CASE 1

- 65 year-old male with diplopia and headaches for 1 month

- On neurological examination: right VI nerve palsy
Glioblastoma multiforme
Pineal region glioblastoma multiforme

- Pineal tumors account for 0.4 to 8% of intracranial tumors
- In the pediatric population they represent 3-12% of brain tumors
- Median age is 22 years old, 1:3 male to female ratio
- Three types: germ cell tumors, pineal cell tumors, and glial tumors
- Germ and pineal cell tumors account for 78-86% of all pineal region tumors

## Pineal region gliomas

<table>
<thead>
<tr>
<th>WHO grade</th>
<th>Histopathology</th>
<th>Number of cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>I (53)</td>
<td>Pilocytic astrocytoma</td>
<td>49</td>
<td>14</td>
</tr>
<tr>
<td></td>
<td>Juvenile pilocytic astrocytoma</td>
<td>2</td>
<td>0.6</td>
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<tr>
<td></td>
<td>Subependymal giant cell astrocytoma</td>
<td>2</td>
<td>0.6</td>
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<tr>
<td>II (167)</td>
<td>Diffuse astrocytoma</td>
<td>86</td>
<td>24.6</td>
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<tr>
<td></td>
<td>Ependymoma</td>
<td>54</td>
<td>15.5</td>
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<tr>
<td></td>
<td>Oligodendroglioma</td>
<td>20</td>
<td>5.7</td>
</tr>
<tr>
<td></td>
<td>Pilomyxoid astrocytoma</td>
<td>1</td>
<td>0.3</td>
</tr>
<tr>
<td></td>
<td>Pleomorphic xanthoastrocytoma</td>
<td>6</td>
<td>1.7</td>
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<tr>
<td>III (64)</td>
<td>Anaplastic astrocytoma</td>
<td>32</td>
<td>9.2</td>
</tr>
<tr>
<td></td>
<td>Anaplastic ependymoma</td>
<td>29</td>
<td>8.3</td>
</tr>
<tr>
<td></td>
<td>Anaplastic oligodendroglioma</td>
<td>3</td>
<td>0.8</td>
</tr>
<tr>
<td>IV (65)</td>
<td>Glioblastoma multiforme</td>
<td>65</td>
<td>18.6</td>
</tr>
</tbody>
</table>

CASE 2

- 57 yo female with an intracranial mass since 2003 suspected of being a meningioma

- Patient denies any neurological symptoms

- Imaging evaluation showed slight increased size (a few millimeters) of the lesion

- Previous history of breast cancer
Cavernous hemangioma
Cavernous hemangioma

- Dural based cavernous malformations are rare and most commonly described in the middle fossa
- Other locations include: tentorium, sagittal sinus, tegmen tympani, Meckel’s cave and the convexity
- They rarely present with hemorrhage
- They are often misinterpreted as meningiomas
- They can show dural tail, calvarial erosion or hyperostosis

CASE 3

- 49 yo female with chest pain and headaches. EMS arrived 2-3 minutes latter, she became unresponsive and asystolic and required CPR. She was coded and intubated and she regained pulse. Pupils were 4 mm and fixed

- Arriving on the ER she was diagnosed as clinically brain death
Pseudosubarachnoid hemorrhage
• Pseudo-SAH reported with...

• brain swelling (anoxic–ischemic brain injury, hyponatremia, metabolic encephalopathy)

• pyogenic leptomeningitis or viral meningoencephalitis

• intrathecally administered contrast material

• leakage of high-dose intravenous contrast medium into the subarachnoid space

• bilateral subdural hematomas

• spontaneous intracranial hypotension
Confounding issues in pseudo-SAH

~10% of patients with aneurysmal SAH have a cardiac arrest, so that a cardiac-arrest history does not mean that CT finding of high attenuation in the basal cisterns or subarachnoid spaces is always indicative of pseudo-SAH.

“Thunderclap headache” has been reported in 15% of patients diagnosed with spontaneous intracranial hypotension, which is a recognized cause of pseudo-SAH.
• Suggested mechanisms of pseudo-SAHI

• In **severe brain edema**, the swollen brain compresses the dural sinuses, compromising the venous drainage from the brain and resulting in engorgement of the superficial veins, which stand out against the edematous low-attenuated brain parenchyma, mimicking SAH.

• In **spontaneous intracranial hypotension**, the sagging brain narrows the subarachnoid spaces (basilar cisterns, Sylvian fissures) and displaces CSF. The resultant subarachnoid spaces become relatively devoid of hypoattenuated CSF and fill with a larger fraction of meninges and blood vessels (middle cerebral arteries in Sylvian fissures; basal veins of Rosenthal in mesencephalic cisterns) that, by increasing their CT attenuation, contribute to the pseudo-SAH appearance.

