

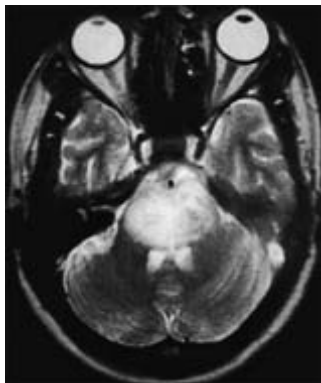
EANS/UEMS European examination in neurosurgery

Variants of questions with answers (compilation - Vyacheslav S. Botev,
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CASE HISTORIES IN BRAIN TUMORS

Case 1

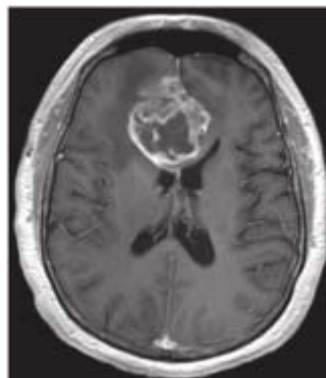
A 30-year-old male with a 3-month history of morning headaches, dizziness and facial numbness was noted to have bilateral papilledema and horizontal nystagmus.



1. What does the MRI scan show?
2. What is the most likely diagnosis?
3. What is the prognosis?

Case 2

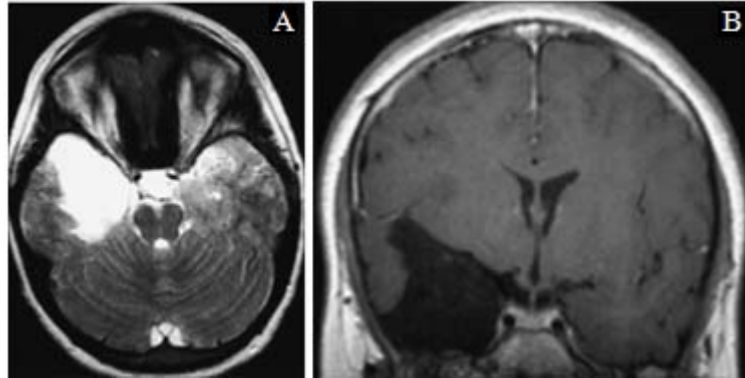
Characteristic appearance of tumor invasion of the corpus callosum on a T1-weighted image with contrast, axial view:



1. What is the classic presentation of this condition?
2. What is the diagnosis and how is it made?
3. What is the prognosis?

Case 3

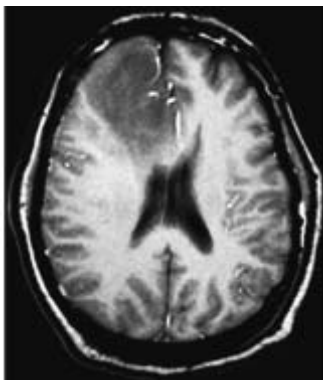
A 4-year-old male is referred to your office for evaluation of the lesion shown (A, B). His parents tell you that they discovered this lesion last year, but elected to follow it because he was asymptomatic. They are now very concerned, however, because he has not been eating well for the past 3 weeks, and that last week he had a seizure for the first time.



1. What is the most likely diagnosis?
2. What sequence of MRI would be helpful to exclude another type of congenital lesion?
3. What are these lesions composed of?
4. Where are they most commonly located?
5. What is the male-to-female ratio?
6. List the indications for treatment of these lesions.
7. Discuss the treatment options for this patient.

Case 4

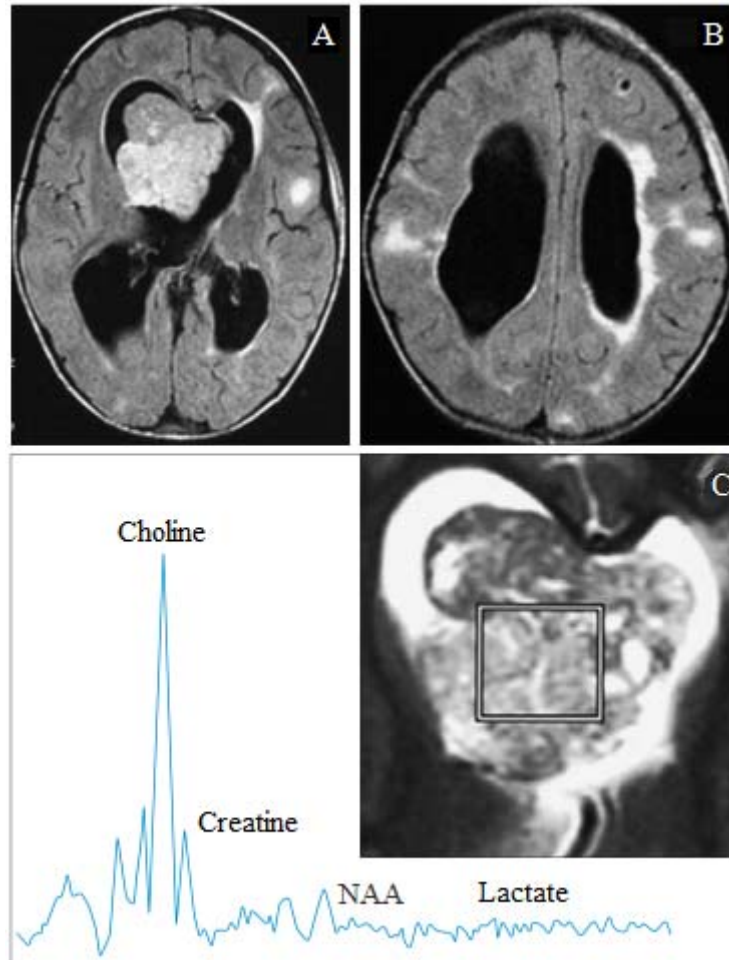
A 38-year-old male suffered a generalized seizure. This T1-weighted MR image was obtained after administration of gadolinium DTPA.



1. What are the findings?
2. What is the most likely diagnosis?

Case 5

A 15-year-old male with a history of mental retardation and generalized tonic-clonic seizures develops ataxia and a change in his seizure pattern despite stable medications. This prompts the brain MRI shown below (A–C). On physical examination, you notice numerous yellow-brown lesions in a malar distribution.



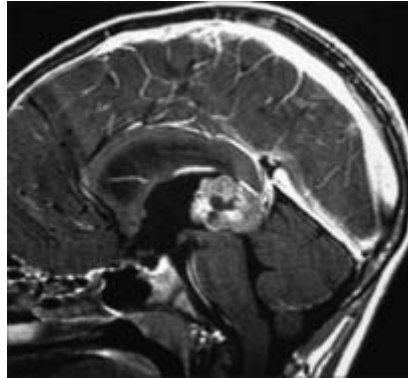
On physical examination, you notice numerous yellow-brown lesions in a malar distribution.



1. What is the most likely diagnosis of the lesion shown on the MRI?
2. How would MR spectroscopy assist with diagnosis?
3. What syndrome does this patient have?
4. How should this lesion be managed?

Case 6

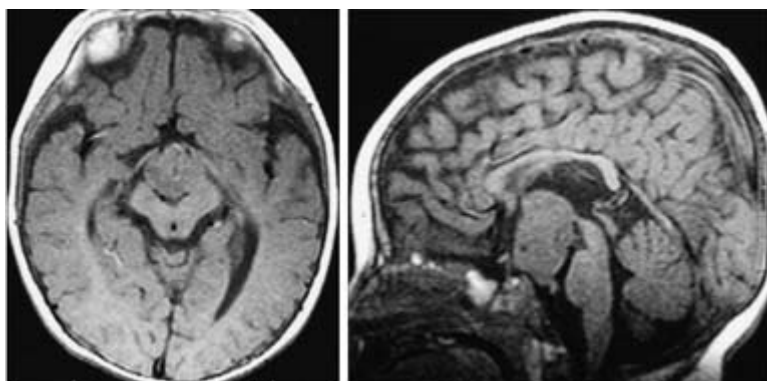
A 16-year-old male is referred to you with complaints of headache, nausea, and intermittent vomiting which have increased over the last 2 months. After handing you his recent contrast MRI, his parents emphasize the memory difficulties he has had over the past several months. His physical examination is remarkable for ataxia and Parinaud's syndrome.



1. What is Parinaud's syndrome?
2. What does the MRI show?
3. Give a differential diagnosis for this lesion.
4. What additional tests might you want to assist with both diagnosis and management?

Case 7

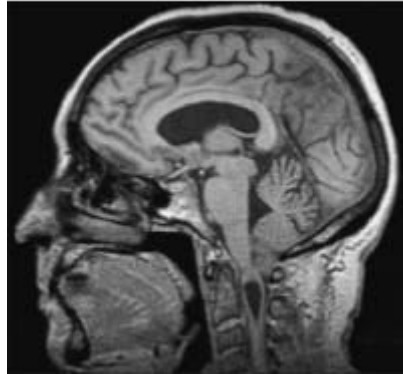
An 11-year-old male presents with precocious puberty and uncontrollable laughing episodes with subsequent generalization. Work-up includes a brain MRI.



1. What type of seizures are these?
2. What does the MRI show?
3. Discuss the treatment options for this lesion.

Case 8

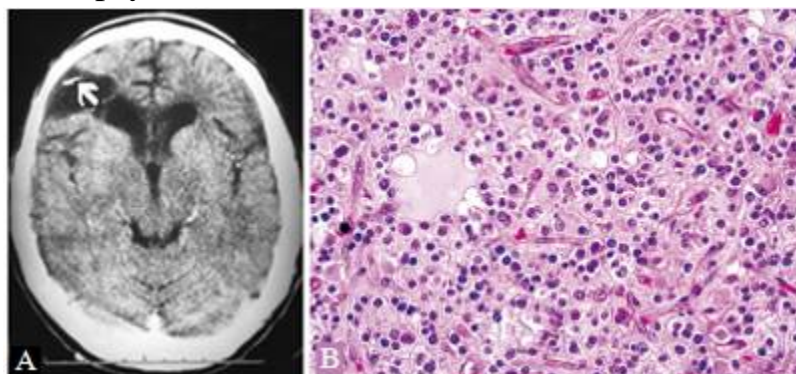
A 48-year-old female gave a 2-year history of severe bioccipital throbbing headaches radiating frontally, precipitated by coughing, sneezing, bending or lifting. The onset was sudden with rapid resolution after 15–30 minutes. There were no additional features to the headache for which she had taken simple analgesics, pizotifen and amitriptyline without good effect. Clinical examination was normal.



1. With what type of headache does the patient present?
2. What does the MRI scan show? Are the headache and MRI abnormality related?
3. Had the neuroimaging been normal what diagnosis would you give the patient? What treatment may provide symptomatic relief in this situation?

Case 9

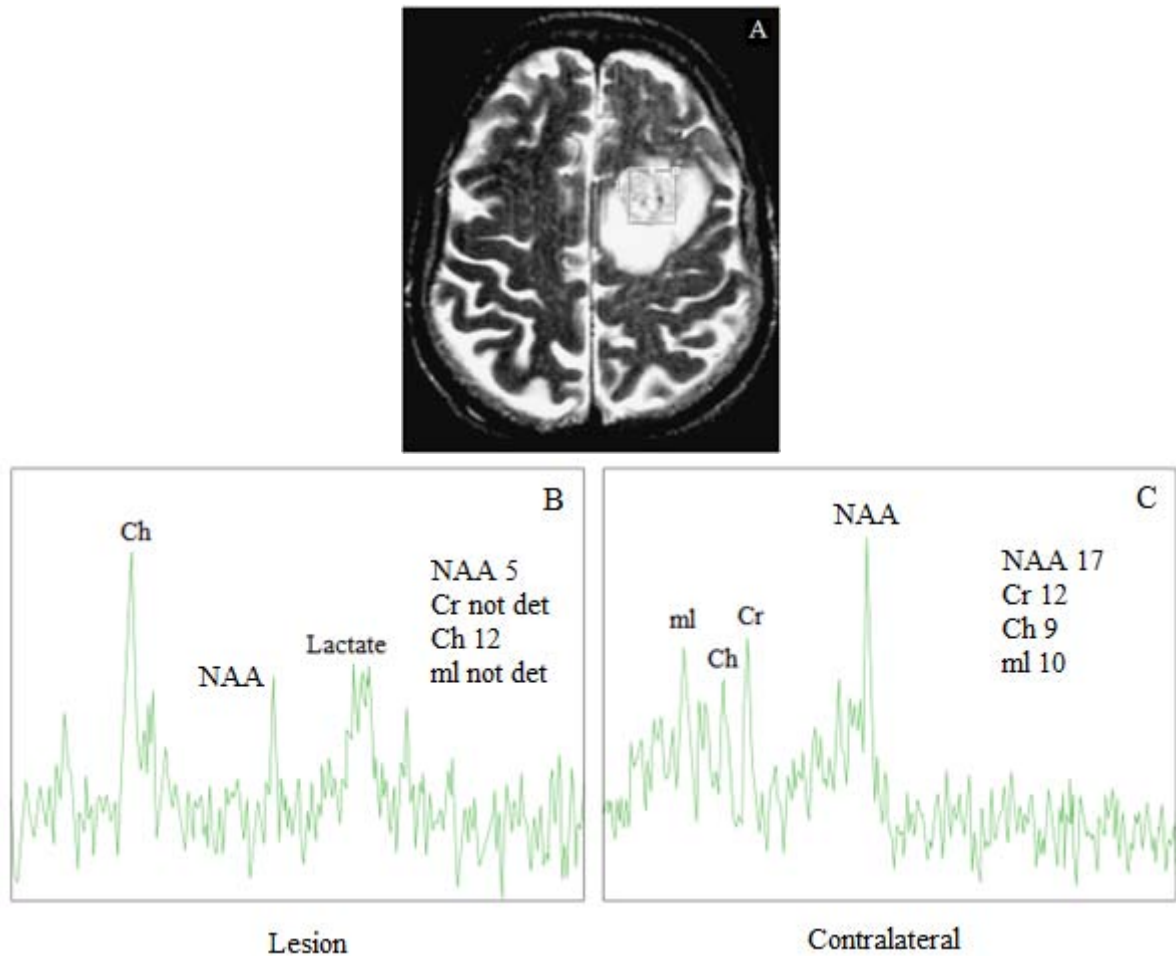
This is the CT scan of a 32-year-old male (A) presenting with right focal motor seizures and the biopsy of the lesion (B).



1. What does the biopsy show?
2. What treatment may be helpful in this condition?
3. What factors may predict a good treatment response?

