Optic Pathway Gliomas, Germinomas, Spinal Cord Tumours

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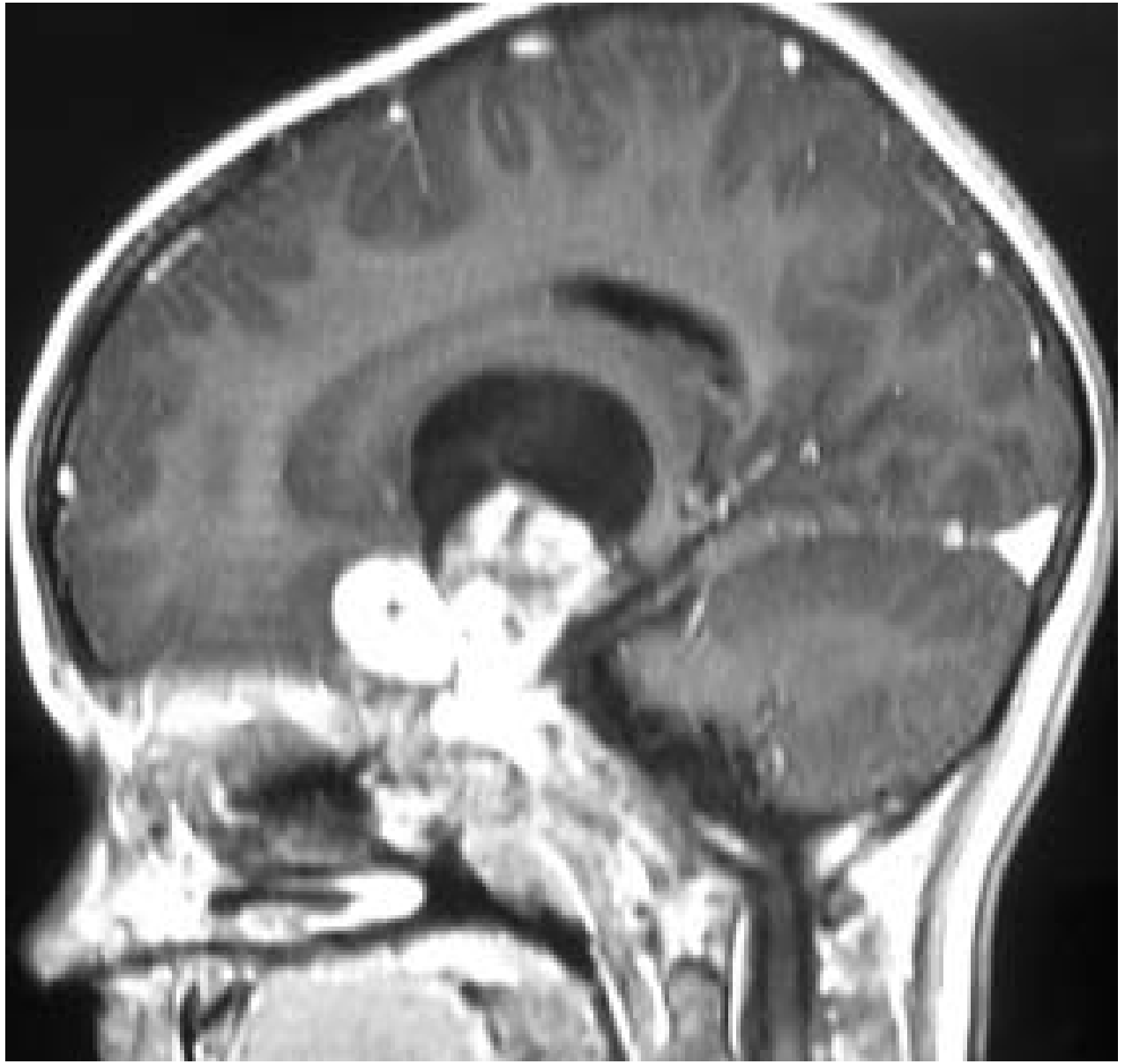
Visual pathway gliomas

Glioma of the optic chiasm.
T1-weighted MRI with gadolinium enhancement, showing intense irregular uptake of contrast.
The tumour extends posteriorly into the third ventricle and anteriorly under the left frontal lobe.



