CASE REPORT

Cranial nerve palsies after shunting of an isolated fourth ventricle

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SUMMARY

An isolated fourth ventricle is characterised by cerebrospinal fluid (CSF) trapping in the fourth ventricle. Although there is no consensus regarding treatment, ventriculoperitoneal (VP) shunting of the fourth ventricle is an option. Complications include infection, mechanical irritation of the brainstem, malfunction and overdrainage. Cranial nerve palsy is a rare complication and has been mostly described in children. We present two adult cases of abducens and facial nerve palsies occurring secondary to this procedure. Placement of a higher resistance valve brought about complete recovery in one patient while withdrawal of the catheter by a few millimetres led to complete recovery in the second patient.

BACKGROUND

An isolated fourth ventricle is an uncommon phenomenon, mostly described in children.^{1–10} Usually, it occurs with ventricular shunts placed for communicating hydrocephalus following haemorrhage or meningitis.^{1–10} There is no consensus concerning appropriate treatment, but cerebrospinal fluid diversion from the fourth ventricle is the most commonly performed procedure. Complications are as for any routine shunt surgery, such as infection, dysfunction, catheter displacement and overdrainage.

Cranial nerve deficits have been described following fourth ventricle catheter placement. Most of the reports concern 6th and 7th nerve palsies, but 5th, 10th and 12th cranial nerve palsies have also been described.^{1 6 7 11–13} These complications, however, have been described mostly in children already having a VP shunt in the lateral ventricle since infancy. Isolated fourth ventricle is much less common in adults, and cranial nerve palsy following fourth ventricle shunting is a rare complication of the procedure. We describe two patients who developed cranial nerve deficits following shunting. The first developed progressive bilateral 6th, 7th and 10th nerve palsies after surgery, while the second patient presented with bilateral 6th and 7th cranial nerve palsies directly after the procedure. Clinical history and physiopathological mechanisms are discussed in the context of recent literature, with a view to improving management of this rare phenomenon.

CASE PRESENTATION

Case 1

A 52-year-old woman was operated for a left petroclival meningioma. Postoperatively, a right

ventriculoperitoneal shunt (VPS) was placed because of progressive hydrocephalus (Sophy 110 mm H₂O). Several months later, the VPS had to be removed because of meningitis, with transitory placement of an external ventricular drain.

After resolution of the infection, a ventriculocisternostomy was attempted, due to suspicion of an obstructive hydrocephalus. Nevertheless, the symptoms recurred associated with progressive dilation of the ventricular system. A left VPS was placed (Codman Hakim valve, 100 mm H₂O). The system was revised a few months later, and replaced by a Medtronic fixed low pressure valve.

Several years later, the patient presented with progressive unsteadiness of gait, nausea and occipital headaches. CT scan showed an isolated fourth ventricle enlargement (figure 1, upper left panel).

In order to not reopen the site of previous surgery, and because of the procedure's ease and our team's experience, neuronavigation was used to place a VPS in the fourth ventricle through a suboccipital burr hole and a transvermian route, with a 'Y' connection to the previous VPS (with fixed low pressure valve). Postoperatively, symptoms improved gradually, but 8 days after surgery, the patient progressively developed diplopia, squint, dysarthria and dysphagia, and was unable to close her eyes completely because of facial muscle weakness. Clinical examination showed bilateral abducens, and facial (House-Brackmann IV) and vagus nerve palsies (figure 1, lower right panel).

MRI showed that the catheter was perfectly placed in the centre of the fourth ventricle, which showed a significant size reduction (figure 1, lower left panel). We suspected overdrainage and the fixed low pressure valve was replaced by a new Codman Hakim programmable valve, with the pressure set at 140 mm H₂O. Postoperatively, the cranial nerve deficits improved. However, the patient presented with increasing unsteadiness, and subsequent pressure adjustments were performed in order to find the optimal resistance to balance between cerebellar symptoms and cranial nerve deficit. Three months after the last operation, the patient had a normal neurological examination with valve pressure set at 80 mm H₂O.