Case 10

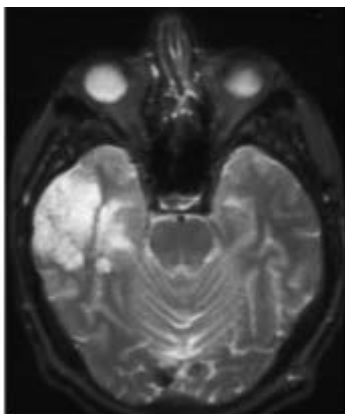
A 74-year-old right-handed surgeon presented with sudden onset aphasia.



1. What are the MRI and MR spectroscopy findings?
2. Are they compatible with a diagnosis of primary brain neoplasm?

Case 11

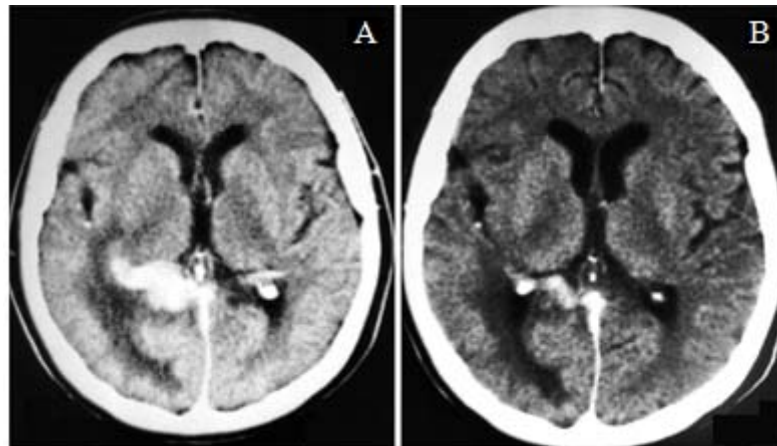
A 27-year-old male presented with a 20-year history of temporal lobe seizures that had been resistant to medication. An MRI was performed.



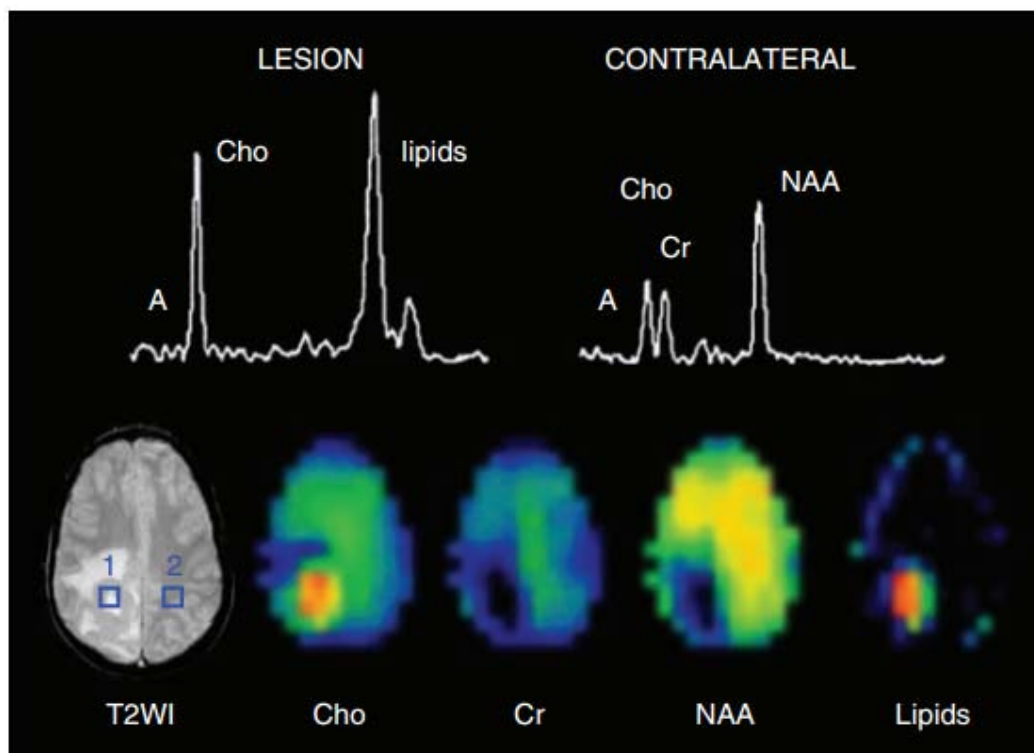
1. What does the MRI show?
2. What is the likely cause?
3. What is the treatment?

Case 12

A 59-year-old female was admitted to hospital with a 3-week history of intermittent confusion. She was systemically well and afebrile. There was no focal neurology. An infection screen was negative. A CT scan was performed (A), she was given some tablets and the CT scan repeated 4 days later (B) after considerable improvement.



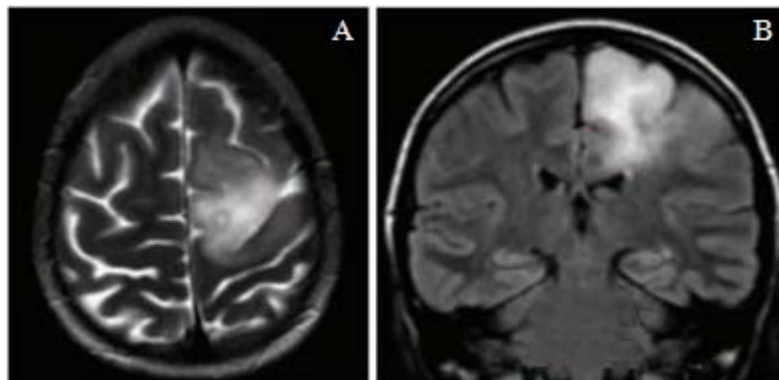
MR spectroscopy:



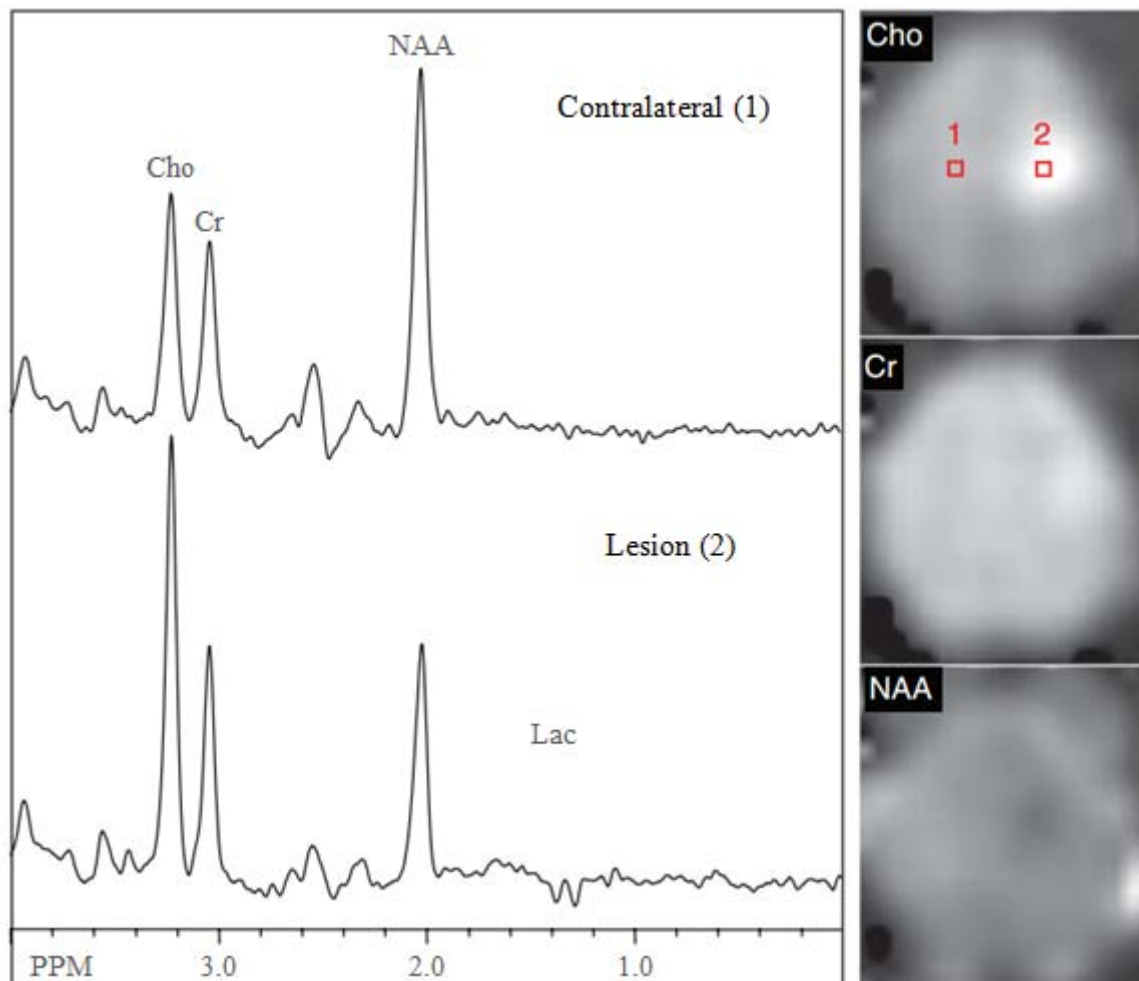
1. What is the likely diagnosis?
2. What treatment was she given?
3. What are MR spectroscopy findings?
4. What is European standard for the treatment of PCNSL?
5. What is the Sedan needle?

Case 13

Axial T2-weighted MR imaging (A) and FLAIR image (B) shows infiltrating mass in the left superior frontal gyrus of a 20-year-old woman presenting with a focal seizure of the right arm.



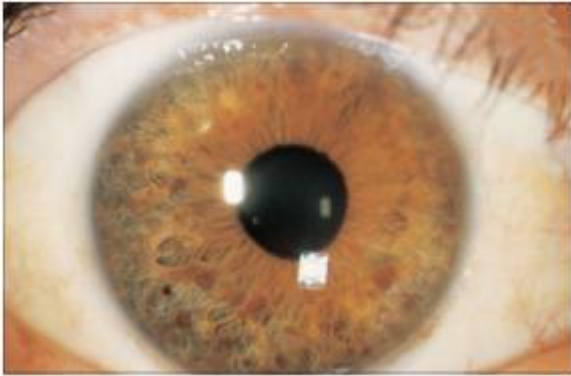
MR spectroscopy. Cho, Cr, and NAA maps with two selected spectra:



1. What is the most likely diagnosis of the lesion shown on the MRI?
2. How would MR spectroscopy assist with diagnosis?
3. How should this lesion be managed?

Case 14

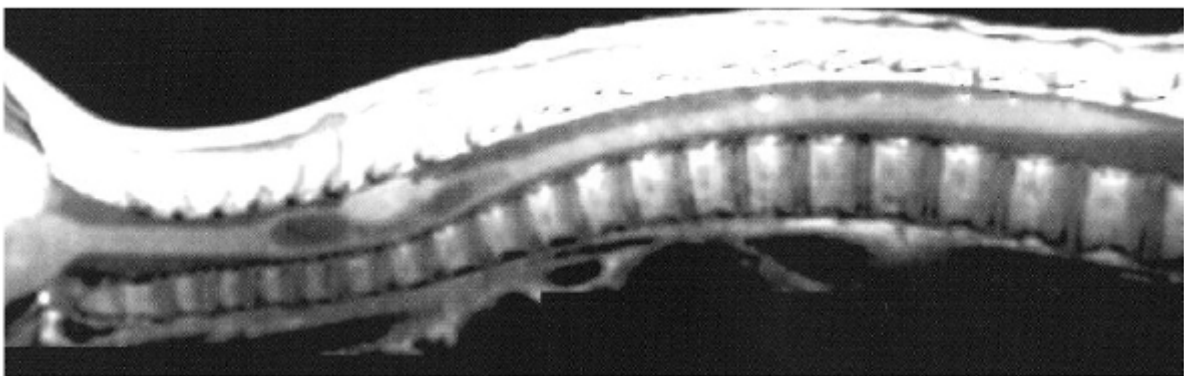
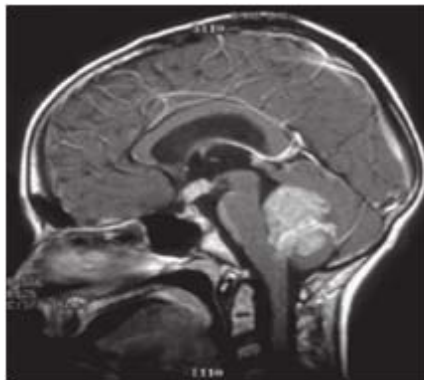
A 37-year-old female presents with complex partial seizures. Examination of her eyes reveals abnormalities of the iris.



1. What are the abnormalities shown, and what other abnormalities would you look for?
2. How is the condition inherited?
3. What could be the cause of her epilepsy?

Case 15

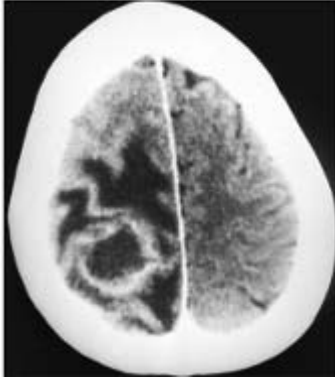
A 3-year-old boy develops ataxia and vomiting.



1. What are the imaging findings?
2. What is the most likely diagnosis?
3. What are the four most frequent tumors in the posterior fossa in children?
4. What is one of the most significant prognostic factors for long-term survival?

Case 16

A 52-year-old female with relapsed stage IVB non-Hodgkin's lymphoma was treated with a stem cell transplant 2 months ago. She then developed focal seizures and a weakness of her left arm and increasing lethargy. She was afebrile and did not complain of headache. A CT brain scan was performed.

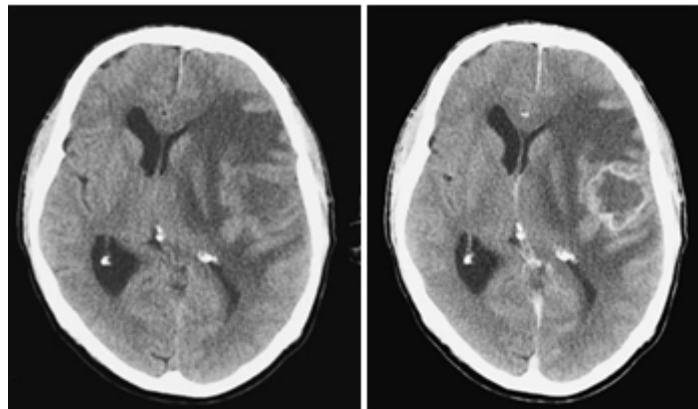


1. What is the differential diagnosis?
2. What are the principles of management?

Case 17

A 58-year-old man collapsed at home. In the emergency department, he was drowsy and dysphasic, with moderate right-sided weakness (MRC grade 4/5). His left pupil was 5mm, the right was 3mm, and both reacted to light. He had attended 2 weeks earlier because of difficulties with his speech.

