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Case Report

Hydrocephalus Revealed by Relapsing Bilateral Fourth Cranial Nerve Palsy

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ABSTRACT

Introduction: Uni- or bi-lateral fourth cranial nerve palsy due to hydrocephalus and/or after VPS placement is a very rare oculomotor manifestation. We report a case of relapsing bilateral fourth nerve palsies demonstrating recurring hydrocephalus. We reviewed the literature (table1) in order to inform the clinician about the clinical assessment, the past medical history and the radiological findings that prompt research for this peculiar entity and to avoid misdiagnoses like palsies of the sixth cranial nerve.

Diagnosis, intervention and outcome: The patient presented with recurrence of diplopia in reading position, partially resolved after a second VPS placement. A diagnosis of bilateral fourth nerves palsies was done after complete neuro-ophthalmological evaluation. A close follow-up demonstrated fluctuating level of diplopia by changing VPS valve resistance. An optimal placement of the VPS offered reduction and stability of diplopia. A final strabismus surgery was necessary to obtain complete symptoms release.

KEYWORDS

Hydrocephalus, Isolated fourth ventricle hydrocephalus, Fourth nerve palsy, Strabismus.

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Case Description

We report the case of a healthy 48-years-old woman presenting with a few-month history of headache, diplopia in reading position and nausea. Tetra ventricular hydrocephalus had been shown at MRI at early phase of the disease course and a prominent enlargement of the fourth ventricle had been highlighted. The first brain imagery shows a tetra ventricular hydrocephalus with a particularly enlarged fourth ventricle. As a strong flow artifact was observed through the aqueduct of Sylvius (Figure 1), a sub-occlusion of the foramina of Magendie and Luschka together with a lower grade stenosis of the aqueduct had been hypothesized. A ventriculoperitoneal shunting (VPS) of the right lateral ventricle with a Medtronic Adjustable Strata Regular® II valve with Delta siphon control device was subsequently performed in another institution with 2.0 opening pressure. Diplopia persisted and dysfunction of the system was diagnosed in our institution.

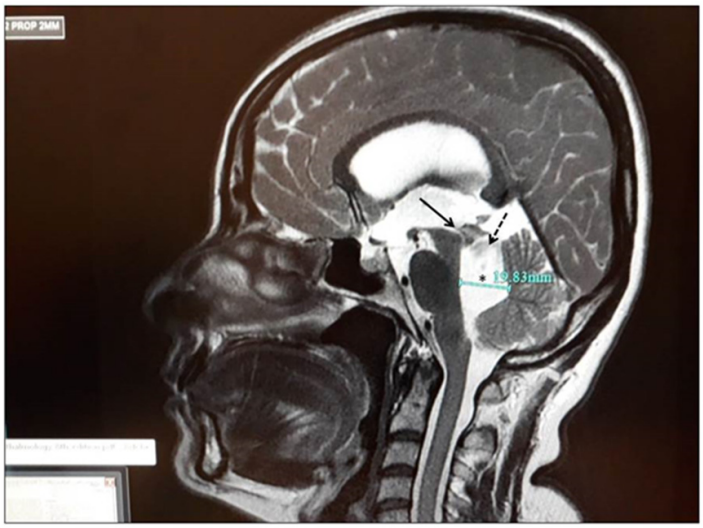

Figure 1: Mid-sagittal T2-weighted MR view showing elective over dilatation of the fourth ventricle together with enhanced flow artifact through the aqueduct of Sylvius (arrow) and at upper part of the ventricle (dotted arrow).

The shunt system was changed and we decided to set valve-opening pressure to 1.5 (standing opening pressure 8.5 cm H_2O). After a month period of symptoms relief, a recurrence of morning headaches with oblique diplopia occurred. An isolated bilateral fourth cranial nerve palsy was assessed considering the features of: i: subjective exyclotorsion over 10°; ii: V syndrome; iii: hypertropia increased in tilting positions, and iii: bilateral exyclotorsion at the fundus examination (Figure 2). Downgrading of the resistance of the Strata valve resulted in symptoms subsidence for only a few days. Though RMI control was unremarkable, an intermittent dysfunction of the ventriculoperitoneal shunting was strongly suspected considering relapses of similar episodes of oculomotor disturbance. VPS was therefore replaced (with a maintained opening pressure of 1.5) resulting in a one-year period of stabilization of the diplopia.

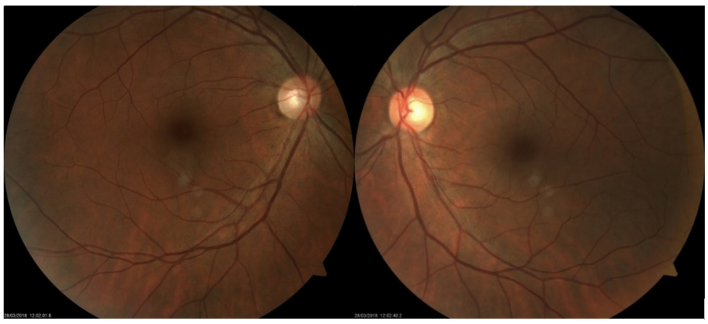

Figure 2: Funduscopy of our patient showing bilateral exyclotorsion.

