CASE REPORT

The isolated fourth ventricle

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hamandik@cardiff.ac.uk**SUMMARY**

Isolated enlargement of the fourth ventricle, or 'encysted' fourth ventricle is a rare late complication following shunt insertion of the lateral ventricles for hydrocephalus. Caudal and rostral obstruction of the fourth ventricle and its subsequent dilation results in compression of adjacent cerebellum and brain stem structures; treatment with further shunt insertion directly to the fourth ventricle is invariably successful. There is potential for diagnostic delay, when clinical symptoms and signs of cerebellar and brain stem compromise are unrecognised or attributed to other factors, and attention on the CT is focused on the lateral ventricular system and the already existing ventriculoperitoneal shunt, which will appear unchanged from previous scans. We report two cases with isolated fourth ventricular obstruction and review the literature to highlight the importance of recognising this condition.

BACKGROUND

We report our experience of two cases with a late complication of hydrocephalus after lateral ventricular shunting.

Although the neurosurgeons and neurologists are familiar with this condition, the wider medical community may be less familiar. Patients will however present to general practitioners, accident and emergency doctors, acute physicians, as well as neurologists and neurosurgeons.

The aim of this article is to contribute to education and awareness of this condition.

CASE PRESENTATION**Case 1**

A 36-year-old woman, presented with a 2-month history of increasing drowsiness, deteriorating mobility, visual disturbance and balance problems. Five years previously she had sustained an open right skull fracture with cerebral contusion from an assault. At that time, a craniotomy was carried out for cerebral decompression and debridement, and ventriculoperitoneal (VP) shunt was also inserted. Postoperative recovery was complicated by persistent wound and bone flap infection and the craniotomy bone was removed, leaving a permanent skull defect. Following a prolonged period of rehabilitation, she was left with dense left-sided hemiparesis with left-sided neglect, but was able to go home. Prior to the recent deterioration she would be alert and orientated, able to hold a conversation and use a wheelchair with a good sitting balance. She was able to transfer independently. She had epilepsy controlled with low dose lamotrigine and infrequent seizures.

On examination she was drowsy but rousable, Glasgow Coma Scale was 14/15 (E4, M6, V4). There was an obvious right skull defect. She had sluggish pupillary reactions with normal visual acuity. She had vertical gaze palsy, convergence paralysis, retraction nystagmus, this combination of signs is known as the dorsal mid-brain or Parinaud syndrome^{1 2} and 'sunset sign' of the eyes (the sclera being visible between the upper eyelid and the iris, seen usually in hydrocephalus due to loss of upward conjugate gaze caused by raised intracranial pressure). She was dysarthric with severe spastic hemiparesis of the left side with 0/5 power on the MRC scale. Sensory examination revealed left hemineglect.

Case 2

A 29-year-old man, presented with five generalised tonic-clonic seizures and reduced consciousness after accidentally falling downstairs at home.

Thirteen years previously, he had had a traumatic head injury with significant cerebellar contusion, he developed hydrocephalus which was treated with a VP shunt to the left lateral ventricle. Following a prolonged period of rehabilitation, he was discharged home with memory deficits and word finding and communication difficulties. He was independently mobile. He developed epilepsy, which was well controlled with carbamazepine 500 mg/day (the last seizure 6 months prior to admission).

Neurological examination revealed horizontal nystagmus on lateral gaze. Remainder of cranial nerve examination was unremarkable and there were no signs in the limbs. He had independent gait but was noted to be mildly unsteady.

One week after his admission, his gait deteriorated abruptly, with repeated falls on the ward and needing assistance of two to mobilise. On examination, he had severe coarse nystagmus in all directions. His speech was dysarthric. He had severe limb and truncal ataxia.

INVESTIGATIONS**Case 1**

CT scan of the brain showed the old right frontoparietal craniotomy and low density in the right cerebral hemisphere, and VP shunt tip in the trigone of the left lateral ventricle (figure 1A). Cerebrospinal fluid (CSF) analysis, from the shunt reservoir: glucose of 3.7 mmol/l and protein of 0.26 g/l, with no cells or growth. Subsequent review of her CT scan of the brain in comparison with the old CT scan identified marked dilation of the aqueduct of sylvius and fourth ventricle (figure 1B).

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Figure 1 CT Brain scans patient 1, upper panel: (A and B) from admission scan, showing in (A) the previous head injury and skull defect, collapsed ventricular system and ventriculoperitoneal (VP) shunt in situ (thin arrow). Block arrow in (B) shows the dilated fourth ventricle, and (C) after fourth ventricular shunt (thin arrow) reduction of fourth ventricle. Patient 2, lower panel: (D) and (E) scan on admission, small right subdural (dotted arrow), VP shunt in situ (thin arrow) and dilated fourth ventricle (block arrow). (F) Repeat scan at 10 days from admission shows marked increase in size of fourth ventricle and (G) following fourth ventricular shunt.

