Case Report

Normal Pressure Hydrocephalous: a Treatable Cause of Dementia

Sudeepta Dandapat*, Kuljeet Singh Anand*, Bhupender Kumar Bajaj*, Bharat Bhushan Sharma**

Abstract

Normal pressure hydrocephalous (NPH) usually occurs in people older than 60 years of age. It generally presents with gait disturbances, urinary incontinence and cognitive decline. The cognitive symptoms may mimic Alzheimer disease while the gait disturbance may mimic festinating gait of Parkinson disease. NPH is often misdiagnosed as one of these diseases. Since NPH can be reversed with shunt surgery, it must be correctly diagnosed.

Key words: Hydrocephalus; normal pressure; dementia; gait apraxia; urinary incontinence

Introduction

The Normal Pressure Hydrocephalous (NPH) first described by Hakim et al [1] and then by Adams et al [2] in 1965, is hydrocephalous in the absence of papilloedema and with normal cerebrospinal fluid (CSF) opening pressure on lumbar puncture. A term such as chronic hydrocephalous has also been used. NPH may exhibit the classic triad (also known as Adam's triad) of urinary incontinence, gait disturbance and dementia. Although NPH is a relatively rare cause of dementia, identifying NPH is important because it is one of the few treatable entities. A case of NPH is reported who improved dramatically after ventriculo-peritoneal shunt surgery.

Report of case

A 70-year nondiabetic, normotensive housewife was referred with complaints of progressive difficulty in ambulation since nine months and deteriorating intellect since six months. Detailed history revealed that she had also been experiencing urinary incontinence in recent months. The patient denied headaches. The difficulty in ambulation was in the form of small-stepped gait with a feeling as if the feet are glued to the floor. Three months after the gait disturbances she had problems with short-term memory. There were no praxic, gnostic or lexic disturbances. There was no history of transient ischaemic attacks (TIA) or stroke in the past. General physical and systemic examinations were unremarkable. Neurological evaluation revealed Mini-mental state examination (MMSE) score 22/30, recall and calculation were impaired; also her reaction time was longer. Her frontal lobe function tests were normal. Cranial nerves, motor and sensory system examination were normal. Deep tendon reflexes were normal with bilateral plantar flexor responses. Gait was slow with short steps and feet were glued to the ground. Haematological and biochemical investigations were within normal limits. Magnetic resonance imaging of brain showed symmetric dilatation of lateral ventricles and third ventricle with evidence of subependymal seepage of CSF (interstitial edema) (figures 1 & 2). CSF flow dynamic study revealed hyperdynamic flow within third ventricle and increased flow through aqueduct in both systolic and diastolic phases. A diagnosis of normal pressure hydrocephalous was made and a CSF tap drainage test was performed in which 50 mL of CSF was drained. The opening pressure was 180 cm of water and cytobiochemical analysis showed 4 lymhocytes/HPF, protein-21 mg/dL and sugar-56 mg/dL (corresponding blood sugar 110 mg/dL). Pre and post lumbar-puncture evaluation of gait and cognitive functions were carried out after first hour and repeated three hourly till 12 hours. There was marked improvement of gait and MMSE score; in recall and calculation. A low pressure Chabbra ventriculo-peritoneal shunt surgery was performed following which there was significant recovery in gait and cognitive functions. On follow-up after

Department of *Neurology & **Radiology, PGIMER and Dr. R.M.L.Hospital, New Delhi – 110001.

Corresponding Author: Dr. Kuljeet Singh Anand, Department of Neurology, PGIMER and Dr. R.M.L.Hospital, New Delhi – 110001. Email : kuljeet_anand@rediffmail.com

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three months of shunt surgery, she continued to maintain improvement in gait and cognitive status, with normally functioning shunt without any complications.

