CASE SEVEN

Short case number: 3_22_7
Category: Neurology
Discipline: Medicine
Setting: Emergency Department
Topic: Intracranial tumours and hydrocephalus [SDL]

Case

Laura Kwa, aged 39 years, presents with intractable headache that has failed to respond to migraine treatment or pethidine. She also complains of mild weakness in her left hand of recent onset. She denies any seizures or previous history of migraine, and has otherwise been well.

Questions

1. Outline the classical presentation of a brain tumour (primary or secondary).
2. List in a table the common primary malignant and benign intracranial tumours.
3. Outline your management of Laura in this case including further history, examination and investigations you would undertake.
4. Outline the principles of medical and surgical management of intracranial tumours.
5. Summarise the difference between communicating and obstructive hydrocephalus.
6. Summarise the presentation of idiopathic intracranial hypertension.
7. Outline the investigation and management of a patient with suspected intracranial hypertension.

Suggested reading:

- Colledge NR, Walker BR, Ralston SH, Penman ID, editors. Davidson's Principles and Practice of Medicine. 22nd edition. Edinburgh: Churchill Livingstone; 2014. Chapter 26.

ANSWERS

1. Outline the classical presentation of a brain tumour (primary or secondary).

Cerebral mass lesions will tend to increase intracerebral pressure, but the amount by which the pressure is raised depends on the rate of growth of the mass. If it is slow, various compensatory mechanisms may occur, including alteration in the volume of fluid in CSF spaces and venous sinuses, thereby allowing some tumours to achieve considerable size. More rapid growth (as in highly malignant tumours or abscesses) does not allow the compensatory mechanisms to take place, so raised intracranial pressure develops early, especially if the CSF circulation is also obstructed. Papilloedema is not always present, either because raised intracranial pressure has developed too recently, or because of anatomic anomalies of the meningeal sheath of the optic nerve. Vomiting, bradycardia and arterial hypertension develop as late features of raised intracranial pressure and usually parallel the other clinical signs; sudden vomiting may be an early feature of tumours of the cerebellum, especially in children.

2. List in a table the common primary malignant and benign intracranial tumours.

PRIMARY MALIGNANT INTRACRANIAL TUMOURS

Histological type	Common site	Age
Glioma (astrocytoma**)**	Cerebral hemisphere Cerebellum Brain stem	Adulthood Childhood/adulthood Childhood/young adulthood
Oligodendroglioma	Cerebral hemisphere	Adulthood
Medulloblastoma	Posterior fossa	Childhood
Ependymoma	Posterior fossa	Childhood/adolescence
Cerebral lymphoma (microglioma)	Cerebral hemisphere	Adulthood

PRIMARY BENIGN INTRACRANIAL TUMOURS

Histological type	Common site	Age
Meningioma	Cortical dura Parasagittal Sphenoid ridge Suprasellar Olfactory groove	Adulthood
Neurofibroma	Acoustic neuroma	Adulthood
Craniopharyngioma	Suprasellar	Childhood/adolescence
Pituitary adenoma	Pituitary fossa	Adulthood
Colloid cyst	Third ventricle	Any age
Pineal tumours	Quadrigeminal cistern	Childhood (teratomas) Young adulthood (germ cell)

3. **Outline your management of Laura in this case including further history, examination and investigations you would undertake.**

The history of the headache and any other neurological changes, including cognition needs to be ascertained. A headache in the morning that wakes a person from sleep and is associated with nausea and vomiting is suggestive of raised intracranial pressure. Other features such as neck stiffness and photophobia suggest involvement of the meninges. A more acute onset of symptoms suggests bleeding or seizures.
Other systematic symptoms need to be sought such as weight loss and general fatigue. These may can be ominous and highly suggestive of systemic cancer.
Other historical features include family history, smoking, past medical problems.
Physical examination includes a full neurological examination to localise the problem to a particular site in the brain.
A head CT scan is indicated.

4. **Outline the principles of medical and surgical management of intracranial tumours.**

Medical
Relief of raised intracranial pressure is often required when surgery is not possible or when life is threatened before investigation has revealed the diagnosis. Dexamethasone, 8 mg 12-hourly either orally or by injection, is used to lower intracranial pressure by resolving the reactive oedema around a tumour. A striking improvement in conscious level is often produced and focal disabilities may regress. In severe and acutely raised intracranial pressure, 16-20 mg of dexamethasone may be given intravenously or 200 ml of a 20% solution of mannitol may be infused.
Surgical
Surgery is the mainstay of treatment, although only partial excision may be possible if the tumour is inaccessible or if its removal is likely to cause unacceptable brain damage. Biopsy by a direct or stereotactic technique should be considered even if the tumour cannot be removed, since the histological diagnosis has important implications for management and prognosis.
Meningiomas and acoustic neuromas offer the best prospects for complete removal without unacceptable damage to surrounding structures. Meningiomas can recur, particularly those of the sphenoid ridge when partial excision is often all that is possible. Pituitary adenomas can often be removed by a trans-sphenoidal route, thereby avoiding the need for a craniotomy.

5. **Summarise the difference between communicating and obstructive hydrocephalus.**

Hydrocephalus (dilatation of the ventricular system) may be due to obstruction of the CSF circulation. Hydrocephalus is said to be 'communicating' if the obstruction is outside the ventricular system (usually in the basal cisterns). Obstruction within the ventricles is most common in the narrow channels of the third ventricle and aqueduct, and may be caused by tumour or a congenital anomaly such as aqueduct stenosis.

6. **Summarise the presentation of idiopathic intracranial hypertension.**

This condition, previously known as 'benign intracranial hypertension' and 'pseudotumour cerebri', usually occurs in obese young women. Raised intracranial pressure develops without a space-occupying lesion, ventricular dilatation or impairment of consciousness. The aetiology is uncertain but there may be a diffuse defect of CSF reabsorption by the arachnoid villi. The condition can be precipitated by drugs, including tetracycline, and rarely vitamin A, retinoids, Addison's disease and withdrawal of corticosteroid therapy.
Characteristically, there is a headache, sometimes with transient diplopia and visual obscuration, but few other symptoms. There are usually no signs other than papilloedema, which may be discovered incidentally at a routine visit to an optician, but a 6th nerve palsy may be present.

7. **Outline the investigation and management of a patient with suspected intracranial hypertension**

A CT is normal, with normal-sized or small ventricles. Once this has been demonstrated, a lumbar puncture is safe and will allow confirmation of the raised CSF pressure and form part of treatment. MR angiography or cerebral venography will exclude cerebral venous sinus thrombosis or stenosis of other cause. True papilloedema may need to be distinguished from other causes of disc swelling by fluorescein angiography.
Ay precipitating condition should be sought, relevant medication should be withdrawn and a weight-reducing diet instigated, if indicated. The carbonic anhydrase inhibitor, acetazolamide, may help to lower intracranial pressure. Repeated lumbar puncture can be considered, but is often unacceptable to the patient. Patients failing to respond, in whom chronic papilloedema threatens vision, may require optic nerve sheath fenestration or a lumbo-peritoneal shunt