Case 2

Brain CT scan, on admission, showed an acute right-sided subdural haematoma, low density and atrophy of the cerebellum and caudate heads and a VP shunt in situ (figure 1D).

Repeat CT scan showed progressive enlargement of the fourth ventricle (figure 1F).

TREATMENT

Case 1

She underwent an urgent fourth ventricular decompression, postoperative CT (figure 1C). She made a good recovery to previous levels of functioning.

Case 2

He underwent a fourth ventricular decompression shunt insertion. A postoperative brain CT scan showed reduction in the size of the fourth ventricle (figure 1G).

OUTCOME AND FOLLOW-UP

Case 1

She was able to go back home and remains well at 4 years follow-up.

Figure 2 MRI T1 sagittal, demonstrating the anatomy of midbrain and brainstem: 1, pons; 2, medulla; 3, cerebellum; 4, fourth ventricle; 5, aqueduct of sylvius and 6, dorsal midbrain.

Case 2

Over the subsequent days, the patient's balance improved and he was discharged home after 2 months of rehabilitation.

DISCUSSION

We present two cases of isolated fourth ventricle. In case 1 recognition of new neurological problems developing was not initially appreciated due to the slowly progressive nature of her deterioration and sequelae of previous severe brain injury. In the second case truncal ataxia was initially suspected to be due to antiepileptic drug toxicity. Both patients made a good recovery to premorbid states after appropriate neurosurgical fourth ventricle decompression and rehabilitation.

Hydrocephalus is a common complication following cerebral insult with infection, trauma or haemorrhage. Treatment is typically by CSF diversion with VP shunt insertion with the proximal end in a lateral ventricle. The UK shunt registry estimates over 3000 CSF shunt operations in the UK every year, with more than 43 000 operations since it was established in 1995³ and presentation to acute medical intake with suspected shunt complications are common.

There is an estimated 140 ml of CSF surrounding the brain and the spinal cord. Around 480 ml are produced every day, and CSF is circulated three times daily. CSF is produced by the choroid plexus (80%), and the brain parenchyma. The lateral ventricles produce most of the CSF, which then passes through the foramen of Monro to the third ventricle, and from there to the fourth ventricle through the aqueduct of sylvius. The fourth ventricle is bounded by the pons and the upper medulla anteriorly and the cerebellum posteriorly. It is in continuity with the central canal of the spinal cord inferiorly. The fourth ventricle has two lateral apertures (the foramina of Luschka) and one median aperture (the foramen of Magendie) through which the CSF passes to the subarachnoid space (figure 2). The CSF then gets absorbed through the arachnoid villi into the cerebral venous sinuses and the venous system.⁴

Isolated fourth ventricle, first described by Dandy⁵ in 1921 is a rare late complication following insertion of lateral ventricular shunt for hydrocephalus in children and adults.^{6–9} Functional collapse of the walls of the aqueduct of sylvius occurs due to change in the pressure gradient across the tentorium, induced by the lateral ventricular shunt. Closure of the fourth ventricular

outlet isolates the fourth ventricle from the rest of the ventricular system; the choroid plexus in the fourth ventricle continues to produce CSF, leading to subsequent dilation of the fourth ventricle and progressive compression of the adjacent structures.^{8–10} Ataxia, nystagmus, dysarthria, pyramidal weakness, cranial nerve palsies, eye movement disorders and symptoms of increased intracranial pressure.^{6 11 12}

Isolated fourth ventricle is reported at around 2.5% of some series, with an interval from VP shunt insertion of 1–7 years.^{13 14} Prematurity, intraventricular haemorrhage and posthaemorrhagic or postinfective hydrocephalus are risk factors, causing ependymal inflammation and closure of the fourth ventricle outlet.⁷

Isolated fourth ventricle is easily overlooked and can be missed on initial medical assessment and radiology review. As ever, knowledge of the condition and high index of suspicion is important.

Learning points

- Isolated fourth ventricle is rare but important complication of lateral ventricular shunting for hydrocephalus.
- Isolated fourth ventricle is easily overlooked late and can be missed on initial medical assessment and radiology review. As ever, knowledge of the condition and high index of suspicion is important.
- Fourth ventricular shunt is the most acceptable procedure with good results.

Competing interests None.

Patient consent Obtained.

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