1. What is the neurosurgical differential diagnosis?
2. A CT scan is performed (pre-contrast and post-contrast). Describe the abnormalities seen.

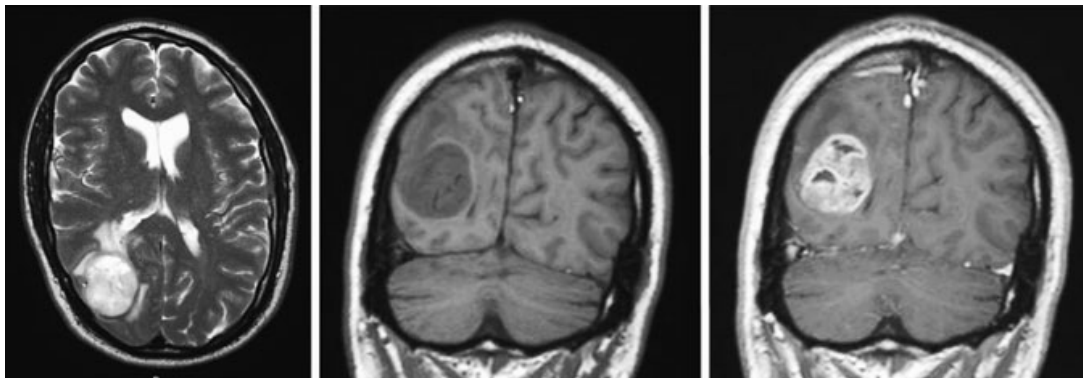


3. What types of edema occur in the brain and which does this patient have?
4. How should this patient be managed?
5. What is the "Stupp regimen"?

Case 18

A previously well 53-year-old man had developed a sudden onset of severe headache 5 days previously. The headache settled within hours and he did not seek medical advice at the time. He later consulted his GP who sought a neurological referral as he was concerned about an acute vascular migraine. The headache had resolved completely by then, but he had a left-sided homonymous hemianopia.

1. Where is the lesion presumed to be?
2. What is the differential diagnosis?
3. The MRI is shown. What is the likely diagnosis?

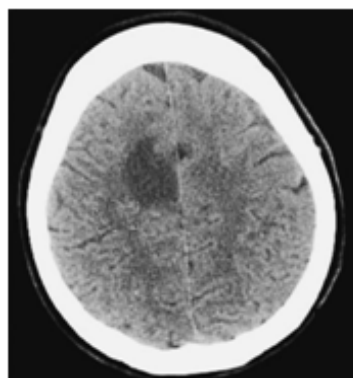


4. What factors influence prognosis if glioblastoma is the diagnosis?

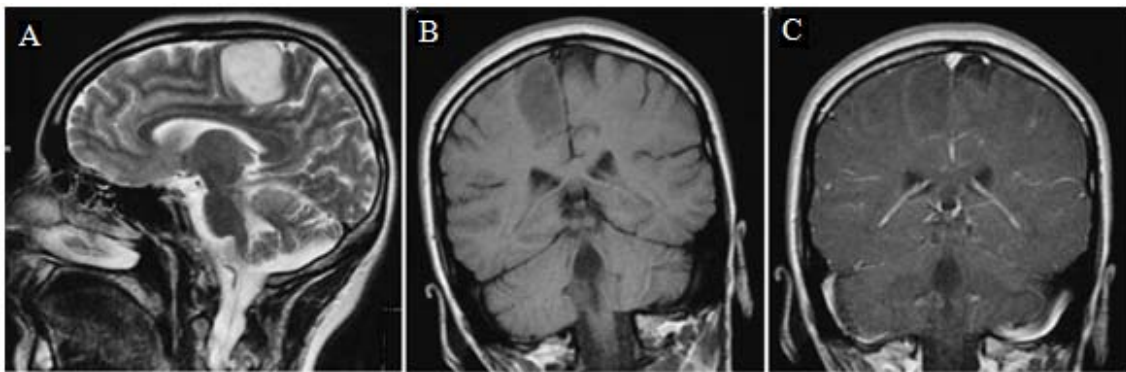
Case 19

A 27-year-old woman presents to the emergency department following a witnessed first grand mal seizure. She was working at her desk, fell to the floor, shook violently for 30 seconds, and was then unconscious. She bit her tongue but was not incontinent. In the emergency department 20 minutes later she is drowsy but responding to voice and obeying commands. Her left arm is weak.

1. What factors lower an individual's seizure threshold?
2. What is the significance of the new left arm weakness?
3. A CT brain is performed. What does it show?



4. Sagittal T2 (A) and coronal pre-contrast (B) and post-contrast (C) MRIs are shown below. What is the tumor location and what information does it provide?

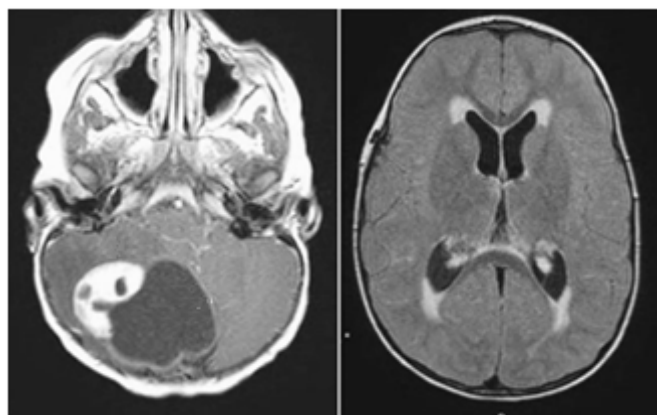


5. What are the management options?

Case 20

A 3-year-old boy has a 6-week history of motor decline. His parents report that previously he was running and playing normally but now is walking slowly and holding on to objects or people to support himself. His GP arranged a non-urgent pediatric outpatient appointment. However, for the past 2 days the child has been irritable and holding his head. This morning he vomited and so came to hospital.

1. What typical diagnosis does the history suggest?
2. What are the clinical findings in a patient with a posterior fossa mass lesion?
3. Explain why a cerebellar lesion causes incoordination ipsilaterally.
4. On examination this patient is alert but miserable. Examination of coordination and funduscopy are not possible. How should he be investigated?
5. An MRI is performed and shown below.
 - (a) What sequences are shown?
 - (b) Describe the abnormalities.
 - (c) What are the possible diagnosis?

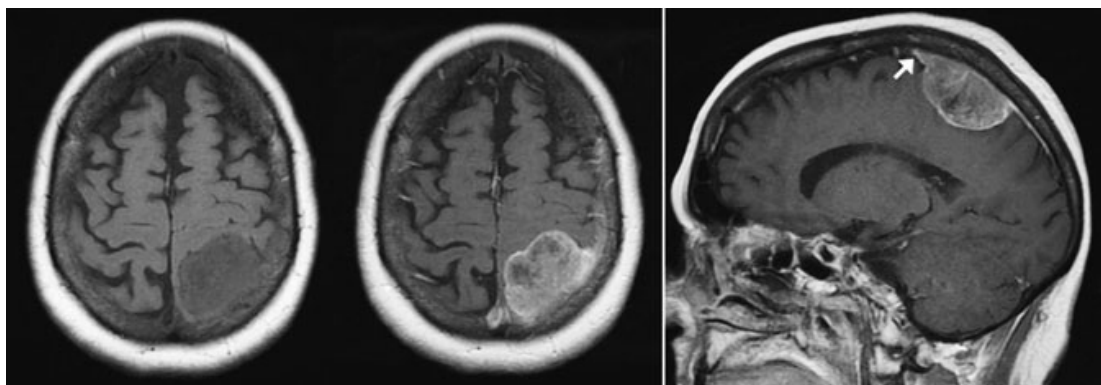


6. How should the patient be treated in the emergency setting?

Case 21

An 52-year-old woman with 6 weeks of progressive right hemiparesis is referred from the medical team. She denies headaches or seizures, and is self-caring and independent, although widowed last year. Her medical history includes type 2 diabetes and hypertension, and she is on aspirin. On examination she walks well with a stick in her left hand. Her right arm is weak and she is unable grasp a pen. She has brisk biceps, supinator, and triceps jerks in her right arm. Her left arm and both legs are normal.

1. Damage to which tract typically leads to an inability to grasp a pen or hold small objects?
2. What is the abnormality on her brain MRI?

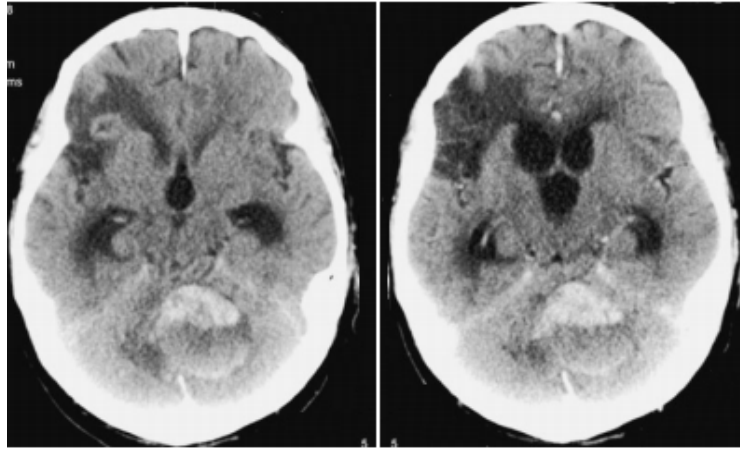


3. Where is this lesion in relation to the motor cortex?
4. Does the location suggest any further clinical signs?
5. Which treatment option would you prefer for this patient?
6. Are there any preoperative considerations?
7. The patient undergoes surgery and recovers with a slow improvement in her hemiparesis. The histology shows an atypical meningioma, WHO grade 2. What implications does this have?

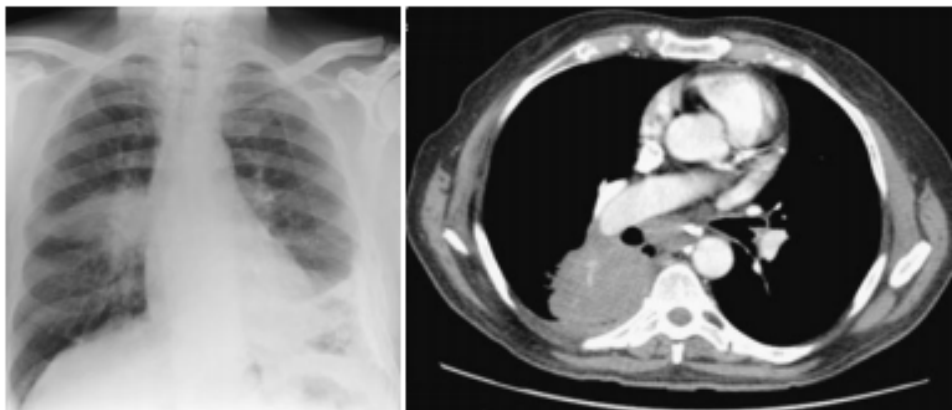
Case 22

A 41-year-old man presents to his GP with a worsening headache which started suddenly 6 days previously. He became very dizzy and his wife noticed him staggering as if drunk. His balance has been poor since then and he has spent the week in bed. His past medical history includes a stroke 6 years previously from which he recovered fully. He is a previous heavy smoker. He takes aspirin and a statin.

1. What is the differential diagnosis?
2. The patient's GP arranges a CT scan and he is then referred to neurosurgery
 - (a) What are the findings on the scans (left, pre-contrast; right, post-contrast)?
 - (b) What is the likely diagnosis?



3. How should this patient be managed acutely?
4. He undergoes a chest X-ray and a CT of his body. What is seen?

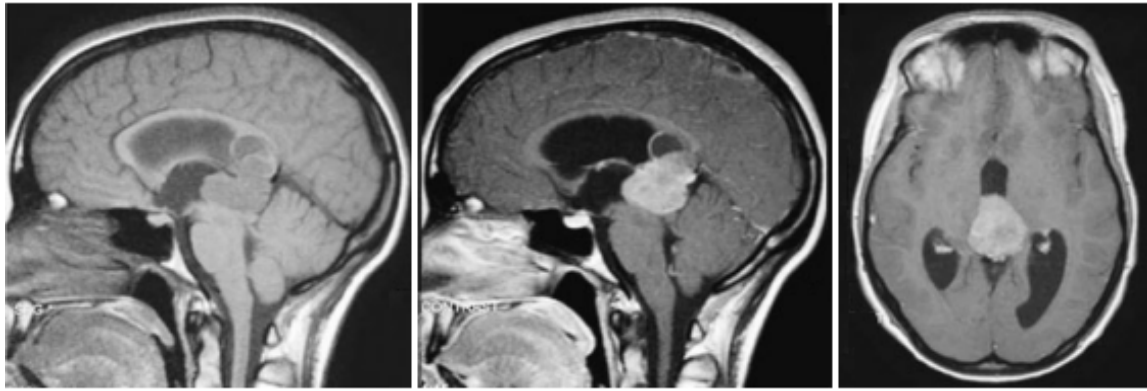


How would you manage him now?

Case 23

A 27-year-old hairdresser presents to her local emergency department with progressive headaches, vomiting, and blurred vision over 2 weeks. She has been waking up at night and vomiting profusely in the mornings. Over the past 24 hours her headaches have been unremitting. She is alert and orientated. Her power is preserved throughout and coordination is normal. She is not photophobic or meningitic. She has reduced visual acuity at 6/12 in the right eye and 6/18 in the left. On fundoscopy there is gross papilledema.

1. This patient has features of increasing intracranial pressure without focal neurological signs or symptoms. What is the differential diagnosis?
2. Describe the abnormalities on the MRI scan.

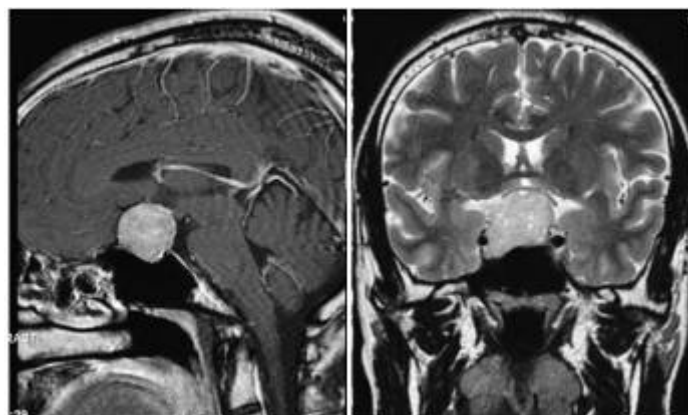


3. What is the differential diagnosis?
4. How should she be managed?
5. What is the value of tumor markers in pineal region tumors?
6. How may surgery be performed? Which anatomical structures are at risk? Should she have any special preoperative investigations?

