Normal Pressure Hydrocephalus Associated with Sleep Apnea: A Case Report and Review

Romaric Lantonkpodé; Cyrille Capel; Turki Alghassab; Pierre-Henri Launois; Olivier Balédent; Johann Peltier

1Department of Neurosurgery, Hospital University Center of Amiens-Picardie, 80054 Amiens, France.
2CHIMERE UR 7516, Jules Verne University, 80000, Amiens, France.
3Image Processing department, Hospital University Center, 80054, Amiens, France.

Introduction: Normal Pressure Hydrocephalus (NPH) is a pathology preferentially affecting an elderly population. Sleep apnea syndrome (SA) is a pathology that alters venous circulation by altering intrathoracic pressure during sleep. Phase-Contrast MRI (PCMRI) allows a quantitative analysis of craniospinal hemohydrodynamics and an analysis of the interactions between arterial blood, venous blood volume exchanges and cerebrospinal fluid (CSF) oscillations. We present the case of a patient with NPH-like symptomatology associated with AS.

Case report: We report a 68-year-old patient with Hakim’s triad associated with hydrocephalus with stenosis of the mesencephalic aqueduct. Associated SA was found. Treatment of SA resulted in substantial clinical improvement for one year in Hakim’s triad. Thereafter, he showed a new clinical alteration requiring endoscopic ventriculocisternotomy, which allowed a perennial improvement. The analysis of craniospinal hemohydrodynamics by PCMRI confirmed the absence of CSF pulsatility at the level of the midbrain aqueduct as well as a hyperpulsatility of the cervical CSF. We also found an alteration of the venous dynamics with an increase in the participation of the accessory drainage routes in the cerebral venous drainage. After treatment, we noted a decrease in the pulsatility of the cervical CSF.

Conclusion: The treatment of sleep apnea syndrome shows an effect on craniospinal hemohydrodynamics which translates into a clinical improvement. This shows the importance of considering these parameters in the management of patients with chronic hydrocephalus.

Keywords: Hydrocephalus; Sleep apnea; Cerebrospinal fluid; Phase-Contrast MRI.

Phase Contrast MRI (PCMRI) is the only tool that allows analysis of craniospinal hemodynamics and hydrodynamics in a non-invasive manner. During a cardiac cycle and in the physiological state, there is an inflow of arterial blood that is not immediately compensated by a flush of venous blood. There is therefore a variation in intracranial vascular volume. Intracranial pressure is preserved by a flushing of CSF from the cisternal subarachnoid spaces to the perimedullary subarachnoid spaces[10].

We report the case of a patient with atypical signs of Hakim’s triad long associated with NPH whose quality of life improved after fitting a device for his AS. We performed craniospinal hemodynamic analysis by PCMRI at each stage of management.

Case report

A 68-year-old patient was referred to neurosurgery by his general practitioner for gait disorders. The symptomatology had been evolving for one year and associated walking and balance disorders, daily urinary leakage and cognitive disorders. His history included multiple cardiovascular risk factors, including hypertension, diabetes, and dyslipidemia. His Body Mass Index (BMI) was 28.

Gait disorders were not associated with a decrease in step length, the step height was altered. No instability was found. Sphincter disturbances were present, rather in the form of polakiuria. The urodynamic assessment showed a bladder hyperreactivity possibly of neurological origin. The patient underwent a complete cognitive evaluation which revealed an MMSE score of 22/29 with an attentional deficit associated with a marked deficit in episodic memory and some praxis difficulties.

MRI revealed a tri-ventricular dilatation (Evans index: 0.4) with a stenosis in the aqueduct of Sylvius (Figure 1). There was no effacement of the vertex sulci. The sylvian valleys were of normal morphology. The angle of the corpus callosum was 90°. There was no focal dilatation of the convex sulci. The DESH score was 4/10. There were periventricular hyper signals suggestive of vascular leukopathy (Fazekas 3) (Figure 2).

There was no papilledema. He was diagnosed with glaucoma.

A polysomnography was performed in a context confirming a severe sleep apnea syndrome (SA), apnea and hypopnea index (AHI) 28/h micro-arousal with an index of micro-arousals at 17.8/h. This sleep apnea and hypopnea syndrome was obstructive and positional with an AHI on his back at 42.6. He was therefore fitted with a device with good compliance. He then showed an improvement on all the parameters of the Hakim triad. Sphincter disorders disappeared. From a cognitive point of view, the MMSE showed an improvement to 26/29. The walking disorders disappeared. The family circle and the patient mention an overall improvement of the patient’s quality of life.