CASE 4

- 8 yo male with recent strep pharyngitis, daily fevers (for approximately 3.5 weeks), intermittent diarrhea, sore throat, and oral/perianal ulcers, increased ICP, ataxia, headache

- Left femur lesion corresponding to fibrous dysplasia

- Elevated inflammatory markers and anemia
SWI no hemorrhage or calcification/ No enhancement post-Gd
Collections of foamy macrophages that contain PAS-positive granules (inset)
Whipple’s disease

- Rare systemic disease affecting gastrointestinal tract, heart or CNS. Approximately 60% of patients show improvement with antibiotics. CNS disease carries a high risk of relapse.

- **Agent is *Tropheryma whippelii***

- CNS MRI findings are involvement of hypothalamus, thalamus, quadrigeminal plate, periaqueductal gray matter and basal telencephalum. Lesions are usually bilateral, nonenhancing and without restricted diffusion

- Even more rare a vasculitis due to leptomeningeal fibrosis and thrombosis (as in our patient)

Case 5

- 6 yo female with headaches, vision changes, episodic aphasia, balance disturbances, dizziness, and vomiting in the last few days
Choroid plexus papilloma, WHO grade I
Choroid plexus papilloma

- Choroid plexus tumors (CTTs) are rare, with an annual USA incidence of 1-3 cases per 10 million people per year but they account for up to 20% of pediatric brain tumors arising in the first year of life.
- Lateral or third ventricle in children, often under 2 years of age; fourth ventricle or the cerebellopontine angle in adults.
- Histologically, choroid plexus may be classified as follows:
  - Choroid plexus papilloma, WHO grade I (60% of CTTs)
  - Atypical choroid plexus papilloma, WHO grade II (15%)
  - Choroid plexus carcinoma, WHO grade III (25%)
- Chance of recurrence is greater with atypical adenoma or carcinoma.
- Gross total removal is best chance for cure. Malignant progression is rare.
- Children with CP carcinoma (CPC) appear to have a high frequency of TP53 germline mutations in association with Li–Fraumeni Syndrome (LPS). This raises the question whether all children with CPC should be tested for TP53 germline mutations in order to institute screening to enhance early detection and treatment of subsequent cancers.

Case 6

- 72 yo male who has a 6-year history of tingling in right greater than left fourth and fifth digits

- Neurologic examination reveals mild hyperreflexia in the upper extremities
Ependymoma, WHO grade II, with ependymal rosettes and perivascular pseudorosettes
Ependymoma

- Intramedullar spinal cord tumors are rare (4-10% of all CNS tumors)
- Ependymomas and astrocytomas are the most common intramedullary tumors
- Ependymomas usually are cervical and have a plane separating them from the normal spinal cord
- Ependymomas present a tendency to be central in the spinal cord. Enhancement and hemorrhage can be seen. Syringohydromyelia is a common finding.

Case 7

- 33 yo female

- During pregnancy a pituitary tumor was discovered due to visual field defects (bitemporal hemianopsia), panhypopituitarism, and migraines.

- She delivered her baby 5 weeks previously.
01/15/2014 (during pregnancy)
Granulomatous hypophysitis
Granulomatous hypophysitis

• Hypophysitis is rare—estimated incidence of one case per 9 million people per year.

• Lymphocytic hypophysitis is the most common form and is characterized by an autoimmune lymphocytic infiltrate that can involve the anterior pituitary, the infundibulum and posterior lobe of the pituitary (infundibulo-neurohypophysitis), or the entire pituitary gland.

• Granulomatous hypophysitis (GH) is the second most common subtype and features widely distributed multinucleated giant cells, granulomas, lymphocytic infiltrates, and fibrosis.

• GH can be a primary phenomenon or secondary to systemic disease. Secondary causes include tuberculosis, sarcoidosis, syphilis, Langerhan’s cell histiocytosis, Wegener’s granulomatosis, Rathke’s cleft cyst rupture, and pituitary adenoma.

• In one series, 72% of patients were women; mean age was 44.

• Radiologically, 93% of patients had pituitary enlargement; 66% had suprasellar extension; 34% had a thickened infundibulum; and 24% had contrast enhancement.

• Majority of cases of GH become apparent due to mass effects from an enlarged pituitary gland. May also show pituitary hypofunction or diabetes insipidus.