Case 24

A 31-year-old man presents via the ophthalmology department. He has a 2-week history of progressive difficulty with his vision, particularly bumping into things that he did not realize were next to him and having to turn excessively to either side to see people that are talking to him. The events culminated in a car accident when he drove into a parked car on his side of the road.

1. What type of visual problem does he have?
2. What is the likely diagnosis from the history? Should any other questions be asked?
3. The MRI is performed. What does it show? What is the differential diagnosis?



4. What are the next steps in the management of this patient?
5. The prolactin level in this patient is moderately elevated (93ng/mL, normal range <20ng/mL). What is the relevance of this result, and how would it affect management?
6. How would you manage the patient's hormonal replacement in the

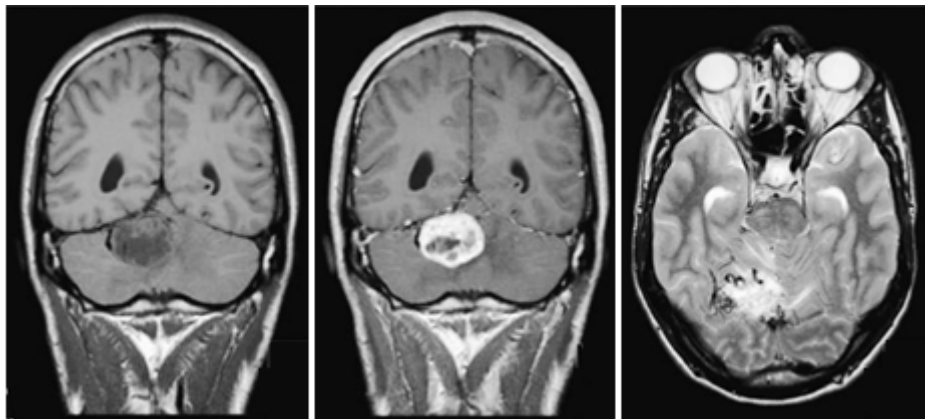
perioperative period?

7. He undergoes trans-sphenoidal surgery which is uneventful. Immediately afterwards he feels that his vision has improved. However, the evening after surgery his urine output increases to a steady 300 mL/hour. What is the differential diagnosis for the increased urine output and the management of each possibility?

Case 25

A 32-year-old man consults his GP with a history of headaches for 3 months, with no vomiting or nausea. They are intermittent and not associated with diurnal variation. His friends have asked him to speak more clearly on occasion, and he has felt that his speech is less fluent. However, he has no imbalance and he has been playing tennis.

1. What is the differential diagnosis?
2. Coronal T1, pre- and post-contrast, and axial T2 MRI sequences are done. What do they show?



3. What treatment would you offer and what is the critical management concern?

Case 26

A 33-year-old man who had a ventriculoperitoneal shunt in infancy for congenital hydrocephalus, which has never been revised, presented to the emergency department with increasing headaches, nausea, and vomiting.

1. What are the important questions to ask in the history?
2. How can a potentially blocked shunt be assessed at the bedside?
3. List two radiological investigations that can be performed to check for a blocked shunt.
4. How should this patient be investigated?
5. The results of the investigations for this patient are as follows:
 - fundoscopy—patient did not tolerate procedure

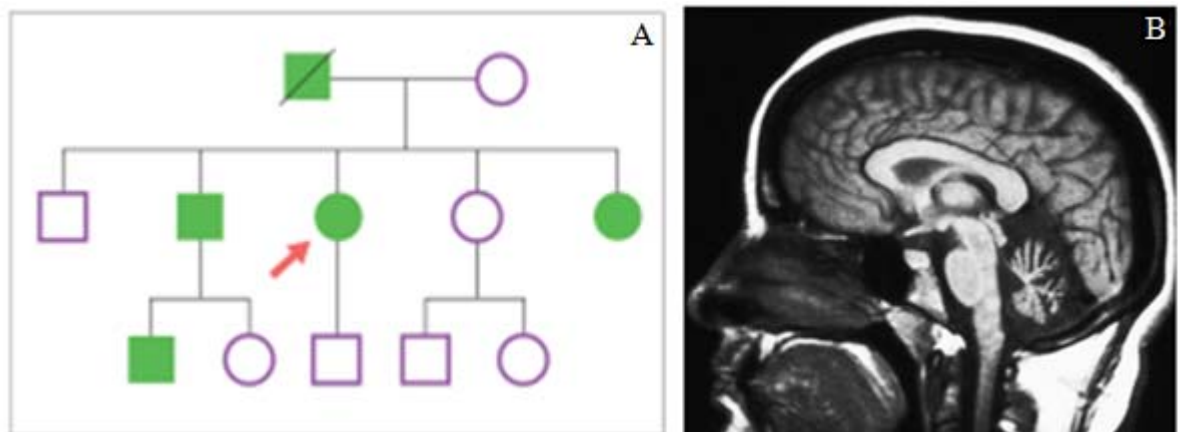
- shunt valve—does not refill on compression
- shunt series X-rays—no abnormalities found
- serum inflammatory markers—normal
- CT brain is shown below.



What will you do next?

Case 27

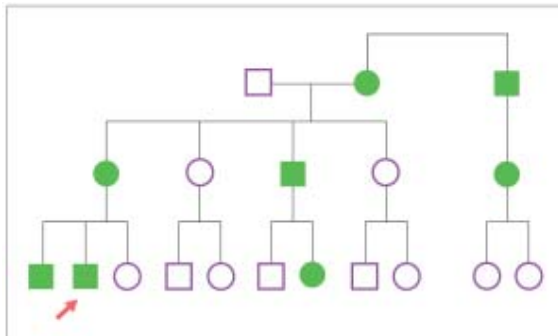
A 40-year-old female with 10 years of progressive ataxia has the pedigree (A) and brain MRI (B) shown.



1. What is the inheritance pattern shown in the pedigree?
2. What does the MRI show?
3. What are the diagnostic possibilities?
4. Is DNA testing available for the differential diagnosis?

Case 28

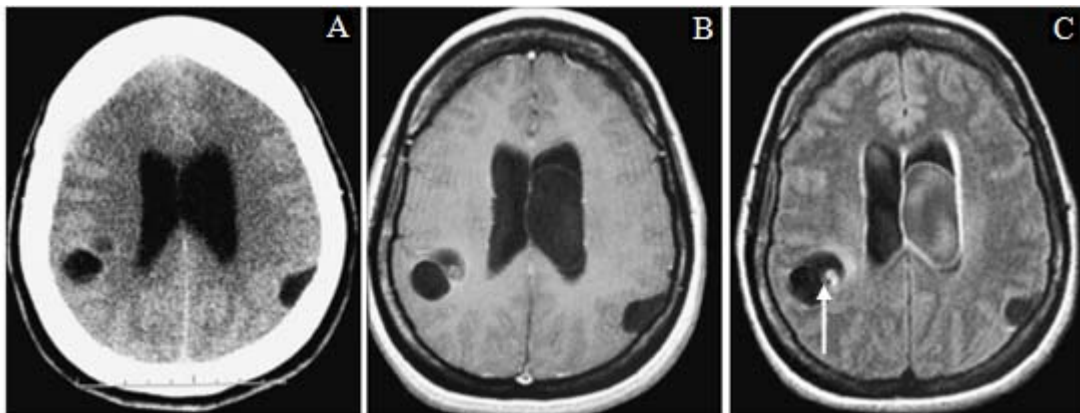
An 18-year-old male has bilateral foot drop, distal muscle atrophy, absent tendon reflexes and moderately slow nerve conduction velocity (NCV). His diagnosis is Charcot–Marie–Tooth (CMT) disease. His pedigree is shown.



1. What type of inheritance does the pedigree show?
2. What type of CMT could he have?
3. The results of his DNA testing show a mutation in the *connexin 32* gene. What is the diagnosis and what are the risks for CMT to his children if he has a daughter and a son?

Case 29

A 39-year-old Honduran female with a long history of mild, chronic headaches presents with a generalized tonic-clonic seizure. Neurologic examination was normal. Nonenhanced CT (a), T1-post-gadolinium MRI (b) and FLAIR MRI (c) were obtained.



1. What are the findings?
2. What is the likely diagnosis?

Case 30

You are called to the bedside of a patient who has just undergone trans-sphenoidal surgery for resection of a pituitary adenoma. The nurse is concerned because the patient has put out 500 ml of urine in the past 2 hours.

1. What other information do you need to plan the most appropriate course of action for managing this patient?

CASE HISTORIES IN BRAIN TUMORS

Answers

Case 1

1. Diffuse infiltration and expansion of the brainstem characteristic of an intrinsic tumor. The basilar artery is enveloped by the mass.
2. Brainstem glioma.
3. This disease carries a poor prognosis in children with an almost 100% mortality by 2 years. In adults it runs a more indolent course, and in a recent series the median survival was 54 months. Radiotherapy is the only treatment which may be helpful. The radiologic appearances are so characteristic that diagnostic biopsy is usually not warranted.

Case 2

1. A short history of dementia and headaches in a middle-aged patient.
2. Butterfly malignant glioma. The diagnosis is obvious on the contrast CT. Stereotactic biopsy would remove all doubt. Resective surgery is not indicated.
3. Radiotherapy may prolong survival, but the prognosis and future quality of life are poor as the tumor spreads around the splenium of the corpus callosum.

Case 3

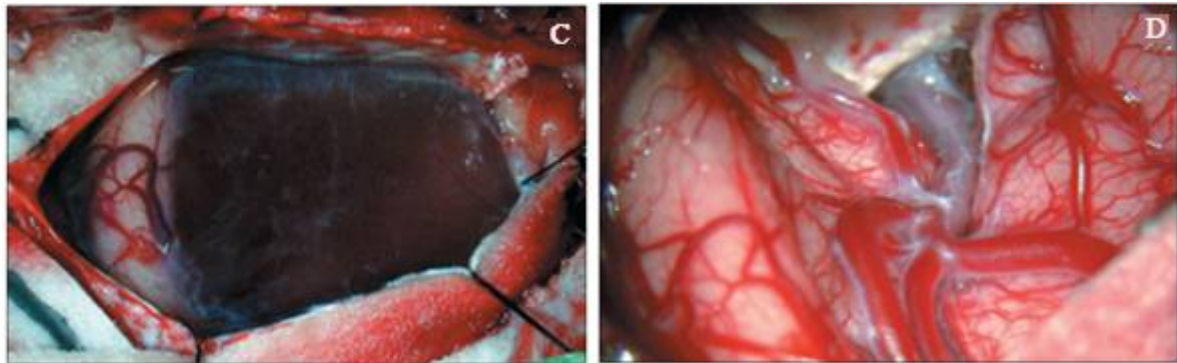
1. The images (A, B) demonstrate an arachnoid cyst. These are congenital lesions that arise during development from splitting of the arachnoid membrane.
2. DWI sequences would help distinguish an arachnoid cyst from an epidermoid cyst, another developmental lesion that may arise when retained ectodermal implants are trapped by two fusing ectodermal surfaces.
3. CSF-like fluid surrounded by an arachnoidal membranous wall.
4. Sylvian fissure and middle fossa.
5. 2:1
6. Most arachnoid cysts that become symptomatic do so in early childhood. Although the clinical presentation varies with location of the cyst, most children present with symptoms and signs of increased ICP (e.g. headache, poor feeding, lethargy) or seizures. Most neurosurgeons recommend not treating arachnoid cysts that do not cause mass effect or symptoms. Accepted indications for operative intervention include increased symptoms (including seizure frequency, ICP, or declining neuropsychiatric testing) and increasing mass effect of the

lesion. Of note, some authors have advocated earlier operative intervention in these children because of the increased risk of subdural hematomas after even mild head trauma.

7. There are three surgical options to treat arachnoid cysts.

(1) Drainage of the cyst by needle aspiration is simple and quick, but there is a high rate of cyst recurrence.

(2) Craniotomy with excision of the cyst wall and fenestration into the basilar cisterns allows direct visualization to treat effectively any loculations and avoids a permanent shunt in some cases (pre- and post-fenestration shown in C, D).



However, a craniotomy has increased morbidity, and subsequent scarring may block fenestration allowing reaccumulation of the cyst.

(3) Shunting of the cyst into the peritoneum is definitive treatment with low morbidity and rate of recurrence. The patient becomes shunt dependent, however.

Case 4

1. The MRI shows a lesion in the right frontal white matter that extends to the cortex anteriorly and the genu of the corpus callosum posteriorly. The cortical sulci are expanded and effaced. The right lateral ventricle is compressed. The signal intensity is the same as, or slightly lower than that of gray matter, and the lesion does not enhance with gadolinium.

2. These findings, along with the clinical presentation of a first seizure at age 38 years, suggest a low grade primary glial neoplasm such as astrocytoma, oligodendroglioma, or mixed tumor. While MRI cannot predict accurately the histologic grade of tumor, higher grade neoplasms such as anaplastic astrocytoma or glioblastoma typically demonstrate neovascularity and consequent enhancement. Glioblastoma and metastatic neoplasms often show central necrosis as well, which appears as a low signal intensity lesion surrounded by a high signal, irregular ring.

Case 5

1. MRI scan (A) shows a giant cell astrocytoma, an enhancing lesion almost always located at the foramen of Monro. They occur in 10–25% of patients with tuberous sclerosis (TSC). Imaging studies of patients with TSC also commonly show subependymal nodules, or tubers (B), low density parenchymal lesions representing heterotopic tissue or defective myelination, and hydrocephalus.
2. Spectroscopy patterns can be helpful to confirm the diagnosis of giant cell astrocytomas (C). Low grade astrocytomas typically have a low creatine to Ch ratio and a low NAA to Ch ratio, with an undetectable lactate peak. More malignant tumors tend to have even lower creatine to Ch ratios and NAA to Ch ratios, and they develop clear lactate peaks.
3. The clinical triad of seizures, mental retardation, and sebaceous adenomas are the hallmark for TSC. A neurocutaneous disorder characterized by hamartomas of many organs including the skin, brain, eyes, and kidneys, TSC has an autosomal dominant inheritance pattern, with the responsible gene located on chromosome 9. Additional associated findings include ash leaf macules and depigmented iris.
4. This symptomatic lesion should be surgically resected. If asymptomatic, paraventricular lesions should be followed closely.