Case 2

An 18-year-old man presented with a right cerebellar haemorrhage due to a ruptured arteriovenous malformation (AVM). Clinically, the patient presented a right static and kinetic cerebellar syndrome. Suboccipital craniotomy was performed, and the AVM was excised.

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Figure 1 Upper left panel: Preoperative sagittal T2-weighted MRI showing isolated fourth ventricle enlargement. Upper right panel: Postoperative axial CT scan showing a catheter tip away from the brainstem. Lower left panel: Postoperative sagittal T1-weighted MRI showing reduction of fourth ventricle enlargement. Lower right panel: Bilateral facial and abducens nerve palsies ('Look to the right', 'Look to the left', 'Smile').

Two days later, a symptomatic cerebellar oedema was observed and decompressed with a suboccipital craniectomy. Three weeks later the patient developed a communicating hydrocephalus treated with a VPS. Over the following months, he underwent two revisions for infection and dysfunction, and another VPS was finally placed in the occipital horn of the right ventricle, with an anti-siphon Medos valve set at 80 mm H₂O. The evolution was progressively positive and the patient attended neurorehabilitation. Nevertheless, during the next few weeks, he presented with progressive unsteadiness. MRI showed an isolated enlarged fourth ventricle, with a displacement of the cerebellar tonsils towards the foramen magnum inducing an iatrogenic 'Chiari-like' image and a cervicothoracic syrinx (figure 2, upper left panel).

Therefore, a drain was placed in the fourth ventricle through the same approach as used for case 1, with a Y connector. Immediately after surgery, the patient developed bilateral abducens and facial nerve palsy. Unlike case 1, the valve adjustment was difficult to perform, so it was decided to install a separate and independent shunt system with a first valve at 120 mm H₂O for the lateral and third ventricle, and a second valve adjusted at 80 mm H₂O for the fourth ventricle. After a few weeks of rehabilitation, the cranial nerve palsies improved. Postoperative MRI showed reduction of fourth ventricle enlargement (figure 2, upper right panel).

During the next 1 year, the patient underwent multiple revisions for infection and dysfunction, and both the supratentorial

and fourth ventricle drains were removed, with a period of external drainage.

A few months later, the patient underwent removal of the drainage system because of a recurrent *Pneumococcus* infection. After treatment of the infection, new drains had to be inserted since symptoms had recurred. The lateral ventricle drain was placed left occipital. Because of numerous previous suboccipital surgeries, the interhemispheric approach was almost impossible due to the scars and adhesions, so we decided to insert the drain to the fourth ventricle through a trans-cerebellar approach on the left side. Both systems were draining independently with two different valves set to the same values as previously. After the procedure, the patient presented a rapidly progressive right abducens and facial nerve palsy House-Brackmann grade IV. The left side was not affected. This asymmetrical clinical finding could be explained by the fact that, with the progressive shrinkage of the fourth ventricle, the tip of the catheter entering the ventricle with an oblique angle was in contact with the right facial colliculus. Therefore we decided to withdraw the fourth ventricle catheter by 8 mm.

After this correction, the patient presented immediately an improvement of his right facial and abducens nerve function but never completely returned to normal.

DISCUSSION

An isolated fourth ventricle is a relatively uncommon complication, occurring in only 2–3% of cases with long-term VPS.^{1 7}

Figure 2 Upper left panel: Preoperative sagittal T1-weighted MRI showing fourth ventricle enlargement. Upper right panel: Postoperative sagittal T1-weighted MRI showing reduction of fourth ventricle enlargement. Lower left panel: Postoperative sagittal T2-weighted MRI showing catheter tip in contact with the brainstem (arrow). Lower right panel: Postoperative axial T2-weighted MRI showing the catheter tip in contact with the middle cerebellum peduncle.

Most of these patients had undergone several shunt revisions.^{1–10} As such, it is assumed that the inflammatory conditions lead to blockage of CSF flow produced by the fourth ventricle choroid plexus.