The follow-up in neurosurgery consultation was regular for one year with a secondary deterioration of his clinical condition, especially on walking, despite a good compliance for his SA. It was therefore indicated a ventriculocisternostomy by endoscopic way. This allowed an improvement both on the instability to walk but also on the cognitive disorders with persistence however of his urinary incontinence.

The study of craniospinal hemodynamics and hydrodynamics showed, before any treatment, a hyperpulsatility of the CSF measured at the level of the high cervical subarachnoid spaces.

This type of hyperpulsatility has been found in idiopathic intracranial hypertension [11]. This hyperpulsatility was associated with hemodynamic disorders and in particular a distribution of the drainage carried out at 53% by the jugular veins and at 47% by accessory drainage routes. One year after carrying out a treatment by ventilation in positive airway pressure, a reduction of nearly 54% of the pulsatility of the high cervical CSF was observed. The distribution of the drainage was essentially jugular. These parameters remained relatively stable before and after endoscopic ventriculocisternostomy. Only the distribution of drainage showed a change after treatment by showing a further increase in the distribution of venous drainage. After ventriculocisternostomy, there was a significant pulsatility of the CSF in the somy. This shows the importance of the intraventricular compliance which was incompetent because of the stenosis of the aqueduct. All analysis of PCMRI are resumed on Table 1.
In our case, the existence of venous drainage resistance could potentiate this venous hypertension and contribute to CSF stasis. Since in addition to his hydrocephalus by stenosis of the aqueduct, and the AS, there is an absence of the right lateral sinus.

Other hypotheses have been put forward over time. The one involving the Glymphatic System (GL) is more and more evoked. It is a conception of the balance between secretion and resorption of CSF in the different brain spaces as well as the clearance of protein waste. It has been reported that the GL is particularly active during sleep. Current theories suggest the relationship between CSF, GL, and sleep. Chong [14] in a systematic review reports that elements of LCS circulation/resorption. GL exchange represent pathways influenced by sleep. Lee [20], using diffusion tensor MRI, assessed the function of the GL along the perivascular spaces in two groups of patients (with and without SA). They were able to demonstrate a dysfunction of the GL in patients with SA. Moreover, this dysfunction would be correlated with the severity of SA. The intricacy of the different hypotheses suggests that polysomnography should be performed in the evaluation of patients suspected of having NPH. The role of the glymphatic pathway in several neurological diseases is an important research topic. The system is particularly related to neurodegenerative disorders, such as Alzheimer’s disease, normal pressure hydrocephalus, glaucoma... [1].

Our patient with both multifactorial NPH and AS, associated with glaucoma, is of course in this register. It would be difficult to prejudice the direction of interaction between his two pathologies in the patient.

Mc Loughlin [21] reported a similar case of severe AS associated with NPH with aqueductal stenosis. On a brain scan performed five years earlier in the same patient, no aqueduct stenosis was visualized nor was hydrocephalus. They concluded that their patient had developed an aqueduct stenosis by local gliosis due to microvascular ischemia. The response to this ischemic lesion involving hypertrophy or proliferation of glial cells in the periventricular and aqueduct region.

In our case, we do not have previous brain imaging before the patient’s first consultation for his present symptoms. We support the hypothesis that the pre-existing NPH in our patient...
due to partial aqueductal stenosis may have been compensated by other compensatory mechanisms. This justifies the important flushing of CSF found from the cerebral subarachnoid spaces to the cervical peri-medullary subarachnoid spaces. The additional presence of SA becomes the decompensating factor of this already established balance. This would justify the clinical improvement of the patient after he has been fitted with a device.

The treatment of hydrocephalus by aqueduct stenosis calls for a first restoration of the CSF circulation by endoscopic ventriculocisternostomy. It is increasingly preferred for this indication over internal ventricular shunt (atrial or peritoneal) for its low safety and reduced risk of complications [22,23]. In this case, the decision to shunt was only secondary in view of the atypical clinical presentation.

Conclusion

NPH and SA are two frequently associated pathologies of adult life. Their interrelated pathophysiological mechanisms suggest the presence of similar risk factors. Atypical clinical presentation of HPN associated with the presence of cardiovascular risk factors should prompt a search for AS.

References

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