Case 6

1. Parinaud's syndrome is a convergence, accommodation, and an upward gaze palsy often with lid retraction. Patients may also have fixed pupils (dissociated light–near response) and nystagmus retractorius. It is common in patients with masses causing direct pressure on the quadrigeminal plate (e.g. pineal tumors) and in patients with hydrocephalus causing compression of the mesencephalic tectum by a dilated suprapineal recess.
2. The MRI demonstrates a heterogeneously enhancing mass in the pineal region causing lateral and third ventricular enlargement.
3. Statistically, the most likely tumor in this patient is a germinoma. However, many different tumors can be seen in the pineal region. They include other germ cell tumors (e.g. choriocarcinoma, endodermal sinus tumor or yolk sac tumor, embryonal carcinoma, and teratoma), pineal gland tumors (e.g. pineocytoma, pineoblastoma), glial tumors (e.g. astrocytoma, glioblastoma, ependymoma, and

oligodendroglioma), or other miscellaneous tumors including meningioma and metastasis.

4. Germ cell tumors characteristically (but not always) give rise to tumor markers beta-HCG and AFP in the CSF. Elevated CSF beta-HCG is classically associated with choriocarcinomas, but also occurs in a minority of germinomas. AFP is usually elevated in endodermal sinus tumors and embryonal carcinomas. When positive, these markers can be followed and used to assess treatment and recurrence.

Markers associated with tumor types

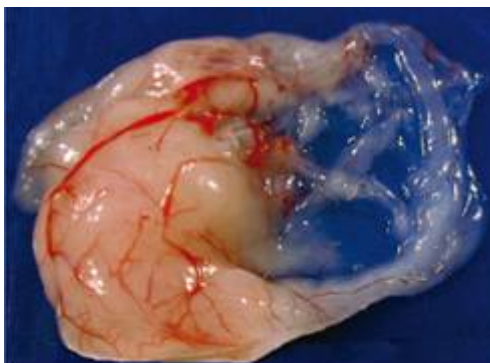
	AFP	β -HCG	PLAP
Germinoma	-	-	+
Teratoma	+	-	-
Endodermal sinus (yolk sac)	+	-	+/-
Embryonal carcinoma	-	-	+
Choriocarcinoma	-	+	+/-
AFP: alfa fetoprotein; β -HCG: beta human chorionic gonadotropin; PLAP: placental alkaline phosphatase			

Because many pineal region tumors have mixed cell types, these markers alone are usually not sufficient for definitive diagnosis (see table above).

Case 7

1. Laughing episodes with secondary generalization are known as gelastic seizures (partial complex seizures). Patients with gelastic seizures commonly have intellectual impairment and psychiatric disturbances.

2.



A hypothalamic (tuber cinereum) hamartoma. They are typically hypodense on T1-weighted MRI, hyperdense on T2-weighted MRI, and do not enhance. They lie between the infundibular stalk anteriorly and the mamillary bodies posteriorly. On cross pathology, resected hypothalamic hamartoma resembles normal cerebral gray matter.

3. Surgical options are considered only if the seizures cannot be adequately controlled with anti-epileptic medications. If possible, surgical resection is preferred. If not, gamma knife radiosurgery can also be used to treat these lesions.

Case 8

1. The patient gives a typical description of cough headache. Cough headache is characterized by severe head pain precipitated within seconds by coughing, sneezing, lifting, straining or any other valsalva maneuver. The headache is usually bilateral and of short duration, typically from seconds up to 30 minutes.

2. The MRI scan shows a type I Arnold–Chiari malformation accompanied by a syrinx. Symptomatic cough headache is usually caused by lesions of the posterior fossa and include basilar impression (e.g. Paget’s disease of the skull), space occupying lesion (e.g. meningioma), midbrain cysts and Arnold–Chiari malformation. Individuals with Arnold–Chiari type I malformations may present with headache as the only symptom. However clinical examination should look for evidence of raised ICP, cerebellar syndrome, syringomyelia or a combination of disorders of the cranial nerves, medulla, cerebellum and spinal cord. Although cough headache may initially be an isolated feature of the disorder, most patients ultimately develop further symptoms and abnormal neurologic signs (usually within 5 years).

3. A diagnosis of benign cough headache is made on the characteristic clinical history described, in the absence of intracranial disease. The syndrome is more commonly seen in men in the age range 40–80 years. The symptoms ultimately improve or resolve spontaneously in the majority of individuals. Relief may be obtained with indomethacin and some patients have responded to therapeutic lumbar puncture. Patients with symptomatic cough headache tend not to respond to indomethacin.

Case 9

1. The biopsy (B) shows the typical appearances of an oligodendroglioma demonstrating the characteristics ‘fried egg’ appearance. The cells have sharp borders without processes, clear cytoplasm and round uniform central nuclei. In addition there is a fleck of calcium, shown clearly on the CT scan (A, arrow), which is another characteristic of these tumors.

2. It is now accepted that these tumors are particularly chemosensitive and have shown 75% responses rates to a combination of procarbazine, CCNU and PCV.
3. Young age, good performance status, extent of surgical resection, loss of heterozygosity on chromosomes 1p/19q. Oligodendrogliomas are relatively uncommon tumors accounting for about 5–10% of primary brain tumors. They usually occur in young and middle-aged adults and present with seizures, focal neurologic deficits, raised ICP, or combinations of these symptoms and signs. They are occasionally multifocal. Intratumoral calcification is common. They may be low grade or high grade (anaplastic) and are frequently amenable to surgical resection because they occur in non-dominant frontal and temporal lobes. They have a better prognosis than ordinary gliomas because they are chemosensitive. Recently, molecular genetic analysis has shown that patients with anaplastic oligodendrogliomas who have deletions of chromosome 1p/19q have a median survival time for low grade oligodendrogliomas of >10 years.

Case 10

1. An axial T2-weighted MRI (A) shows a left frontal mass with surrounding high signal, vasogenic edema. MR spectroscopy obtained from the tumor (B) and the contralateral normal white matter (C) show that Ch and lactate are increased within the mass, and that NAA is decreased. MR spectroscopy is a technique for analyzing the chemical composition of a specified brain region. It measures amounts of several metabolites including NAA, myo-inositol, Ch, creatine, lactate, as well as lipids and other amino acids.
2. In normal brain, NAA, a marker for normal neurons, is elevated relative to Ch, a marker for cell membrane turnover. In patients with high grade tumors, Ch is increased and NAA is decreased. In this case, considerable lactate is present as well, indicating some degree of necrosis. While these MR spectroscopy findings could indicate either a high grade neoplasm or cerebral infarct, the MR imaging findings suggest a tumor. The patient underwent craniotomy with intra-operative motor and speech mapping, with removal of a glioblastoma.

Case 11

1. The MRI is a T2-weighted MRI (CSF is white, gray matter is lighter than white matter). This is usually better than T1 at showing pathology. In this scan there is a tumor in the temporal lobe with no associated mass effect.

2. In view of the 20-year history of seizures beginning in childhood, and the MRI appearances, the tumor is almost certainly benign. In the past, these tumors were often categorized as 'low grade' gliomas, but recent histopathologic evidence has demonstrated that these tumors contain ganglioneuronal elements and are termed dysembryoplastic neuroepithelial tumors (DNTs). These tumors commonly occur in the temporal lobes and result in temporal lobe epilepsy. They are often cystic on MRI or CT scan, and may be associated with cortical dysplasia.

3. Surgical removal of a DNT has a high chance of resulting in remission of the epilepsy, even in cases where there is associated cortical dysplasia.

Case 12

1. Primary CNS lymphoma.

2. Steroids (dexamethasone). The scan and response to steroids is typical of PCNSL. This tumor is rare but steadily increasing in incidence, even without the AIDS epidemic. It is essentially a local disease with <10% of patients having evidence of systemic disease at autopsy. Many lesions are periventricular, which predisposes to dissemination through the CSF. There is usually intense and homogeneous contrast enhancement on CT scanning. PCNSL is exquisitely sensitive to the lympholytic effects of steroids and so they should not be started before diagnostic biopsy. The treatment of this condition is based around high dose MTX chemotherapy followed by radiotherapy. Unlike systemic lymphoma, the conventional CHOP regimen provides only short-lived benefit.

3. Absent Cr signal in CNS lymphoma.

MRSI

(PRESS: TR/TE = 2000/144 ms; 32×32 matrix; FOV = $240 \times 240 \times 15 \text{ mm}^3$) was acquired at the level of the mass in the right parietal lobe. Cho, Cr, NAA, and lipids maps with two selected spectra are illustrated. The spectrum from the mass (1) shows very high elevation of Cho and mobile lipids signal, associated with absent Cr and NAA signals. The metabolite maps nicely illustrate the abnormal changes.

4. European standard for the treatment of PCNSL:

- When the presence of PCNSL is suspected, no steroids are given before tissue diagnosis is established.

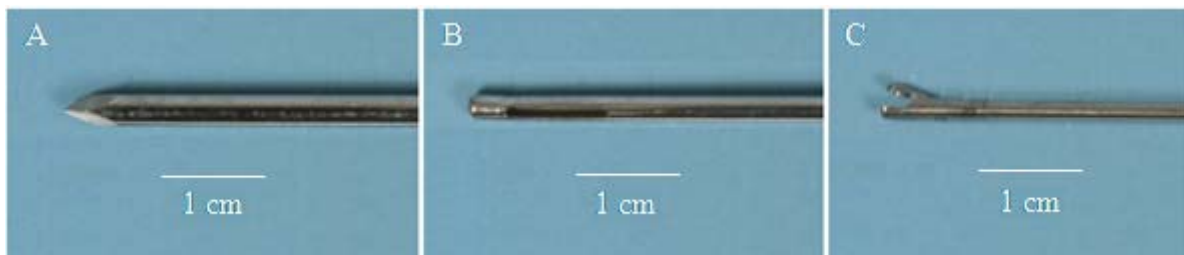
- Tissue diagnosis is established by stereotactic biopsy.
- Risk-adapted chemotherapy protocols of variable intensity are the current gold standard of treatment.
- Surgical resection for lymphoma is unusual and is the consequence of highly individual circumstances.

5. **Technique of Frame–Based Stereotactic Biopsy**

Nowadays, CT- and MRI-compatible stereotactic frames are available.

After calculation of the target point, the values of the coordinates are transferred to the stereotactic apparatus. After a 0.5 cm skin incision need use a twist drill trepanation of the skull with perforation of the dura (A). Afterwards, the biopsy probe is advanced to the target point and two to three biopsies are taken.

Another biopsy is taken along the trajectory before and behind the target point, depending on the size of the lesion. Subsequently, histological results from the cross-section of the lesion are obtained. In most cases usually used a side-cutting aspirator called a Sedan needle for biopsy sampling (B). This instrument provides a 1 cm biopsy cylinder. In brain stem lesions a 2 mm forceps is used (C).



Case 13

1. This is a brain tumor, most probably astrocytoma.
2. Spectroscopy patterns can be helpful to confirm the diagnosis of astrocytoma. Note that Cho significantly increased and NAA decreased in the tumor (2) compared with the contralateral normal spectrum (1). Note also appearance of a small lactate peak in the spectrum from the tumor, suggesting increased glycolytic tumor metabolism.
3. The neoplasm was surgically removed and a grade III anaplastic mixed oligoastrocytoma (MOA) was diagnosed.

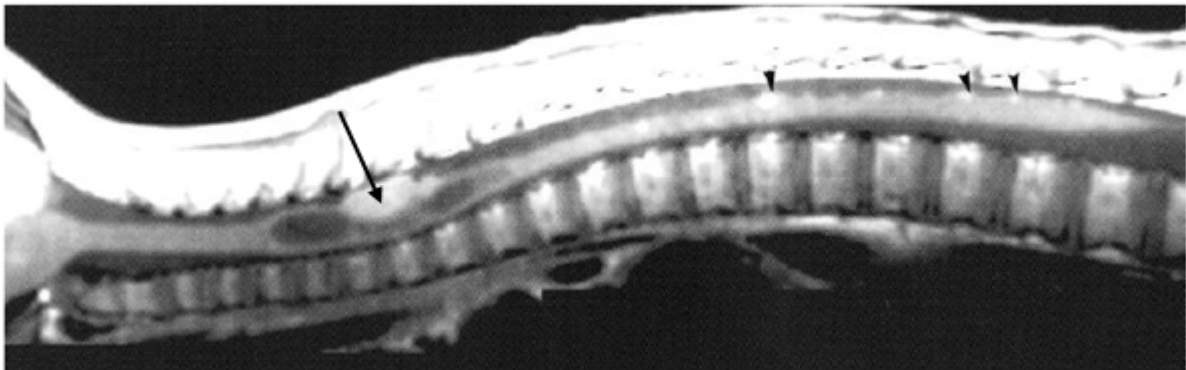
Case 14

1. These are Lisch nodules, hamartomas within the iris. They are a pathognomonic feature of NF1. Other cutaneous features of this condition are cutaneous neurofibromas, plexiform neurofibromas, café-au-lait spots, and axillary freckling. Hypertension can result from renal artery stenosis or pheochromocytoma.
2. NF1 is inherited as an autosomal dominant condition with abnormalities in the neurofibromin gene on chromosome 17.
3. The incidence of CNS tumors especially optic nerve glioma is increased in NF1. In addition, there is an increase in the incidence of cortical dysplasia in NF1.

Case 15

1. MRI shows enhancing mass in the cerebellar vermis extending into the fourth ventricle.

Contrast T1-MRI: spinal canal shows large mass (*arrow*) in junction of cervical and thoracic spine with syrinx; multiple small enhancing nodules (*arrowheads*) over spinal cord surface:



2. Medulloblastoma dorsal to the fourth ventricle originating in the vermis with drop metastases.
3. Pilocytic astrocytoma, medulloblastoma, brainstem glioma, and ependymoma.
4. Residual tumor volume after neurosurgical resection.

Comment

This is the classic age, location, and imaging appearance for the medulloblastoma.

Medulloblastoma is, depending on the age and gender of the patient, one of the most frequent primary neoplasms of the posterior fossa. Medulloblastoma is

especially frequent in boys in their first decade of life. Overall, medulloblastoma (25%) is second to pilocytic astrocytoma (35%). Brainstem gliomas (25%) and ependymomas (12%) are the third and fourth most frequent tumors of the posterior fossa in children. In total, these four variants represent 97% of all posterior fossa tumors.