Follow up and outcome

After replacement of the changed VPS in our institution, a close follow up over one year highlighted stable reduction of diplopia without complete resolution. Surgical correction of the strabismus

Authors of the case report	Description of the patient and onset of complains	Etiology of the hydrocephalus	Side affected	MRI findings	Treatment	Outcome
[1] Dimosthenis Mantopoulos, David G Hunter, Dean M.Cestari	29 yo man Vertical diplopia worse in down gaze 10 Y after shunting	Initial VPS placement for subarachnoid hemorrhage years before	Bilateral	Discovery of a dysfunctional VPS in good position with a broad enlarged third and fourth ventricle	New ventriculostomy of the third ventricle	Spontaneous resolving of diplopia
[2A] Pandey PK, et al.	20 yo woman vertical diplopia immediately after shunting	VPS placement for tubercular meningitis	Unilateral	Discovery of functioning VPS in good position. Good sized ventricles.	Strabismus surgery by 6 mm left inferior oblique recession	Diplopia disappeared
[2B] Pandey PK, et al.	16 yo boy vertical diplopia immediately after shunting	VPS placement for solitary pontine tuberculoma	Unilateral	Discovery of functioning VPS in good position. Good sized ventricles.	Strabismus surgery by 6 mm left inferior oblique recession	Diplopia disappeared
[3] Giesemann AM,et al.	82 yo man Complains of vertical diplopia after VPS	Non obstructive chronic hydrocephalus	Bilateral	Complication of the surgery with trauma of the pretectum	Surveillance	Diplopia persists
Our case	48 yo lady	Intermittent malfunction of the ventriculoperitoneal shunt was hypothesized	Bilateral	Discovery of a functional VPS in good position with a tetra ventricular hydrocephalus and particularly enlarged fourth ventricle	VPS replacement Strabismus and strata II valve resistance adaptation followed by surgery after 1 year	Diplopia fluctuated with Strata II valve resistance till stabilization And diplopia resolved after strabismus surgery

Table 1: Review in the literature of cases of isolated fourth nerve palsies as unique sign of hydrocephalus.

by a Harada-Ito procedure on the right eye had thereafter an excellent functional outcome with complete subsidence of the symptoms.

Discussion

Based on a thorough review of the literature, IV nerve palsies due to cerebro-spinal fluid (CSF) flow dysfunction has been scarcely reported [Table 1].

Hydrocephalus is defined as a disturbance of the flow of cerebrospinal fluid that leads to an increase in volume occupied by this fluid in CNS. So-called 'Communicating' hydrocephalus is featured by a disturbance of the CSF resorption in arachnoid villi leading to increased fluid volume within an unexpandable CNS without obstacle throughout the CSF pathways. In turn, the 'non-communicating' hydrocephalus either is featured by a detectable obstacle along CSF pathways within ventricles or occurs within the foramina impairing the outflow of ventricular CSF in the subarachnoid spaces. A peculiar sub-entity of the latter condition is the fourth ventricle hydrocephaly where there is the elective or preferential enlargement of the fourth ventricle. This entity of hydrocephalus has been reported as a complication of spontaneous or iatrogenic intraventricular hemorrhage, meningitis, or because of chronically excessive drainage by the VPS [4].

Most frequent ophthalmological signs of hydrocephalus are papilledema, enlarged blind spot at the perimetry, failure of upward gaze and unilateral or bilateral VI nerve palsy. In turn, as seen with our patient, fourth nerve palsy gives an oblique double vision increasing in down gazes and in the tilting positions (Bielschowsky test). The main differential diagnostic for these symptoms is the skew deviation. The exyclotorsion of the affected eye and the three steps method is usually conclusive in distinguishing the two entities (Figure 2).

Uni- or bi-lateral IV nerve palsy due to hydrocephalus and/or after VPS placement is a very rare oculomotor manifestation. The mechanism hypothesized in the literature [4] is either a stretching of the IV nerve resulting from an impingement of the superior medullary velum or a shearing of the midbrain due to the enlargement of the fourth ventricle. Since the cerebellar hemispheres are fixed by adhesions to the dura, the sole structure able to move in such mechanically constraining condition is the brainstem. In our patient all ventricles were enlarged since the initial phase of the disease course, but with a prominent enlargement of the fourth ventricle. The aperture pressure at lumbar puncture was normal which suggests significant obstruction of Magendie's and Luschka's foramina together with lower grade stenosis of the aqueduct of Sylvius. In the frame, we also noticed signs of CSF per-ependymal transudation in brain parenchyma surrounding the foramina. CSF analyses were normal. After thorough electronic review of the literature, only four cases of fourth nerve palsy in patients with hydrocephalus have been reported up to now [Table 1] who had all radiological/clinical features matching our hypothesis of preferentially elevated CSF pressure within the fourth ventricle. Furthermore, as CSF pressure fluctuates in hydrocephalus because of variable imbalance between CSF production and resorption, the diplopia fluctuated and the fluctuations closely matched changes in the resistance of the adjustable VPS valve. This observation strongly supported the hypothesis of CSF pressure-related changes in IV nerve dysfunction.

Conclusion

In conclusion, occurrence of diplopia enhancing in reading position or descending the stairs in a patient with history of hydrocephalus should suggest a uni- or bi-lateral IV nerve palsy, mainly in cases in whom overdilatation of the fourth ventricle when compared to other ventricles is assessed at CT or MR brain imaging. Risk factors such as previous VPS placement or meningitidis must be asked for. We advocate the proposal of a neuro-ophthalmology examination in all patients with hydrocephalus and diplopia to better define the oculomotor dysfunction, to establish the possible relationship between those entities and to improve the clinical follow up before or after VPS placement.

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