Hong Kong College of Physicians
Case Report for Interim Assessment
Specialty Board of Advanced Internal Medicine (AIM)

For AIM Training, case reports should be submitted in the prescribed format together with the application form for Interim Assessment at least EIGHT Weeks before the date of Interim Assessment

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Date(s) and place (hospital) of patient encounter: 14/8/2023 (NDH MED)
Date of report submission: 13/9/2023

Case report

Note: Failure to follow the prescribed format (including the number of words) results in a FAILURE mark (score between 0 and 4) for the Case Report.

Title: A Case of Idiopathic Normal Pressure Hydrocephalus with the Classical Triad of Symptoms

Case history:

Mr. A is a 61-year-old gentleman with a history of anxiety disorder, bipolar affective disorder, benign prostate hyperplasia with lower urinary tract symptoms, and hypertension.

He attended the emergency department twice in February 2023 for a 6-month history of non-specific dizziness, which was non-vertigo and unrelated to postural change. He mainly experienced dizziness when he walked as he felt unsteady. Physical examination showed normal cranial nerves, full limb power, intact sensations, and the absence of past-pointing or dysdiadochokinesia. There was mild unsteadiness on tandem walking. There were no signs of Parkinsonism. Blood tests, including complete blood count, liver function test, renal function test, active vitamin B12 level, folate, thyroid function test, and Venereal Disease Research Laboratory Test (VDRL), were all unremarkable. Plain computed tomography (CT) of the brain showed dilatation of bilateral lateral ventricles and the third ventricle. It also showed disproportionate subarachnoid space enlargement, mainly affecting the Sylvian fissures.

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A short course of rehabilitation was done. His Modified Barthel (MBI) Index Score was 100 out of 100, and his Modified Functional Ambulatory Category (MFAC) was 6 out 7 after rehabilitation. He was able to walk without walking aids and was discharged.

He was followed up in the medical outpatient clinic two months later. The attending consultant reviewed a detailed clinical history. He further revealed a history of progressive worsening of memory and progressive unsteady gait in the past two years. He also had three episodes of fall and urinary incontinence in the past year. He was then admitted to the medical ward for further workup for suspected normal pressure hydrocephalus (NPH).
The neurology team was consulted, and early Magnetic Resonance Imaging (MRI) of the brain with contrast and neurosurgical referral was suggested. Lumbar puncture was attempted, but it was unsuccessful. Cognitive assessment with the Montreal Cognitive Assessment Hong Kong version (HK-MoCA) score was 28/30 (2 marks were deducted for failure to recall). He was assessed by physiotherapists and had a MFAC category 5 out of 7 and was prescribed a rollator as a walking aid. He was discharged and was referred to the neurosurgery clinic for further assessment.

MRI Brain with contrast was done in May 2023. It showed a serial increase in the degree of ventriculomegaly involving the lateral, third, and fourth ventricles. The Evans index was 0.32 (reference range: 0.20-0.25: Normal; 0.25-0.30: Possible or early ventriculomegaly; >0.3: Ventriculomegaly). It also showed a widening of temporal horns, a narrowing of callosal angle to 78 (lower limit: 90, measured at posterior commissure on coronal view), and an upward bowing of corpus callosum with mildly reduced thickness. There was also a disproportionate dilatation of bilateral sylvian fissures with relative effacement of cerebral sulci at the vertex. The cerebral aqueduct appeared dilated with a prominent flow void. The mean cycle volume was 0.085 ml (upper limit: 0.045 ml - 0.072 ml), which suggested hyperdynamic CSF flow. Overall, MRI features were suggestive of normal pressure hydrocephalus.

He was re-admitted two months later for unsteady gait and dizziness. Lumbar puncture was reattempted successfully. The opening pressure was 22.6cm H2O. Clear cerebrospinal fluid (CSF) was obtained, and CSF

analysis showed normal biochemical and microbiological results. 25ml of CSF was drained during the procedure. His mobility improved to MFAC category 6 with the aid of a walking stick after the lumbar puncture, and he was discharged.