Optic pathway gliomas (OPG):
classification, NF1 and screening, overall approach to management

- 3-5% of primary intracranial neoplasms in children; 90% of OPGs occur in children
- **Histology:** typical pilocytic astrocytomas. Malignant change rare.
- **Classification**
  - 25-33% are pre-chiasmatic (i.e. optic **nerve** gliomas), usually intraorbital. Remainder involve chiasm and tracts +/- third ventricle
  - 15-35% occur in children with NF1with different biology – lower incidence of progression of chiasmatic gliomas. Different pattern of growth with expansion or inclusion, rather than invasion, of nerve.
  - OPGs less likely to progress in NF1 (e.g. 2/17 vs 12/19 in one series) but very long term event free survival not significantly different (e.g. around 60% 25 yr survival)

MR Screening for optic glioma in NF1 is of doubtful value (i.e. controversial) and management recommendations may not apply to asymptomatic gliomas found on screening: MONITOR VISION.

Overall approach to management

- Progression generally slow. Spontaneous regression sometimes. Progression more likely in younger children and uncommon after age of 6-8 years.
- Management decisions always need to weigh treatment toxicity vs benefit and put off until tomorrow treatment that is not needed today.
- Visual outcome is key but difficult to monitor, especially in young children
- Management of endocrine aspects, including growth, also vitally important.

Visual pathway gliomas

Anterior optic (nerve) gliomas

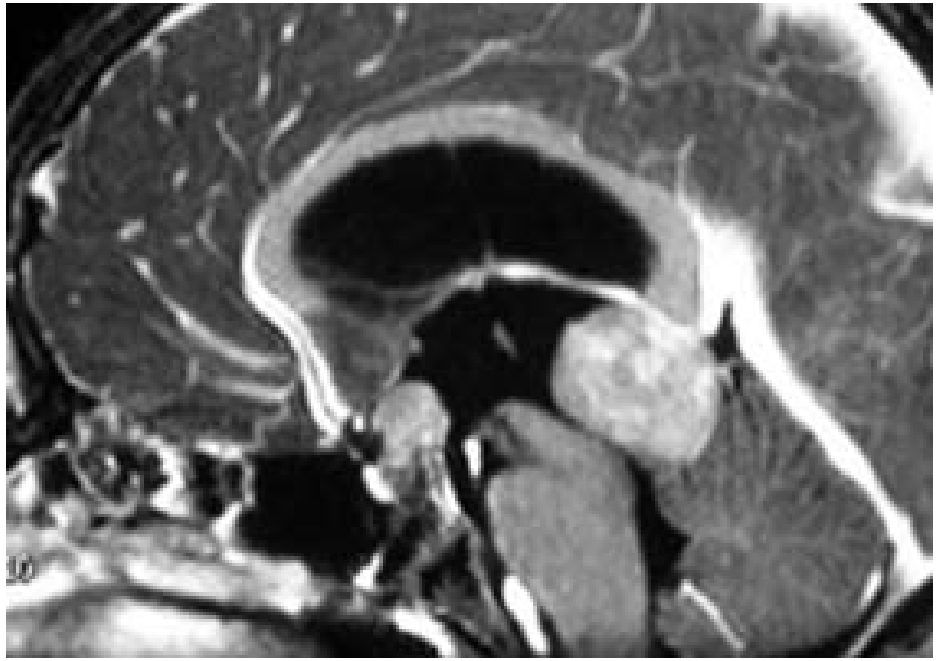
- Isolated anterior optic nerve tumours usually seen in NF1; rarely, if ever, progress to involve chiasm.
- Present with proptosis and or monocular visual loss (easily missed as children adjust to loss of vision). Fundoscopy: papilloedema or optic atrophy
- Management
  - Observe if asymptomatic
  - ? Chemotherapy (see next slide) if progressing but has useful vision
  - Excision if/when no useful vision: excellent outcome.