Medulloblastomas most frequently arise dorsal to the fourth ventricle either in the midline (vermis, 75% to 90%) or in a somewhat more lateral position (10% to 15%; also known as lateral medulloblastoma). Consequently, the fourth ventricle is pushed anteriorly and serves as an anterior tumor border. The compression of the fourth ventricle may result in an obstructive hydrocephalus. Clinical symptoms are either related to the obstructive hydrocephalus or to local tumor infiltration and may include ataxia, gait disturbance, nausea, vomiting, and headaches. Medulloblastomas have a high cellularity and are consequently dense on computed tomography.

On magnetic resonance imaging the lesions can be differentiated from ependymomas, which are typically located within the fourth ventricle (in contrast to the medulloblastomas, which are primarily located dorsal to the fourth ventricle). Medulloblastomas usually display an ill-defined dorsal border because of infiltration of the adjacent vermis or cerebellar hemispheres; they are T1 hypointense to isointense and T2 isointense or hyperintense. Contrast enhancement may be strong but is occasionally absent.

Cerebrospinal fluid (CSF) metastases may occur when the tumor has invaded the fourth ventricle. Tumor metastases may be seen within the third ventricle and lateral ventricles or along the spinal cord. A preoperative work-up should include the entire spinal axis. Prognosis depends of the residual tumor bulk after neurosurgical resection. The smaller the residual component, the better the prognosis. The residual tumor component is even of higher prognostic significance than the initial tumor size. In addition, the histology, immunohistochemistry, and neurogenetic results will also help to determine prognosis. Adjuvant treatment is decided by the combined information collected by imaging, immunohistochemistry, neurogenetic analysis, and residual tumor bulk after neurosurgery. Prognosis significantly improved in the last decade, with good long-term prognosis in most cases.

Case 16

1. The scan shows a ring-enhanced mass lesion in her right frontoparietal lobe with surrounding edema. The radiologic differential diagnosis is wide but in the context of an immunosuppressed patient the most likely differentials include fungal abscess, cerebral toxoplasmosis, nocardia and bacterial abscess. Intracerebral metastases from non-Hodgkin's lymphoma are extremely rare.

There are many potential causes of a cerebral ring enhancing lesion and remembering them all can prove tricky. Well, that is unless you know the very handy and aptly named **MAGIC DR** mnemonic:

M - Metastasis

A - Abscess

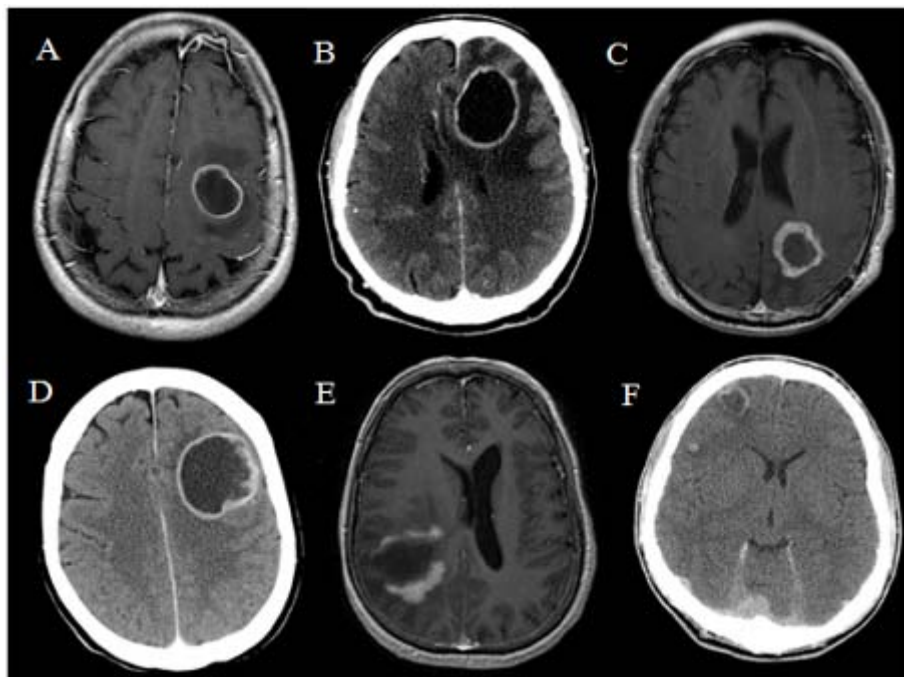
G - Glioblastoma multiforme (GBM)

I - Infarct (subacute phase)

C - Contusion

D - Demyelinating disease (e.g., tumefactive MS)

R - Radiation necrosis



Although you can't possibly know by looking at the single images, for what it is worth, the above cases are.

A = Metastasis, **B** = Abscess, **C** = Radiation necrosis,
D = GBM, **E** = Demyelination, **F** = Contusion.

2. Broad spectrum anti-bacterial, anti-fungal and anti-protozoal cover. Do a biopsy if no improvement is observed within 1 week to guide further treatment. Perform aspiration and surgical decompression if there are symptoms and signs of rising ICP. CNS infections are an uncommon complication of cancer and are usually seen in patients with lymphoma and leukemia, often after chemotherapy and bone marrow transplantation. The usual florid symptoms apparent in immunocompetent patients with CNS infection are not seen in these patients. In particular, headache and fever may be absent (as in this patient). Brain abscesses are usually due to fungi, nocardia or toxoplasma, i.e. a different group of

pathogens compared to immunocompetent patients, and need to be covered with appropriate antibiotics. Surgical intervention should be reserved when the conscious level declines or when there is no improvement after 1 week of appropriate treatment. Often other foci of infection, e.g. lung abscesses due to nocardia, allow precise diagnosis without the need for a brain biopsy.

Case 17

1. The dysphasia and a right hemiparesis suggest a left cerebral lesion. Unequal pupils may indicate impending transtentorial herniation from mass effect. The speech difficulties two weeks earlier suggest a rapidly expanding mass lesion such as a malignant tumor or a subdural hematoma. The reasons for collapse are unclear from the limited history but he may have suffered a seizure.

2. There is a mass in the left hemisphere surrounded by an extensive area of low density which represents edema. There is midline shift and compartmental hydrocephalus demonstrated by the enlarged lateral ventricles on the right due to compression of the ventricular system at the foramen of Monro. There is herniation of the uncus of the left temporal lobe.

The mass exhibits ring enhancement. The differential diagnosis is between a high grade glioma, an abscess, and metastasis. In the absence of raised infective markers, a tumor is more likely.

3. Three main types of edema occur in the brain.

Cytotoxic (intracellular) edema occurs mainly in traumatic and ischemic brain injury. It results from defective sodium ATP-driven transmembrane channels in the affected cells, leading to sodium (and thence water) retention. It is not responsive to corticosteroids.

Interstitial edema occurs in hydrocephalus and is due to high CSF pressures in the ventricular system, resulting in CSF egress into the adjacent brain parenchyma.

Vasogenic edema is due to increased capillary permeability from breakdown of the blood–brain barrier. It is seen principally with tumors and abscesses. It is responsive to corticosteroid therapy. Different types of edema may coexist. In this patient it is predominantly vasogenic.

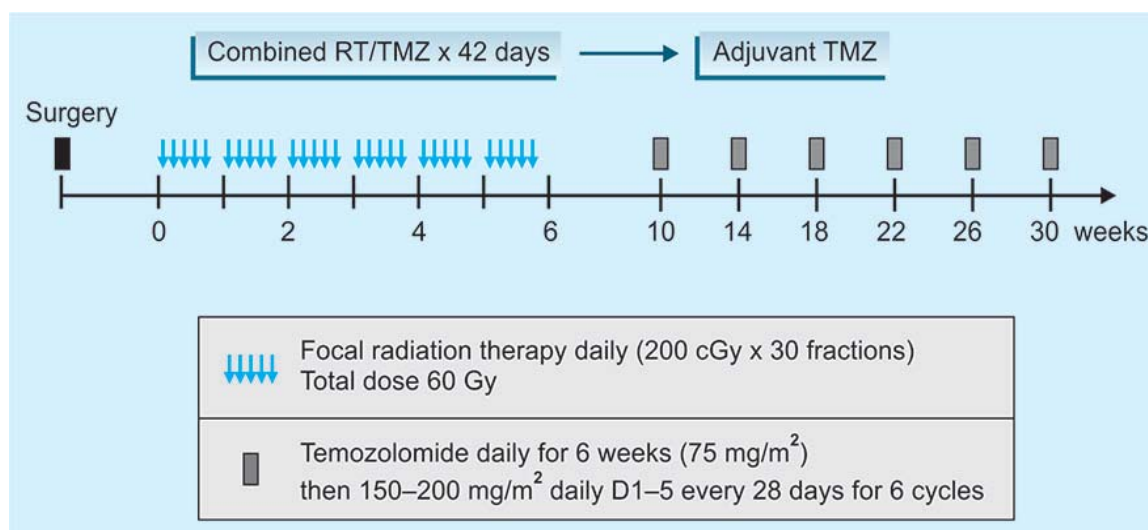
4. The patient is at risk of rapid deterioration due to raised intracranial pressure. He requires corticosteroids (e.g. dexamethasone 8mg bd) to reduce vasogenic tumor edema and decompressive surgery if he is to survive. He underwent a craniotomy and debulking of the tumor, and the histology showed a

glioblastoma (WHO grade 4). Treatment for glioblastoma after surgical resection consists of radiotherapy and temozolomide for patients in good performance status. The prognosis, even with treatment, is very poor and stands at approximately a year.

5. Chemotherapeutic agents have been used for many years in the management of malignant brain tumors, but clinical benefits remain uncertain.

Standard therapy for newly diagnosed glioblastoma commonly referred to as the “Stupp regimen”*.

Nitrosoureas are still the standard drugs used in the initial chemotherapy regimen such as carmustine (BCNU) as a single agent (200 mg/m^2 IV q8 weeks) or in PCV combination, which comprise procarbazine (60 mg/m^2 days 8-21), CCNU (110 mg/m^2 PO day 1), and vincristine (1.4 mg/m^2 days 8-29) q6 weeks.



Temozolomide ($150\text{--}200 \text{ mg/m}^2$ days 1–5 q4 weeks) is an oral alkylating drug that has activity against recurrent malignant glioma. Currently, various clinical trials are under way to study the effectiveness of a variety of molecular-targeted therapies either used alone or in combination with cytotoxic drugs like Temozolomide.

Problems of drug administration.

Toxicity: The ideal cytotoxic drug selectively kills tumor cells; but tumor cell response related directly to the dose. High drug dosage causes bone marrow suppression. Often marrow depression occurs before an adequate therapeutic dose is reached.

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Case 18

1. To cause a homonymous visual field deficit the lesion must be posterior to the optic chiasm. Therefore it may be in the right optic tract, thalamus, optic radiation, visual cortex, or adjacent structures causing impingement.
2. A sudden-onset severe headache probably represents an intracranial bleed. Given the presumed location of the lesion, this is probably an intraparenchymal hemorrhage. The causes include vascular lesions such as cavernomas, arteriovenous malformations, and other lesions that can bleed including malignant tumors. Alternatively, if there is no mass lesion amyloid angiopathy, hypertension and small-vessel disease could be precipitating factors.
3. The T2 weighted MRI (left) shows a lesion of mixed signal intensity adjacent to the primary visual cortex of the right occipital lobe. There is surrounding edema, represented by high signal. There is not widespread evidence of white matter disease elsewhere in the brain which, if present, would suggest underlying hypertension or vasculopathy. Therefore appearances are consistent with a tumor. Pre- and postcontrast T1 images (centre and right) show that the abnormality enhances heterogeneously. This makes a malignant tumor likely. Metastasis is the most common malignant brain tumor, whereas glioblastoma is the most common primary brain tumor. Given the patient's previous unremarkable medical history, the latter is the likely diagnosis.
4. Glioblastoma has a poor prognosis — median survival of 9–15 months with treatment. Prognosis is influenced by age at presentation and degree of neurological disability. Younger patients with better performance status survive longer. Treatment factors affecting prognosis include extent of surgical resection (greater resection giving longer survival) and the provision of adjuvant radiotherapy and temozolomide chemotherapy, both of which prolong survival. Certain tumor attributes also influence prognosis; the best studied is MGMT promoter methylation status which, if present, confers additive benefits of temozolomide treatment. This patient is relatively young with a Karnofsky score of 90. Maximal surgical resection followed by radiotherapy and temozolomide is appropriate if glioblastoma is confirmed at surgery. The patient undergoes a macroscopic resection and glioblastoma is confirmed. He tolerated a full course of radiotherapy with concomitant temozolomide, followed by adjuvant temozolomide for 5 days a week every month. He recovered well but developed

headaches a year later. They are progressive and worse in the morning. His visual field improved a little after the surgery but has returned.

Case 19

1. Various factors influence an individual's chance of having a seizure: the presence of a systemic inflammatory process, commonly sepsis; electrolyte imbalances, particularly of sodium; certain drugs, including some antidepressants and tramadol; sleep deprivation; rarely, flashing lights.
2. This is a potentially important focal sign which should recover (Todd's paresis). It may relate to a pre-existing mass lesion. The deficit may persist longer if it is due to an acute lesion, such as hemorrhage, commonly a cavernoma or arteriovenous malformation rather than an aneurysm, as aneurysms present with ictal headache rather than a fit with focal cortical neurological deficit. Alternatively, malignant tumors may present with a hemorrhage.
3. The posterior right frontal lobe low attenuation is probably a low-grade tumor. Absent contrast enhancement will usually distinguish it from a high-grade (malignant) tumor. With clear edges, a small cortical infarct is another possibility. Diffusion-weighted MRI can help decide between infarct and tumor.
4. The tumor is in the posterior right frontal lobe, close to the motor cortex. Neither CT nor MRI show evidence of hemorrhage, so the arm weakness is likely to be Todd's palsy. Coronal sequences show no tumor contrast enhancement, so the diagnosis is low-grade glioma (probably astrocytoma).
5. Anticonvulsants will be indicated to prevent further seizures. Low-grade gliomas can be treated conservatively or surgically. Conservative management with radiological surveillance avoids surgical risk in a potentially eloquent area. However, the patient will have no histological diagnosis and hence prognosis, and the uncertainty can be difficult to manage. Surgically, biopsy alone (for histological diagnosis) or resection of the tumor are options. Definitive histology allows for possible further non-surgical treatment (chemo- or radiotherapy), although it is not usually offered for a low-grade tumor. Biopsy may occasionally take a non-representative sample and provide erroneous information. Near the motor cortex most surgeons would use image guidance or have the patient awake during surgery for intraoperative assessment of the involved brain prior to resection. This requires a cooperative relaxed patient and an experienced anaesthetist.