Fourth ventricle shunting is the most popular procedure for an isolated fourth ventricle.¹⁴ Other techniques, namely aqueductoplasty, endoscopic aqueductal stenting and other shunting procedures, have been proposed over the past two decades. Concerning aqueductoplasty, some authors report a very high restenosis rate (39%), without stent placement.¹⁵ Transaqueductal stenting shows promising results, but we believe that treatment of an isolated fourth ventricle should be based on individual anatomy. Specifically, in our patients, the presence of narrow supratentorial ventricles in our first patient, no proper alignment of the aqueduct and foramen of Monroe in both cases, and multiple previous surgeries, were arguments against an endoscopic procedure.

Concerning the chosen shunting procedure, a suboccipital transvermian route was preferred in order not to reopen the site of previous surgery, and because of the procedure's ease and our team's experience.

Postoperative cranial nerve palsies are uncommon after fourth ventricle shunting and most of the reports concern children.^{1–10} Lee *et al*⁴ described immediate sixth and seventh nerve palsies after shunting, which were attributed to a direct injury to facial colliculus from the catheter at the time of the procedure. Eder *et al*¹ suggested another mechanism in two cases who presented with sixth and seventh nerve palsies several weeks after surgery,

and in whom the catheter tip was initially located in the centre of the fourth ventricle. In fact, MRI performed several weeks after surgery revealed a collapsed ventricle and the catheter tip appeared to 'touch' the brainstem. Finally, Pang *et al*⁷ described a patient with progressive and bilateral 6th, 7th, 10th and 12th cranial nerve palsies following shunt placement. In his report, the brainstem retracted considerably from the clivus, and traction injury to the cranial nerves was suspected.

In both our cases, the cranial nerve palsies being progressive and bilateral seem to correlate more likely with a gradual nerve traction than direct facial colliculus damage. Moreover, the catheter tip was not in contact with the brainstem. Finally, multiple cranial nerve and 10th nerve involvement suggests that backward displacement of the brainstem and traction on the cranial nerve is the most probable cause of the clinical presentation of our first patient.

Surprisingly, the second patient presented progressive sixth and seventh nerve palsies on the right after the last revision of the fourth ventricle shunting. The progressive and unilateral presentation does not suggest a direct injury during the placement of the catheter; however, this clinical finding is more in favour of a secondary mechanical impingement of the right facial colliculus due to size reduction of the fourth ventricle. The fact that the facial and abducens nerve function improved rapidly after shortening the catheter favours this hypothesis.

In order to avoid these complications, first, we suggest avoiding rapid emptying of the fourth ventricle, and advise trying not to induce massive CSF loss during surgery. We propose to insert

a programmable valve, independent from the previous lateral VPS, and to start to adjust it at a relatively high resistance, which can be decreased slowly and progressively over time.

A flow-regulated valve might also be an option, because pressure-regulated valves might be difficult to adjust, and changes occur between supine and upright position. Some have advocated the use of a 'Y' connector between the fourth ventricle catheter and the existing lateral ventricle drainage.¹¹ Because the compliance and pressure of the 'isolated' fourth ventricle and supratentorial ventricles are different, a 'Y' connection between the lateral and fourth ventricle does not seem to prevent siphoning of the isolated fourth ventricle. Post-shunting imaging almost always reveals a collapsed fourth ventricle.⁷ Therefore, we think that an independent drainage system should be implanted in each compartment.

Finally, we favour an interhemispheric approach, which will have less probability of inducing mechanical pressure on a cranial nerve nucleus. Nevertheless, if a trans cerebellar approach

cannot be avoided, it is important to predetermine the length of the fourth ventricular catheter preoperatively, and to anticipate the decrease of size of the fourth ventricle in order to make sure that the catheter will never be long enough to press on the brainstem.

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Patient's perspective

(Written by the father of patient 2): We hope our son's medical history and complications will contribute to better management of this rare condition in the future.

Learning points

To avoid complications following isolated fourth ventricle drainage:

- The length of the fourth ventricle catheter should be measured preoperatively.
- A flow-regulated or high pressure programmable valve should be placed independently from the lateral VP shunt.
- A 'Y' connection between the lateral and fourth ventricle does not seem to prevent siphoning of the isolated fourth ventricle.
- An imagery-guided procedure should ideally be performed.

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