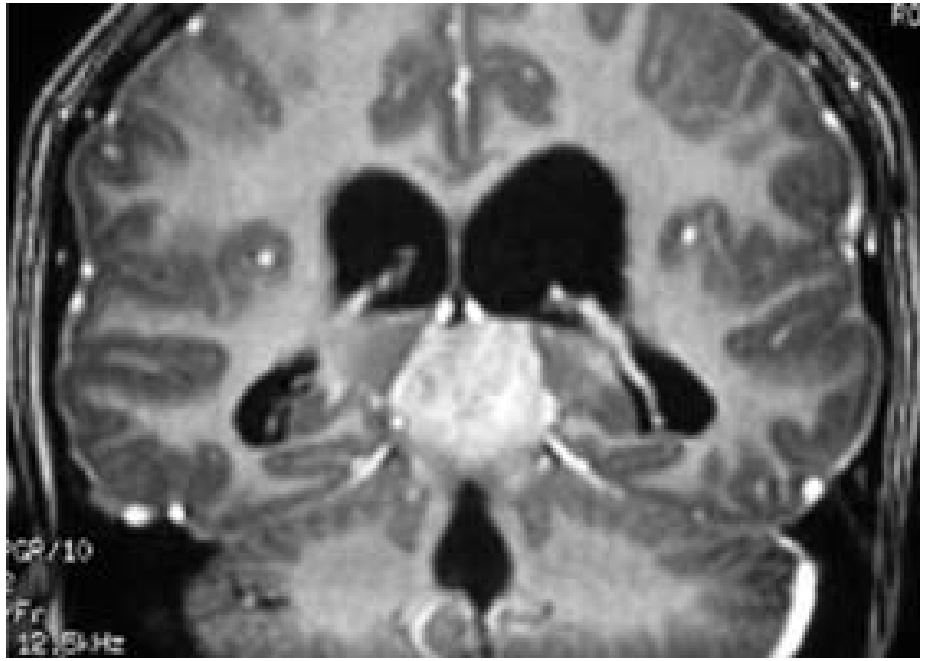
Visual pathway gliomas

Posterior optic (chiasm and tract) gliomas

- Present with loss of visual acuity, e.g. scotoma plus contralateral visual field defect. May mimic optic neuritis.
- Pendular nystagmus secondary to visual loss only under age of 2 years.
- Rarely monocular nystagmus mimicking spasmus nutans.
- May present with diencephalic (Russell) syndrome or obesity, DI, hypogonadism, precocious puberty (NF1), hydrocephalus (not NF1)
- Fundoscopy: usually optic atrophy
- Management
  - Asymptomatic and static: observe
  - Severe functional deficit or tumour progression clinically or on imaging:
    - Non-NF1 under 8 years or NF1: Chemotherapy for with carboplatin and Vincristine (or Vinblastine) to delay radiotherapy. Tumour response in up to 80%. Then radiotherapy for progression if non-NF1.
    - Non-NF1 aged 8 years or more: conformal photon or proton therapy (? Try chemo first)

Germ cell tumours





Pineal germinoma. (Left) T2-weighted sagittal MRI after gadolinium enhancement shows a tumour of heterogeneously high signal compressing and pushing down and forward the quadrigeminal plate causing hydrocephalus.
(Right) Frontal cut showing the mass and hydrocephalus from aqueductal stenosis.

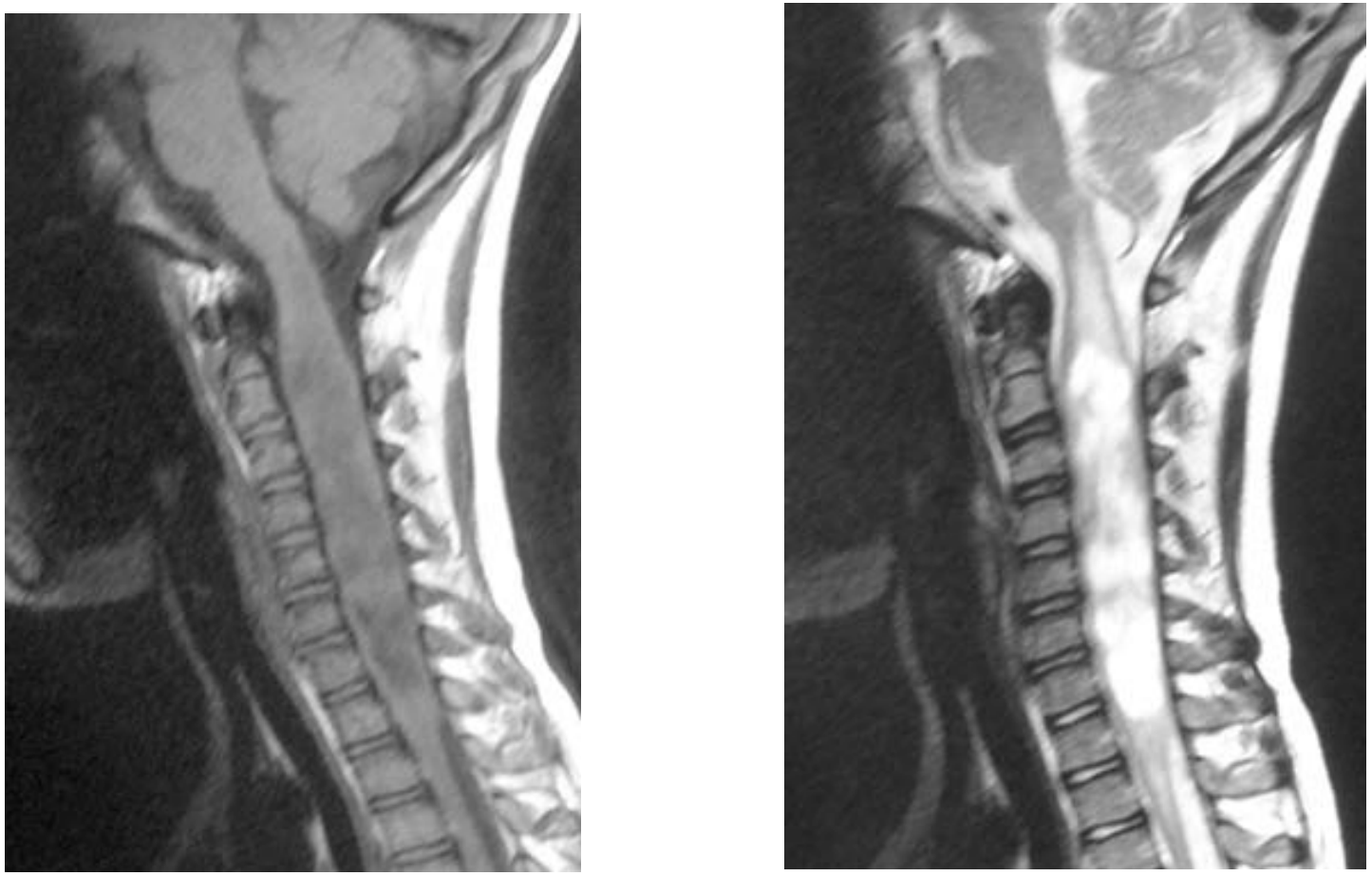
Germ cell tumours: presentation, imaging and histology

