**Investigations**

1. **Blood testing** → mainly to exclude other causes of headache or investigation complications of tumours:
   * **Clotting** → bleeding disorders
   * **Calcium and U&Es** → may be abnormal in **hypercalcaemia or SIADH**
   * **ESR/CRP** → exclude other causes of headache
2. **Imaging** → diagnosis largely relies on this:
   * CT
   * MRI → good for posterior fossa tumours
   * Spine imaging → may be required for lymphoma
3. **Lumbar puncture** → only after CT
4. **Biopsy** → occasionally considered. Gives definitive diagnosis and histological types

**Differential diagnosis for a CNS mass/lesion**

|  |  |
| --- | --- |
| Cause | Distinguishing features |
| Tumour (1o or 2o) | Presentation over a protracted period (symptoms as above), WBC, ESR and CRP generally normal. MRI demonstrates a heterogeneous appearance. Unlikely to produce fever of meningism |
| Aneurysm | Posterior communicating artery aneurysm causes a painful 3rd nerve palsy |
| Abscess | May be associated with other infections (e.g. sinusitis, otitis media). Signs of meningism and fever are common. Blood abnormal (WCC, ESR/CRP). Imaging may reveal ring-enhancing lesions |
| Subdural haematoma | May follow trauma. More common in the elderly and those with clotting deficiencies, e.g. warfarin use, alcoholics |
| Granuloma | May be a history of TB |
| Cyst | Headaches, blunted consciousness, incontinence, weak legs, drop attacks |

**Clinical Manifestations**

* **Raised ICP 🡪 Secondary to the mass of the tumour or local brain oedema**
  + Headaches (worse on waking, lying down or coughing)
  + Vomiting
  + Papilloedema
  + Reduced GCS
* **Local Irritation** 
  + Can Lead to seizures which can be partial or secondary generalised
* **Focal neurological signs** 
  + Negative symptoms (i.e. loss of brain function)
    - Frontal lobe
      * **Motor:** hemiparesis, broca’s dysphasia
      * **Sensory:** anosmia
      * **Personality:** subtle changes including indecent, indolent, irritable and socially inappropriate
      * **Executive function and cognition:** apathy, concrete thinking, preservation (unable to switch train of thought), executive dysfunction (poor task planning), reduced verbal fluency
    - Temporal lobe
      * Dyphasia
      * Contralateral homonymous hemianopia (or upper quadrantopia if Meyer’s loop is affected) – Remember **PITS**
      * Amnesia
    - Parietal lobe
      * Hemisensory loss
      * Unable to recognise objects via touch
      * Dysphasia
      * Sensory inattention
    - Occipital lobe
      * Contralateral visual field defects
      * Polyopia **(**seeing multiple images)

**Treatment**

* **Surgery:** 
  + Used when the tumour can be resected or to debulk the tumour pre radiotherapy
* **Radiotherapy** is useful for symptomatic tumours that are unresectable
* **Radiosurgery** as for radiotherapy
* **Chemotherapy** can be used post op for gliomas or metastases or as the sole mechanism of treatment when surgery is impossible

**Prognosis**

* Prognosis is poor but improving. Overall there is **<50% survival at 5 years for CNS primaries**. Benign tumours are curable by excision.
* It is important to have **palliative care** involved early on to ensure relief of patients symptoms, e.g. N&V, pain, constipation. Aim to ensure the patient is comfortable at all times.

**Brain tumours**

* Represent about 2% of all tumours diagnosed in the UK
* Risk = 1 in 150
* Most commonly in those aged 50 – 70
* Most common types: **Glioma and Meningioma**
* **Risk factors:** 
  + Exposure to ionising radiation and certain industrial chemicals
  + Certain syndromes: **Neurofibromatosis, Von-Hippel Lindau syndrome** and **tuberous sclerosis**
* **Divided into high and low grade**

|  |  |
| --- | --- |
| **High grade (malignant)** | **Low grade (usually benign)** |
| * **Glioma** → from glial cells (**non-neuronal**), e.g**. ependymomas** (choroid plexus), **astrocytomas** (astrocytes) and **oligodendrocytogliomas** * **Primary cerebral lymphoma** → typically in **HIV patients** * **Medulloblastoma** → highly malignant, usually arises in the **posterior cranial fossa** from the cerebellum. Thought to originate from embryonic cells | * **Meningioma** * **Acoustic neuroma** → from Schwann cells of CN VIII. Common in **neurofibromatosis** * **Neurofibromas** * Pituitary and pineal tumours * **Craniopharyngomas** |

**Common secondary CNS tumours include:** Lung, breast (leptomeningeal deposits), stomach, prostate, thyroid, colorectal, melanoma, renal

**Management of raised ICP**

* A/B → may require ventilation. **Hyperventilation** can be used to immediately recused ICP
* C → correct hypotension, **elevate head of the bed to 30-40o**
* Medication →
  + **Acetazolamide / Mannitol (**or other osmotic agents) can be used to reduced ICP, however these may only be useful in the short term
  + **Topiramate** is used in idiopathic intracranial hypertension
  + **Dexamethasone** (**10mg IV** followed by **4mg/6hours**) is only useful in treating raised ICP **caused by cerebral oedema associated with tumours**
* **Fluid restrict**
* **Investigations**
* **Surgical intervention will usually be required**

**Huntingtons Disease**

* **Autosomal dominant neurodegenerative disorder caused by a CAG repeat within the huntington gene – degeneration of cholinergic and GABAergic neurones in the striatum of the basal ganglia (defect in chromosome 4)**
* **Clinical feature:**age 35 – 40 @onset, insidious then progressive signs, chorea 🡪 irritability 🡪 dementia 🡪fits 🡪 death
* **Neuropsychiatric symptoms:** personality changes, irritability, impulsiveness, dementia
* **Motor changes:** chorea, deficits in fine motor coordination, slowed saccadic eye movement

**Chorea, athetosis and dystonia**

* **Chorea**: irregular, random and variable movements which have a flowing or dancing quality which may appear semi purposeful

|  |  |
| --- | --- |
| **Acquired** | **Hereditary** |
| * **Post infectious** (**Sydenham’s chorea post rheumatic fever** – remember PECCS – polyarthritis, erythema marginatum, carditis, chorea, subcutaneous nodules) * **Polycythaemia rubra vera (JAK2, pruritis, splenomegaly etc)** * **SLE** * **Thyrotoxicosis** * **Drugs** → L-DOPA, phenytoin, neuroleptics | **Huntington’s disease**  A number of rare, inherited disorders |

* **Athetosis:** slower and more writhing in quality than chorea. It represents transition from one dystonic posture to another. It is typically associated with **congenital brain damage** (cerebral palsy). The **hands and feet** are typically affected
* **Dystonia:** involuntary, sustained,painful muscle contractions resulting in **abnormal posture.** Often caused by antipsychotic medications: classified as generalised or focal
  + **Focal:**
    - **Blepharospasm:** involuntary eye closure
    - **Oculogyric crisis:** eye rolled upwards (seen in post-encephalitic parkinsonism)
    - **Spasmodic torticollis** 🡪 scm contraction may turn the head to one side or move it forward or backward
    - **Laryngospasm**
    - **Trismus**
    - **Writers cramp**