However, two weeks later, he was re-admitted for unsteady gait. He was transferred to the neurosurgical ward for further management. A Lumbar Infusion test was performed. It showed a normal opening pressure with a moderate CSF outflow resistance of 19mmHg/ml/min and a rapid drop in CSF pressure after free drainage was allowed. A gait assessment was performed before and after the infusion test, and he showed improvement and became self-ambulatory again.
The Lumbar Infusion test results indicated a moderate impairment of CSF re-absorption; therefore, he was likely to respond favorably to shunting. He agreed to right ventriculoperitoneal shunting with a Certas Plus programmable valve. The operation was performed on 6/9/2023, and he was still in rehabilitation at the date of this report.

Discussion and literature review

This case illustrated a patient with the classical triad of symptoms of idiopathic normal pressure hydrocephalus (iNPH).

Proposed Pathophysiology

The choroid plexus produces most cerebrospinal fluid (CSF) in the lateral ventricles. It then flows into the third ventricle, the fourth ventricle, and the subarachnoid space. The arachnoid villi eventually absorb them back into the systemic circulation via the sagittal sinus.

NPH can be classified as idiopathic or secondary. Common causes of secondary NPH include meningitis, intraventricular hemorrhage, and subarachnoid hemorrhage. When there are no apparent causes, it is classified as idiopathic NPH.
The exact pathophysiology of iNPH is unclear. It has been hypothesized that a decrease in CSF absorption causes an increase in intracranial pressure. The ventricles dilate over time and lead to normalization of intracranial pressure. The CSF flow is directed into the brain parenchyma via the Virchow-Robin spaces. It causes parenchymal changes, eventually leading to tissue compression and deep white matter ischemia. Some studies have shown an increase in cerebral blood flow could lead to clinical

improvement (1).

Clinical features

There is no age predilection for secondary NPH. Idiopathic NPH is more common in patients over 60 years old, and the prevalence increases with age. Both males and females are equally affected.

The three cardinal features of normal pressure hydrocephalus include 1) gait abnormality, 2) cognitive disturbance, and 3) urinary incontinence, but patients may not have all three symptoms.

1. Gait abnormality is the most important and prominent clinical feature in NPH. The characteristics of the gait abnormality include small-stepping, wide-base, reduced velocity and stride, diminished height of the steps, and increase in sway (2). It is often described as a "glue-footed" or magnetic gait. Patients tend to fall when turning.
2. Cognitive impairment in NPH has an insidious onset, which takes months to years to develop. Features include attention deficit, impaired executive function, poor memory, impaired visuo-perceptual and visuospatial functions, psychomotor slowing, and apathy (3).
3. Urinary symptoms usually begin with urinary frequency. In the early stage, incontinence may be related to impaired mobility and difficulty reaching the toilet. In the late stage, cognitive impairment may play a role in urinary incontinence, as patients usually lack concern for voiding.

This case illustrated the importance of careful history-taking. Although the literature has developed many terms to describe gait abnormality in NPH, patients may perceive their gait abnormality as other symptoms. Our patient described his symptoms as "dizziness" twice. Only upon further history taking was it revealed that the "dizziness" he described was referring to unsteadiness in walking. Also, he did not initially provide a history of mild memory decline and urinary symptoms until he was directly asked about these symptoms during the follow-up.

Diagnostic Evaluation and Treatment

Evaluation of patients with suspected normal pressure hydrocephalus includes 1) exclusion of NPH mimics, 2) imaging to support the diagnosis, and 3) provocative testing.

1. Exclusion of mimics

Idiopathic NPH is a diagnosis of exclusion. Several neurodegenerative diseases mimic NPH and should be considered as an alternative diagnosis. Patients with Dementia with Lewy bodies (DLB), Progressive Supranuclear Palsy, and Parkinson's disease may have gait abnormality at initial presentation mimicking NPH. However, these disease entities have their unique clinical features that allow one to distinguish them from NPH. For instance, patients with DLB usually have prominent psychotic features, and patients with Parkinson's disease typically have characteristic motor symptoms like bradykinesia, tremor, and rigidity. Alzheimer's disease and vascular dementia are important differential diagnoses for patients with cognitive impairment. Gait disturbance is a hallmark symptom of NPH, and cognitive impairment without prominent gait abnormalities is highly suggestive of an alternative diagnosis.