- Presentation:
  - symptoms of hydrocephalus
  - Parinaud's syndrome (complete syndrome= loss of upgaze, pupils unresponsive to light, ptosis)
  - Polydipsia, polyuria and/or precocious puberty
- Imaging
  - Commonest pineal region tumour. Peak incidence early in second decade
  - Maybe suprasellar, bifocal or 'ectopic' (= invading floor of 3rd ventricle without pineal lesion)
  - Iso- or hyperintense and uniformly enhancing with contrast on T1 +/- calcification
  - Spread around 3rd and to lateral ventricles (enhancing rim). Distant metastases rare.
- Histology:
  - Germinomas, (33-50% of all pineal tumours) divided into secreting and non-secreting by blood & CSF αFP and βHCG.
  - Non-germinomatous: embryonal cell Ca, teratoma, choriocarcinoma, endodermal sinus tumours

Germ cell tumours: management

- Treat hydrocephalus
- Measure tumour markers
- Biopsy (endoscopic) **only if tumour markers negative**. Excision rarely appropriate and has significant morbidity, esp if 3rd ventricle involved.
- **Stage** with brain and spinal imaging and markers
- Germinomas: **radiosensitive** and >90% cured by low dose radiation to tumour bed plus periventricular field as primary treatment. Little or no evidence of long term cognitive problems. **Also chemosensitive**: chemotherapy plus irradiation for recurrence preferred by some
- Non-germinomas: require more intensive treatment, higher recurrence.
- Avoidable risks Risk factors for poor outcome: **unnecessary biopsy, incomplete staging,**
- Endocrinology of key importance

Spinal cord astrocytoma



Intramedullary astrocytoma of the spinal cord. (Left) T_1-weighted MRI showing enlarged cervical cord with multiple low-signal areas. (Right) High signal from the same areas on T_2-weighted sequence.

Spinal cord astrocytoma: presentation

- **Back pain** (diffuse/radicular, esp. nocturnal, **may remit**) +/- rigidity.
- **Back deformity,** typically scoliosis (may be mild/absent)
- **Segmental weakness** (Limp/stiffness/claudication may be longstanding/interpreted as clumsiness/femoral anteversion etc) + UMN/LMN/mixed signs
- Urinary urgency → retention/incontinence as late features
- May mimic transverse myelitis/irritable bowel syndrome
- May present as hydrocephalus (meningeal fibrosis, haemorrhage from tumour, basal arachnoiditis)
- **Presentation notoriously subtle and easily dismissed as trivial**

(see Wilne & Walker, Education and Training edition of Arch Dis Child 2010)

- **Spinal cord compression is as urgent as ↑ICP**

Spinal cord astrocytoma: investigation and management

- Plain X-Ray – may show scoliosis, bony destruction/erosion, widening of canal
- MR investigation of choice – tumour may be difficult to distinguish from inflammation
- Management: total or subtotal excision if possible.
- Chemotherapy (e.g CisPlat, VCR) may improve outlook if remnant progresses
- Low grade astrocytomas may do well even with subtotal resection
- High grade tumours (e.g. glioblastomas) mostly do poorly

END