Case 20

1. Motor decline can be non-specific, but the poor balance points to a cerebellar problem. Holding the head, being irritable, and vomiting suggest raised intracranial pressure. These are typical features of a posterior fossa mass lesion compressing the fourth ventricle and causing hydrocephalus. Although any mass lesion can cause this picture, a tumor is the likely diagnosis given the progressive symptoms and age.
2. A cerebellar hemispheric lesion causes loss of coordination ipsilaterally. If the lesion involves the cerebellar vermis (midline), there may be truncal ataxia with a broad-based gait. Hydrocephalus in children can cause downward deviation of the eyes ('sun-setting'), bilateral sixth nerve palsies, and under the age of 18 months bulging of the anterior fontanelle if it is open. Fundoscopy may be difficult to perform in children, but will show papilloedema in most patients with raised intracranial pressure. Visual acuity may be reduced.
3. Efferent outputs from the cerebellum to the limbs either 'double cross' (the dentato-rubro-thalamic tract and the globose-emboliform-rubral tract) or do not cross (the fastigial-vestibular and fastigial-reticular tracts). Hence cerebellar lesions cause ipsilateral symptoms.
4. He requires urgent imaging of the brain. MRI is preferable as it will delineate the pathology, but if there is neurological compromise due to presumed hydrocephalus a CT scan could be performed if more readily available.
5. (a) These are axial T1 post-contrast (left) and axial FLAIR (right) sequences. (b) The T1 image shows a large mass lesion with a solid component and large cystic component arising from the right cerebellar hemisphere and extending cross the midline. The solid component has a strong uniform enhancement pattern with some discrete non-enhancing spots in it as compared with the non-contrast scan (not shown). The fourth ventricle cannot be seen and is effaced by the mass. The FLAIR sequence shows lateral ventricular enlargement, and the whiter areas around the tips of the ventricles are indicative of 'trans-ependymal flow' of CSF indicating high pressure. (c) This is a posterior fossa tumor with obstructive hydrocephalus. An avidly enhancing solid component with a cyst suggests pilocytic astrocytoma. Other childhood tumors are ependymoma and medulloblastoma (primitive neuroectodermal tumor (PNET)). Pilocytic astrocytoma is benign and often curable with surgery. Ependymoma and medulloblastoma are malignant and require adjuvant chemoradiotherapy.

6. Within 24 hours, dexamethasone helps reduce the peritumoural edema, usually allowing CSF flow through the fourth ventricle to alleviate the hydrocephalus. If the headache does not resolve or the GCS worsens, treating the hydrocephalus with an external ventricular drain or endoscopic third ventriculostomy is required. Even with improvement on steroids, most pediatric neurosurgeons would consider a third ventriculostomy a few days after presentation and plan tumor resection thereafter. This often facilitates the definitive surgery. Spinal MRI is also required to exclude CSF metastases which are harder to identify postoperatively because of the presence of CSF blood products. In this case, clinically there is raised intracranial pressure but the hydrocephalus is not severe on imaging. The patient does not require emergency CSF drainage but is commenced on dexamethasone. Radiologically this is pilocytic astrocytoma and excision should be curative. If the tumor were malignant, surgery with maximal resection would be followed by adjuvant chemoradiotherapy.

Case 21

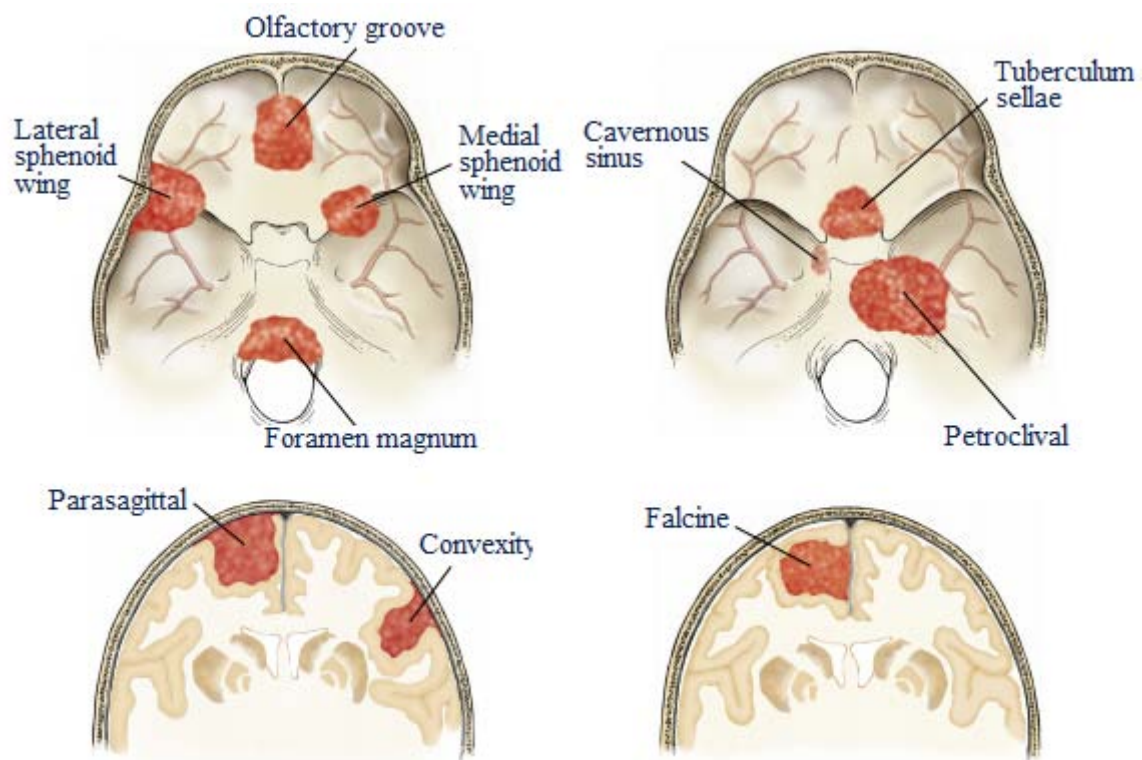
1. Impairments of fine motor movements typically arise from lesions of the pyramidal (corticospinal) tract.
2. An enhancing extra-axial mass overlies the left parietal lobe. It is durally based with a dural “tail” (arrow), suggesting a meningioma.
3. The motor symptoms are due to mass effect on the pre-central gyrus.
4. Whilst initial sensory examination may be normal, cortical sensory loss may be elicited by asking the patient to carry out tasks of sensory discrimination such as naming a number drawn on her palm or asking her to remove objects from a pocket. These tasks require cortical integration of somatosensory stimuli with spatial awareness and decision-making.
5. Management of meningiomas may be expectant (observation with treatment of seizures if required), surgery, or radiosurgery. This patient has a partial deficit and treatment is appropriate. Radiosurgery is valuable to control small (<3cm) tumors that are surgically inaccessible. It will prevent progression of the tumor but will not remove it. Open surgery has a high cure rate, related to the Simpson grade which is highest for meningiomas of the cranial convexity. Therefore, despite her age, surgery was undertaken.

6.



Meningiomas may bleed considerably during surgery. They derive their blood supply principally from the dura but also recruit vessels from the adjacent brain. Neurovascular embolization may be performed preoperatively for large tumors. This is rarely necessary for tumors of the convexity as the dural blood supply will be

encountered early on in the operation and is readily dealt with. Preoperative embolization has a stronger role for skull base tumors or those where the blood supply will be encountered late in the surgery.



Common sites of meningiomas growth in relationship to adjacent skull, brain, and dural reflections.

7. Atypical meningiomas are not malignant but have a higher recurrence rate, even after complete excision. Patients should be followed closely with a view to early treatment of recurrence if necessary. Treatment could involve further surgery or radiotherapy.

Case 22

1. The differential diagnosis for a sudden-onset headache includes any type of intracranial bleed or migraine. Subsequent problems with balance indicate pyramidal motor pathway or cerebellar involvement. A bleed causing hydrocephalus would also lead to unsteadiness.
2. a) There is a well-defined weakly enhancing mass in the left cerebellar hemisphere crossing the midline. It is heterogeneous on the non-contrast CT, but high density suggests recent hemorrhage. The fourth ventricle is occluded, causing hydrocephalus. There is an old area of right frontotemporal encephalomalacia indicating his previous stroke.

(b) The most common cause of a posterior fossa tumor in adults is metastasis. Malignant tumors can present with intra-tumoral hemorrhage. Malignant primary brain tumors (glioblastoma) of the cerebellum are most uncommon.
3. There is hydrocephalus but the patient is alert and does not need immediate surgery. He receives dexamethasone 8mg bd to reduce vasogenic edema around the tumor; this will improve his headaches and may improve the hydrocephalus. His aspirin is stopped in anticipation of surgery. An urgent effort should be made to locate a primary tumor and the extent of the metastatic disease for the following reasons.
 - 1) If prognosis is especially poor, cranial surgery may not be in his best interests.
 - 2) This information will inform discussion with oncologists, the patient, and the family. If a histological diagnosis is needed tissue may be obtained more easily from other sites (e.g. via a CT-guided biopsy) than from a posterior fossa craniotomy.
4. There is a right hilar mass posteriorly situated on the CT, consistent with a primary lung tumor. Metastatic lung cancer has a poor prognosis. If the patient does not undergo surgery, he will probably die of hydrocephalus in the next few days or weeks. Surgery may extend his life by a few weeks but little more. Staging requires an MRI of the brain to look for small metastases, and CT of the chest, abdomen, and pelvis. Tissue should be sampled from wherever is most convenient to inform a multidisciplinary discussion between oncologists, neurosurgeons, thoracic surgeons, and palliative care specialists to decide whether or not to treat the patient aggressively. In practice, when the first presentation of malignancy is with metastatic disease, it is difficult to offer only palliative care without a tissue diagnosis. He underwent a posterior fossa craniotomy which confirmed a secondary tumor from lung adenocarcinoma. He recovered well from the operation and after 6 days was discharged to a hospice before going home. He subsequently only survived a further 4 weeks.

Case 23

1. The differential diagnosis is a large space-occupying lesion in a non-eloquent area such as the right frontal or temporal lobe. The short history makes a malignant tumor more likely than a benign tumor. The second possibility is hydrocephalus. There are many causes of hydrocephalus presenting in adulthood; most will also have symptoms of the causative condition, such as meningitis, subarachnoid hemorrhage, or cerebellar tumor. Presentation with hydrocephalus alone suggests small mass lesions around the third ventricle, cerebral aqueduct, and fourth ventricle. These include colloid cysts of the third ventricle, pineal region tumors, and fourth ventricular tumors including choroid plexus tumors and ependymoma. Aqueduct stenosis commonly presents in the neonatal period, but may present in a delayed manner in older patients.
2. There is a well defined homogenously enhancing mass in the posterior aspect of the third ventricle, with a small cyst superiorly. There is marked associated hydrocephalus. The basal CSF cisterns, including the prepontine cistern, are patent.
3. This is a pineal region tumor; germinomas and teratomas are most common. Other pineal region tumors include pineocytoma or its malignant counterpart pineoblastoma, astrocytomas, and meningiomas. In an older patient one might suspect a metastasis.
4. Steroids should be given, but there is no visible perilesional edema and improvement with steroids may be minimal. The hydrocephalus should be treated with either an EVD or an endoscopic third ventriculostomy (ETV). ETV allows physiological CSF resorption and avoids external hardware with attendant risk of infection. Depending on the angle of approach, it may also allow biopsy of the tumor if positioned posteriorly in the third ventricle after creating the ventriculostomy.
5. Tumor markers in the serum and CSF can aid diagnosis although they do not provide a definitive diagnosis. They are also important in monitoring response to treatment. An ETV produced good resolution of her symptoms. Her tumor marker studies are negative and therefore she is scheduled for surgery.
6. The pineal region is surgically challenging. The most common approach is the supracerebellar infratentorial approach, which was used here. With the patient in a sitting position a craniotomy was performed over the cerebellum to allow it to fall away from the tentorium. Access to the pineal region is then through the corridor above the cerebellum and below the tentorium. The pineal region contains a number of large veins, including the internal cerebral veins that join to form the vein of Galen which feeds into the straight sinus in the tentorium. During the approach the craniotomy will be near the transverse sinus which can bleed torrentially if opened.

Because of the patient's sitting position and the chance of inadvertent opening of dural venous sinuses, there is a risk of intraoperative air embolism. Cerebral venous pressure in the sitting position is lower than atmospheric pressure and hence open veins may suck air in. The anesthetist must be prepared: the treatment involves emergency repositioning of the patient or a central venous line in the right atrium to evacuate air with a syringe. Before surgery, a bubble echo study should be performed to look for an atrial septal defect, the presence of which would increase the chances of a systemic air embolism.

Case 24

1. Bumping into objects in the periphery and needing to turn excessively to see people points to a visual field defect affecting the temporal fields of vision.
2. A bitemporal visual field defect is caused by compression of the central part of the optic chiasm and is the typical finding in patients with large pituitary tumors. Features of pathological hormonal secretion by the tumor should be sought. Constant lethargy, loss of sex drive, and impotence should raise the possibility of pituitary failure. In men, galactorrhea, loss of body hair, and impotence may suggest excessive prolactin secretion from a prolactinoma. Most pituitary tumors large enough to present with visual failure will be non-functioning.
3. The most likely diagnosis is a large pituitary tumor. The differential diagnosis (of other tumors in this location) includes craniopharyngioma and meningioma.
4. This patient has visual failure due to the tumor and requires treatment before vision deteriorates. Further assessment is required.
 1. A formal visual field examination (Goldmann fields) to document the extent of his visual failure.
 2. A full pituitary hormone screen. Pituitary function should be assessed by an early-morning cortisol, follicle-stimulating hormone, luteinizing hormone, thyroid-stimulating hormone, growth hormone, and prolactin levels. The three hormones commonly secreted by pituitary tumors are cortisol, growth hormone, and prolactin. Cortisol and growth hormone cause Cushing's disease and acromegaly, respectively. An urgent prolactin level is vital in this patient to exclude a prolactinoma as the initial treatment is then medical (with cabergoline or bromocriptine).
5. This is probably due to compression of the pituitary stalk by the tumor, resulting in loss of hypothalamic dopamine-mediated inhibition of prolactin release from the pituitary gland. The result is a prolactin level elevated to that of a few times normal. This level is also seen with small prolactin-secreting tumours (microprolactinomas). If a large tumor causing visual failure is a macroprolactinoma the serum prolactin level is typically increased a few hundredfold. The prolactin level is not grossly elevated, the diagnosis is a non-

functioning pituitary macro-adenoma, and the patient should undergo surgery. In the context of sellar expansion and an optic chiasm which is clearly elevated, the surgical approach should be trans-sphenoidal.