2. Imaging

CT and MRI of the brain are important imaging modalities to support the diagnosis of NPH. Ventriculomegaly is an important but non-specific feature. Evans index is used to quantify vestibulopathy. An Evans index >0.3 is suggestive of dilated ventricles. However, the Evans index is increased in nearly 20% of elderly above 70 years old. Disproportionate enlargement of the subarachnoid space hydrocephalus (DESH) is a more specific radiological feature of NPH. The idiopathic NPH Radscale Score has been developed to standardize the assessment of radiological features of NPH. A score over 8 suggests iNPH as the likely diagnosis (4).

3. Provocative testing and treatment

Surgical CSF shunting is the main treatment modality for NPH. In carefully selected patients, 80-90% of them respond to CSF shunt surgery (17). A systematic review including 64 studies and 3063 patients found that 82% of patients undergoing shunt surgery showed clinical improvement, and the mortality rate was 0.2% (5).

Ventriculoperitoneal shunting is the most performed in which CSF is redirected from the lateral ventricle into the peritoneum via a catheter. Ventriculoartrial shunting is less common, involving CSF shunting into the heart's atrium. Lumb-peritoneal shunting has been attempted, but further studies are needed to validate its efficacy.

Given the lack of a gold standard for the diagnosis of NPH and the invasiveness of the surgery, proper patient selection for shunt surgery is essential. Therefore, several provocative tests have been developed to identify patients most likely to respond to a CSF shunt surgery. These tests include the CSF tap test, external lumbar drain test, and lumbar infusion test. (6)

CSF tap test involves the removal of 40ml of CSF by lumbar puncture. Gait and cognitive assessments are performed before and after the procedure to look for any clinical improvement. (6)

External lumbar drain test involves the placement of a temporary catheter into the lumbar space. CSF is drained at a rate of 5-10ml/hour for 2-7 days (6). Patients are observed for any clinical improvement during the trial period.

Lumbar infusion test assesses CSF re-absorption. It involves the placement of two lumbar drains at different lumbar levels. A fluid is slowly infused into the lumbar CSF space via one of the drains. The other drain is used for monitoring the outflow resistance. Patients with an increased outflow resistance are likely to be shunt-responsive. Our patient's outflow resistance was 19 mmHg/ml/min. Boon et al. found that patients with CSF outflow resistance of 18 mmHg/ml/min or greater were more likely to respond to shunting. (7)

CSF tap test has a high positive predictive value (94%) but a low negative predictive value (58%) (8). External lumbar drain test and lumbar infusion test have a higher specificity (80%) than sensitivity (50%) (6). Therefore, while a positive provocative result suggests shunt responsiveness, a lack of response on these tests does not exclude a patient from this surgical procedure.

A multidisciplinary approach involving neurologists, neurosurgeons, radiologists, and allied health teams is required, and the potential risks and benefits of a shunt surgery should be discussed with the patients (6).

Summary

In conclusion, this case illustrated the typical presentation of idiopathic normal pressure hydrocephalus with gait disturbance, mild cognitive impairment, and urinary incontinence. The patient showed clinical improvement with the CSF tap and lumbar infusion tests and was a good surgical candidate.

Tables and figures (where applicable) (no more than two figures)

Nil

Passed at Council Meeting on 24 Oct 2019

Reference (not more than 10)

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10. Williams MA, Malm J. Diagnosis and Treatment of Idiopathic Normal Pressure Hydrocephalus. Continuum (Minneap Minn). 2016;22(2 Dementia):579-599. doi:10.1212/CON.0000000000000305

No of words in Case History and Discussion (excluding references):__1818_____.

(should be between 1000-2000)

Declaration

I hereby declare that the case report submitted represents my own work and adheres to the prescribed format. I have been in clinical contact with the case selected. The case report has not been submitted to any assessment board or publication and it is NOT related to my second specialty(ies), if any. My consent is hereby given to the College to keep a copy of my case report, in written and/or electronic, at the College Secretariat and allow the public to have free access to the work for reference.

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(signature of Trainee)

Endorsed by Supervisor *

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