Reminder of important clinical lesson

Transient obscuration of vision

Robert Corns, Lamia Nayeb, Andrew Martin

Department of Neurosurgery, St George's Hospital, London, UK

Correspondence to Robert Corns, clifodan@hotmail.com

Summary

A previously well 13-year-old girl presented with a 4-month history of progressively worsening headache and visual disturbance. At fourth presentation, examination showed bilateral papilloedema and a left homonymous hemianopia. CT and MRI scans revealed a tumour obstructing the third ventricle with consequent hydrocephalus. Following uncomplicated endoscopic surgery, both headache and visual function improved. Several features of the history were highly suggestive of raised intracranial pressure and were overlooked initially.

BACKGROUND

Headache is a symptom frequently reported to general practitioners (GPs) and raised intracranial pressure is a rare but important cause of this. There are certain features which are characteristic and this case report highlights some of them.

CASE PRESENTATION

A 13-year-old left-handed schoolgirl presents with a 4-month history of increasingly severe headache, worse in the morning. There was no associated nausea and vomiting. She had visited her GP on three occasions prior to the detection of the papilloedema. The absence of neurological deficit or associated nausea and vomiting was falsely reassuring.

She also reported transient visual symptoms, described as bright flashing lights in the periphery of her vision or 'graying' of her vision, especially when moving from sitting to standing.

Her medical history was unremarkable. She was a normal vaginal delivery at term with subsequent normal development. Menarche was 1 month prior to presentation, that is, normal; there were no features to suggest endocrine disturbance. She had normal schooling and was doing well.

On examination, she was alert and orientated. She had bilateral papilloedema, visual acuity was right 6/18 correcting to 6/6 with pin-hole. Left was 6/6. Formal field testing revealed a bitemporal hemianopia. There was no other focal neurological deficit.

INVESTIGATIONS

Initial CT scan revealed a calcified, supra-sellar cystic tumour with biventricular hydrocephalus. MRI confirmed these findings, which were in keeping with a craniopharyngioma. Pituitary function tests were normal (figures 1–3).

DIFFERENTIAL DIAGNOSIS

- ▶ Benign headache
- ▶ Migraine
- ▶ Raised intracranial pressure due to
 - Hydrocephalus
 - Tumour
 - Other

- ▶ Benign intracranial hypertension
- ▶ Optic neuritis

TREATMENT

The following day she underwent a transventricular endoscopic drainage of the cyst, biopsy and fenestration of the septum pellucidum. A catheter connected to a subcutaneous reservoir was left within the cystic for later percutaneous aspiration or conversion to a shunt, should they be needed. Histology confirmed this to be a craniopharyngioma. A postoperative CT shows resolution of the hydrocephalus and catheter left within the tumour cyst.

Figure 1 Unenhanced axial CT showing a calcified cystic lesion (black arrow) in the region of the pituitary gland with associated hydrocephalus (enlarged temporal horns, white arrow).

Figure 2 (A) Coronal T2 FLAIR, (B) axial T2 and (C) sagittal T1 MRIs showing a cystic lesion arising in the pituitary sella and extending superiorly into the third ventricle.

OUTCOME AND FOLLOW-UP

The patient's headaches resolved and vision normalised postoperatively. She was discharged home soon afterwards without complication and has received stereotactic conformal radiotherapy.

DISCUSSION

Several associated features of headache should be regarded as 'red flag' signs and prompt further investigation including: papilloedema, focal neurological deficit, new persistent and unremitting headache, with or without vomiting. Crucially, associated transient obscuration of vision is a very significant finding, as it is pathognomonic of papilloedema and always requires further investigation.

Raised intracranial pressure (ICP) develops as the consequence of either a space-occupying lesion or increased cerebrospinal fluid (CSF) volume within the rigid confines of the skull. Hydrocephalus may be caused by a relatively

small lesion obstructing the normal drainage of CSF. Cerebral oedema surrounding tumours and other lesions may further exacerbate the increase in ICP. The headache of raised ICP is classically worse in the morning and on bending, coughing or straining. The underlying mechanism of Valsalva manoeuvres (with coughing or straining) causing venous stasis leading to episodic increased ICP.

Vomiting is often a feature of raised ICP but its absence, as in the case, can be falsely reassuring. Papilloedema occurs when CSF pressure is high enough to cause swelling of the optic discs. Unchecked, increasing ICP will ultimately result in brain herniation and death.

Altered consciousness, lethargy and seizures are late features of raised ICP. In infants, increasing head circumference, bulging fontanelles and suture separation are features specific to this age group.

Causes of raised ICP in adults include space-occupying lesions, such as haemorrhages, tumours (primary or metastatic) and brain abscess. Focal oedema developing around these can contribute to intracranial pressure rise. Meningitis and encephalitis result in diffuse brain oedema and raised ICP, as can subarachnoid haemorrhage, diffuse axonal injury and water intoxication. Excess CSF results in hydrocephalus and raised ICP. Haematomas act as space-occupying lesions and also bring about raised ICP. Benign intracranial hypertension is an idiopathic condition which results in increased CSF pressure without increased CSF volume.

The causes of raised ICP in children are similar to those in adults; notably, brain tumours are more likely to be primary in nature, with metastatic disease being uncommon in this population. While brain tumours are overall rare across the lifespan, they are nonetheless the second most frequent malignancy of childhood after leukaemia and lymphoma.

Craniopharyngiomas constitute 15% of paediatric brain tumour.¹ Craniopharyngiomas are tumours which arise from embryonic epithelial tissue in the pituitary gland and typically present with headache, vomiting, visual disturbance and hydrocephalus.²⁻⁷ They may also present with tumour-induced endocrine disturbance including Addison's syndrome, hypothyroidism, hypogonadism and growth retardation.

The management of craniopharyngiomata remains controversial.⁸ It is a benign tumour and therefore complete excision can lead to a cure. However, complete surgical resection is often associated with unacceptable, devastating hypothalamic injury. Treatment is determined by size of the tumour and involvement of surrounding structures

Figure 3 Unenhanced axial showing the postoperative appearances with decompressed ventricles and catheter left within the tumour cyst (this can be aspirated, should the cyst recur).

such as the hypothalamus and optic chiasm; treatment often involves a tissue diagnosis and radiotherapy.^{9 10} They are often cystic which can be dealt with by endoscopic and/or stereotactic drainage. Survival rates for children at 5 years are as high as 83–99%; however, complete cure is usually elusive.¹¹

Learning points

- ▶ This young lady was seen on three occasions prior to diagnosis and a number of important 'red flag' indicators of serious underlying pathology were overlooked, including:
 - Transient obscuration of vision
 - Papilloedema
 - New headache which is persistent and unremitting
 - Focal neurological deficit (bitemporal hemianopia).
- ▶ A thorough history and neurological examination may have detected these features earlier.

Competing interests None.

Patient consent Obtained.

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