6. This depends on his preoperative status. If he has been shown biochemically or clinically to have impaired pituitary function preoperatively, he will have been started on steroid replacement and this should continue postoperatively. The dose should be increased on the day of surgery and for the subsequent days and then returned to maintenance around day 4. If he has normal preoperative pituitary function, perioperative steroid replacement should still be given. However, in this case it is reasonable to measure the morning cortisol level prior to giving steroids replacement on days 4 and 5 postoperatively; if it is normal, steroid replacement is stopped.

7. There are two possibilities. First, the perioperative intravenous fluid regimen may have been excessive and he is offloading fluid physiologically. The management of this is expectant. The second more important diagnosis is diabetes insipidus (DI) due to loss of antidiuretic hormone (ADH). This is a common transient complication of pituitary surgery and may result in severe dehydration and hypernatremia.

If he is developing DI, he will be clinically dehydrated and thirsty. The serum sodium level should be checked urgently. If the diagnosis is DI, the patient should be given a supply of water and instructed to drink according to his thirst. He may be given synthetic ADH (desmopressin, DDAVP) as a subcutaneous injection. The sodium level should be monitored and fluid balance charts maintained until at least day 3 postoperatively.

Case 25

1. Headaches and changes in speech coordination raise the possibility of a cerebellar mass with hydrocephalus. It would be unusual for a supratentorial mass to produce raised intracranial pressure with headaches and focal deficit in relation to mild speech disturbance (which in this case is more suggestive of dysarthria than dysphasia). The slow and minimally progressive nature of the symptoms suggests a benign lesion.

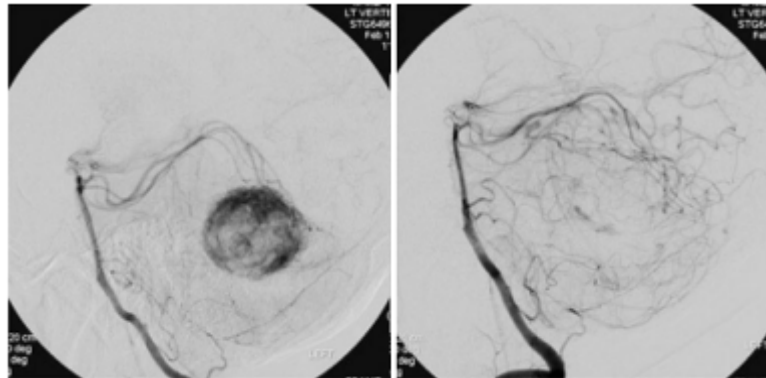
2. The coronal T1 pre- and post-contrast and axial T2 images confirm a well-circumscribed avidly enhancing mass. The axial T2 image through the upper part of the mass shows a number of surrounding black flow voids indicating blood vessels. Enlarged surrounding vessels are a typical feature of cerebellar hemangioblastoma, and the diagnosis should be questioned if they are not seen around a lesion of this size. There is no merit in conservative management for this patient as his symptoms will inevitably progress.

3. Hemangioblastomas are benign (WHO grade I) and therefore may be cured by complete surgical excision. Although usually sporadic, they do occur as part

of von Hippel–Lindau disease, one of the neurocutaneous syndromes, where they may be multiple. They commonly occur as a cystic mass with a non-enhancing wall and a solid enhancing nodule; removal of the nodule alone is the surgical treatment.

Solid hemangioblastomas, such as this one, are less common and enhance throughout. The tumor is highly vascular and an attempt should be made to remove it in one piece (en bloc).

Catheter angiography is useful, as knowledge of the blood supply may aid surgery, and embolization of the feeding vessels, where possible, can reduce vascularity, as shown for the patient in figure below:



The pre- and post-embolization lateral angiograms show an impressive reduction in tumor 'blush' from high vascular flow through the superior cerebellar artery. His surgery was uneventful, with en bloc removal and minimal blood loss. He made a full recovery and was discharged on day 5 postoperatively.

Case 26

1. In any patient presenting with a shunt-related problem, the essential questions to ask are as follows.

About the shunt:

- location of the distal end of the shunt (peritoneum, pleura, superior vena cava)
- type of shunt (pressure setting, programmable or not)
- the diagnosis requiring the shunt
- when the shunt was first inserted
- date and nature of subsequent revisions (which part of the shunt was revised, whether it was removed, etc.).

About the symptoms:

- associated with raised ICP (blocked shunt) — headaches, nausea, vomiting, visual disturbance
- associated with infection — pyrexia, altered mental state
- associated with problems at the distal end of the shunt (e.g. abdominal pain).

Patients and their relatives are often very aware of the presenting symptoms when the shunt has blocked in the past, and it is always worthwhile paying

attention to these symptoms when assessing the patient.

2. Several bedside tests can reveal information about whether the shunt is functioning adequately.

- Papilloedema indicates raised intracranial pressure and is therefore consistent with a blocked shunt. It does not indicate which part of the shunt is blocked.
- Pressing on the reservoir may indicate whether the proximal (ventricular catheter) end is working. Press the valve and release: if the valve refills, the ventricular catheter is patent; if it depresses and does not fill, blockage of the ventricular catheter is a possibility, although a low ICP may also be possible. Even if the valve fills, a blocked shunt is not excluded as the shunt tubing distal to the valve may still be blocked. This is generally an unreliable technique, especially in older shunts, as one may be pressing on a ventricular access reservoir or the valve may have become incompetent.
- Tapping the shunt is a commonly performed test which involves passing a narrow (usually 25G) needle into the reservoir of the shunt valve. A manometer and syringe may be connected to it. There are various results from such a test:
 - CSF easily aspirated at high pressure — suggests distal blockage.
 - CSF easily aspirated at low pressure — shunt may be working, does not exclude blockage.
 - CSF difficult to aspirate — suggests proximal blockage.

3.

- X-ray shunt series enables the shunt tubing to be examined for breakages or disconnections.
- CT brain will enable the size of the ventricles to be assessed. Ventriculomegaly (especially if there is an increase in size of the ventricles compared with a previous CT scan performed when the patient did not have a blocked shunt) is consistent with a blocked shunt. Some patients can have 'stiff ventricles' that do not dilate when under pressure and so a normal CT scan does not exclude a shunt blockage. Ultimately the decision to explore a shunt for potential blockage depends on careful assessment of the patient's symptoms.

4. The patient clinically has a blocked shunt. The fundi should be examined for papilloedema. The shunt valve can be examined to see if it refills after compression. The patient also requires a shunt series X-ray to identify problems with the shunt tubing, and a CT scan to assess the size of the ventricles. The serum inflammatory markers should also be checked as shunt infection can present with similar symptoms, and there may be both infection and blockage (the former leading to the latter). Tapping the shunt can be avoided as it carries the risk of introducing infection and the clinical suspicion of shunt infection is low.

5. The shunt catheter is positioned in the ventricle and the ventricles are not particularly dilated. An old CT when the shunt was known to be working was not available for comparison. Shunt infection is unlikely because the shunt was last operated on a very long time ago. The shunt valve is not filling, which is

consistent with obstruction of the proximal (ventricular) catheter, but this finding in itself is not sufficient evidence for blockage. Confirmation could be sought by measuring the CSF pressure. This can be done by tapping the shunt valve with a needle and attaching a manometer, but this carries the risk of introducing infection into the shunt system. An LP carries less risk of introducing infection. In this patient an LP was considered safe and therefore was performed. It revealed an opening CSF pressure of > 40 cm H₂O. The patient proceeded to surgery, during which the proximal catheter was found to be blocked. This could not be removed as it had adhered to brain over many years. Therefore a new catheter was inserted with the old one still in place. The patient's symptoms resolved postoperatively.

A CT head should be performed after shunt revision so that a baseline scan (with a functioning shunt) is available for comparison if the patient presents with shunt-related problems in the future. The postoperative CT scan of this patient, in which both the old and the new catheter can be seen, is shown in figure below.



Pearls

Patients with a blocked shunt may have normal-sized ventricles on CT. The diagnosis of a blocked shunt must not be excluded on the basis of a CT scan showing normal-sized ventricles.

The decision to explore a shunt is ultimately based on the patient's symptoms.

A patient and his or her family are often the best source of information for how a shunt blockage has presented in the past for that particular patient.

Case 27

1. The inheritance pattern shown in the pedigree is autosomal dominant. There is more than one generation affected with male-to-male transmission, ruling out X-linked inheritance and autosomal recessive inheritance. Transmission by a male also eliminates mitochondrial inheritance.

2. The MRI shows severe midline cerebellar atrophy.

3. This patient has a hereditary ataxia and most probably has one of the spinocerebellar ataxias (SCAs). The hereditary ataxias are a group of

neurogenetic disorders producing slowly progressive incoordination of gait, associated with dysmetria, dysarthria and poor coordination of eye movements. These are slowly progressive and often associated with cerebellar atrophy. The ADCA includes 10 SCAs, SCA1–11 (SCA 9 not known), plus DRPLA and two episodic ataxias. Great overlap is present in the clinical manifestations of these SCAs, both in terms of age of onset and physical findings. There are a few distinguishing features for each type. SCA2 shows early slow saccadic eye movements and hyporeflexia. In SCA4 a sensory neuropathy is common. SCA5 has an early-onset ataxia with a slow prolonged course. SCA6 has a later onset of an isolated ataxia with a slow prolonged course. SCA7 often has visual loss with retinopathy. SCA10 presents with ataxia and sometimes seizures. SCA11 is an uncomplicated cerebellar ataxia. DRPLA would be another diagnostic consideration, but is less likely in this patient who does not have seizures, dementia or chorea.

4. There is highly specific DNA testing available for SCA types 1–3, 6, 7 and for DRPLA. All have trinucleotide expansions in their genes. Tests are not presently clinically available for SCA4, 5, 8, 10, 11.

Case 28

1. The pedigree could show either an autosomal dominant or an X-linked inheritance pattern. There is no male-to-male transmission seen in this pedigree which would confirm autosomal dominant transmission and eliminate X-linked transmission. Transmission by a male eliminates mitochondrial inheritance.

2. He could have CMT1A, CMT1B or CMTX. Charcot–Marie–Tooth disease is the most common genetic cause of neuropathy. It is a group of disorders that produce both motor and sensory neuropathy. There is considerable genetic heterogeneity, that is, many different genes can cause the same phenotype. It can be inherited in an autosomal dominant, autosomal recessive or X-linked manner. The pedigree in this case eliminates autosomal recessive types of CMT. The autosomal dominant form has been divided into two major categories based on the NCVs: CMT1 with slow NCVs and CMT2 with normal or near normal NCVs. CMT1 subtypes A and B are clinically indistinguishable and are designated solely on molecular findings. CMT1B tends to be more disabling and with very slow NCVs, 5–20 m/s. However, some cases of CMT1B are clinically identical to CMT1A. CMT2 has NCVs in the normal, or mildly abnormal range and is unlikely in this case with moderately slow NCVs. Males affected with X-linked CMT (CMTX) have the full syndrome and females may be clinically normal or have mild to moderate signs and symptoms.

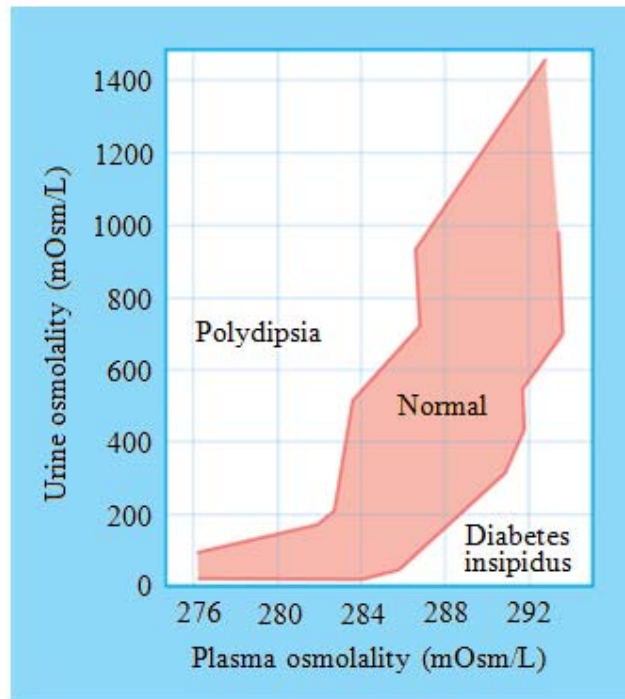
3. The connexin 32 mutation is seen in CMTX. This means that the patient has X-linked CMT. The risk for his daughters of inheriting the gene is 100%, as he only has one X chromosome, which must have the mutation. For his sons the risk is 0% because they will only inherit his Y chromosome and not his X chromosome.

Case 29

1. The CT (a) shows enlarged lateral ventricles and several intraparenchymal and subarachnoid cysts without surrounding edema. The MR images (b, c) show an intraventricular thin-walled cyst, two parenchymal cysts at the right parietal gray–white junction, and a left parietal subarachnoid cyst. One of the parietal cysts contains an enhancing serpiginous nodule (arrow).
2. The constellation of intraparenchymal, intraventricular, and subarachnoid thin-walled cysts in a Central American patient presenting with seizure indicates CNS cysticercosis in its vesicular form. The visualization of an enhancing intracyst nodule is diagnostic of the cysticercotic scolex in this setting. Infection of the CNS by the larval stage of the pork tapeworm, *Taenia solium*, results in formation of intraventricular, intraparenchymal and subarachnoid cysts. The living organism is protected from the immune system and is often asymptomatic for long periods (vesicular stage). When the larva dies, an inflammatory reaction ensues and seizures or focal neurologic symptoms can occur. At this stage (vesiculo-nodular), peripheral edema and ring-enhancement are commonly seen on CT and MRI. Involuting cysts may show enhancement without edema (granular nodular stage). Involved cysts eventually calcify (nodular calcified stage). FLAIR images are particularly helpful in demonstrating intraventricular cysts, since intraventricular CSF signal is suppressed whilst proteinaceous fluid remains bright.

Case 30

There are three recognized patterns of post-operative DI that can be caused by damage to the posterior lobe of the pituitary or stalk during trans-sphenoidal decompression. The majority of patients experience transient DI which lasts 12–36 hours post-operatively, characterized by supra-normal urine output and polydipsia. A minority of patients experience prolonged DI which can last months or permanently in up to one-third of this group. A triphasic pattern of DI is observed in an even smaller percentage of patients: characterized first by DI, followed by normalization or SIADH, and finally DI which can be either transient or permanent. Other laboratory information helpful in making the diagnosis of DI includes Na^+ level and urine specific gravity.



For patients who can tolerate p.o. intake the optimal treatment for DI is to encourage the patient to drink water. However, when UO exceeds the amount that can be comfortably replaced with p.o. or IV fluids (i.e. >300 ml/4 hours), then a vasopressin preparation is appropriate treatment.