survival**).
- Treatment includes maximal safe surgical resection followed by CSI (**3600 cGy**) and boost dose to the posterior fossa to **5400 cGy** with **vincristine** given concurrent with radiation (for children > 3years) and postradiation chemotherapy (vincristine, prednisone and lomustine). Children's cancer group protocol. Event free survival was 74.2%.
- Radiotherapy is associated with the following risks:
 1. **Cognitive impairment** develops in all children (IQ decreases from 101 to 91 in average in children younger than 7 years)
 2. **Neuroendocrine dysfunction** (growth failure, hypothyroidism, precocious puberty and adrenocortical dysfunction).
 3. **Hearing deficit** (radiation to the auditory apparatus and ototoxic chemotherapy (cisplatin). 15%. Can be reduced by using oxaliplatin (less ototoxic) and 3 dimensional conforming radiation delivery techniques.
 4. **Secondary malignancy** (meningiomas, GBM, leukaemia) 5 times increase in the incidence comparing with controls.
 5. Psychological side effects (short stature, decreased hearing, thin hair, ataxia affects the child s social interactions.
 6. Radiation necrosis

2. Cerebellar astrocytoma:

- 15% of paediatric brain tumours, 30% of paediatric posterior fossa tumours
- Pilocytic astrocytoma in 70-80%, fibrillary astrocytoma in 20%, AA-3%, GBM <1%
- **Classically hemispheric cystic tumour with enhancing intramural nodule**, but can have enhancing wall or can be solid (90% of solid tumours are located in the vermis) **About 40% of cerebellar astrocytomas are in the vermis**
- . Presentations A. hydrocephalus with headaches, vomiting , papilledema and rectus palsy B. occipital headaches and neck tilt or torticollis may indicate impending tonsillar herniation
C. Cerebellar signs (dysmetria and incoordination “deterioration of hand writing” in case of hemispheric lesions or truncal ataxia “regression of walking” in vermal lesions.
- Management A. CT , MRI : Classically hemispheric cystic tumour with enhancing intramural nodule, but can have enhancing wall or can be solid (90% of solid tumours are located in the vermis) .About 40% of cerebellar astrocytomas are in the vermis
B. Management of hydrocephalus as for medulloblastoma
C. Surgical removal of the tumour:
 1. In the case of a cyst with enhancing nodule-remove the enhancing nodule only,
 2. In the case of solid tumour-remove all solid tumours.
 3. In the case of cyst with irregular enhancing wall one should aim at removing the enhancing wall

4. Regularly enhancing wall could be due to tumour or vascular hyperplasia as demonstrated in two recent studies. Intraoperative frozen section of the wall could help in the decision making

All patients need postoperative MRI scan within 3 days looking for residual tumour

D. If the tumour is low grade and was totally or subtotally removed-Follow up by MRI at 6 month then yearly interval. If the tumour reoccurs reoperate and aim at total removal. If the tumour is high grade give postoperative radiotherapy if child is older than 5 years or chemotherapy for younger children. In this case image the spine and take CSF for cytology to decide on local or craniospinal radiation.

3. Ependymoma:

- 10% of brain tumours in children, 2/3 in the posterior fossa. These tumours arise from ependymal cells around the ventricles or from ependymal rests
- Presentations (Hydrocephalus, vomiting due to irritation of area postrema, neck tilt “ tonsillar impaction in the upper spinal canal”, cerebellar signs and cranial nerve and long tract signs in case of brain stem invasion (rare).
- Arise from **ependymal cells or rests composed** of polygonal cells with **rosettes and Pseudorosettes. Poor correlation between anaplastic features and prognosis. see histology above**
- Diagnosis CT, MRI (**Mostly midline tumours arising from the brain stem and filling the 4th ventricle, tend to extend through foramen of Magendie into the upper cervical canal and through foramen of Luschka into the C-P angle tend to encase blood vessels and nerves**). Inhomogeneous enhancement. **Calcification is common** If possible do spine MRI before surgery or in the first 3 days after (CSF dissemination in 17%)
- Management: Start steroids, management of hydrocephalus (argue the options), prone position, head pins in children older than 3 years, 20-40 lb, occipital burr hole+-insertion of EVD, midline incision, occipital craniotomy, opening the dura through Y shaped incision, Be aware of occipital and circular sinus in children, central debulking followed by tumour removal, the part attached to the brain stem or encasing the nerves and vessels is left behind, haemostasis, water tight dural closure. For the role of electrophysiological monitoring of BAER, EMG Recording of facial , hypoglossal, abducens nerves read Albright’s page 616-618
- Complications of posterior fossa surgery:
 1. Positioning (pressure necrosis of the retina, meralgia paresthetica, brachial plexus injury, injury to the eyes by antiseptic solutions, pin complications “infection, extradural haematoma, air embolism”)
 2. Air embolism (**subclinical 26% in sitting position and 10% in prone position**): Monitor **end tidal CO2, precordial Doppler**, oesophageal stethoscope, some centres use oesophageal echo (most accurate), central venous line. In case of suspected air embolism emerge the field with water, wax the bone, 100% oxygen, head down if possible, aspirate the right ventricle through the central line, cardiac medications to support the heart and if CPR is needed close the wound with large sutures or sponge and Opsite turn to supine position.
 3. Postoperative haematoma
 4. Hydrocephalus: repeated LPs or V-P shunt if the patient continues to have headaches, decreased level of consciousness

5. Pseudomeningocele: LPs. If it doesn't resolve consider V-P shunt
6. Meningitis bacterial or aseptic (chemical). Exclude bacterial meningitis, restart steroids
7. Pseudobulbar palsy and cerebellar mutism **5-20%** of surgery for large vermian tumours (irritability, emotional lability, ataxia, mutism, hemiparesis occurring 1-2 days after surgery). Resolves in most cases in few weeks to 6 months. The anatomical substrate responsible for this syndrome is not known (dentate nucleus, middle cerebellar peduncle?)
8. Lower cranial nerve injury with the need for tracheostomy and gastrostomy (parents of children with posterior fossa tumours should be warned about this possibility)

- Adjunctive therapy 1. Radiotherapy (4500-5600 cGY) is indicated for
 - a. Large residual tumour
 - b. recurrent tumour
 - c. Anaplastic features on histology

In the case of CSF dissemination, craniospinal irradiation is indicated

2. Stereotactic radiosurgery: for local residual or recurrent tumour
 3. Chemotherapy: for young children with residual or recurrent tumours. Some studies showed improved survival with cisplatin based chemotherapy, other studies showed no effect.
- **5 year survival is around 60%**. Prognostic factors are
 1. The extent of surgical resection was the only clinical factor that was associated with survival in some studies
 2. Tumour grade (patients with high-grade tumours have significantly poorer survival compared with those with low-grade tumours in one study 71% vs., 29% (controversial). The main cause of death in ependymoma patients is intracranial failure at the primary site
4. Atypical teratoid/rhabdoid tumour: CP angle, highly malignant in children younger than 3 years (surgery+ chemotherapy). The majority die within 2 years.
 5. Hemangioblastoma: cerebellar cystic with enhancing nodule. In 30% solid. Rare in children .Von- Hippel- Lindau syndrome needs to be excluded
 6. Epidermoid, dermoids and arachnoid cysts : CP angle
 7. Acoustic schwannoma: rare in Children. Investigate for NF2.
 8. Others (metastasis, chondrosarcoma, meningioma) are rare in children