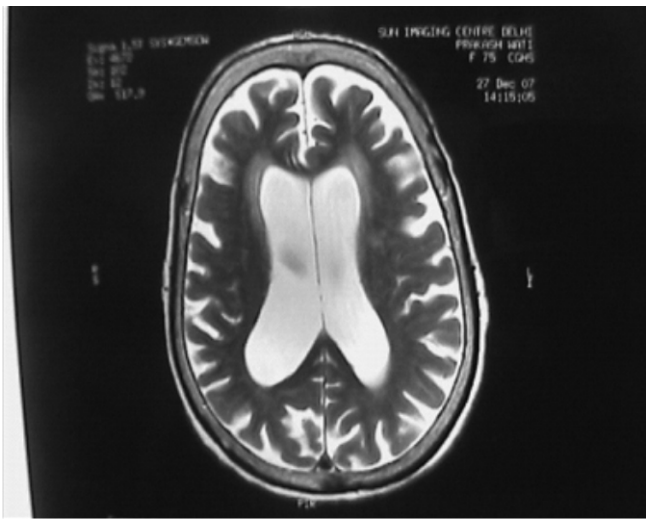

Figure 1- Dilated lateral ventricles with periventricular T2 hyperintensity

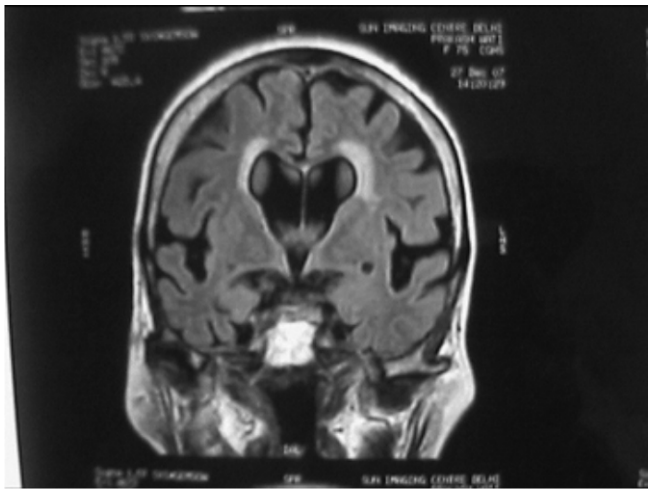

Figure 2- Dilated lateral and third ventricles with periventricular T2 hyperintensities

Discussion

Patients with NPH may have a known or probable cause in form of trauma, meningitis, subarachnoid haemorrhage, surgery or tumours. The resulting syndrome is commonly referred as secondary NPH. In more than half of NPH patients, however, there is no identifiable cause [1,2]. These are idiopathic cases. Idiopathic NPH appears to involve the interplay of several factors such as decreased ventricular compliance, decreased cerebral blood flow to periventricular regions and decreased absorptive capacity of brain to CSF.

NPH is fundamentally a clinical diagnosis. It is often diagnosed with a good history and physical examination, together with a CT or MRI and a lumbar puncture for CSF pressure determination.
Diagnostic modalities to predict shunt responsiveness include positive lumbar puncture/ lumbar drain trial test and presence of pressure signs on MRI such as periventricular transependymal effusion. CSF flow dynamics characteristics across the aqueduct of Sylvius can be studied using phase-contrast MRI. Bradley et al [3] found that increased flow void in the aqueduct as seen on proton density-weighted conventional spin-echo images was associated with favourable shunt outcome. Intracranial pressure (ICP) monitoring [4] has been described to have some use in NPH. The presence of B waves, which are transient elevations of mean and pulse pressure, for more than half the monitoring time suggest a better outcome with shunting. A radionuclide cisternogram [5,6,7] may provide useful objective data of altered CSF dynamics, but it has been found to be a poor predictor of response to shunting because it is non-specific. Positron-emission tomography (PET) and single-photon emission computerised tomography (SPECT) also have predictive value [8].

The decision to treat is often straightforward. The mainstay of treatment is ventriculoperitoneal shunt employing preset non-programmable device and the outcome is usually quite satisfactory. Technical difficulties include the selection of correct valve pressure to achieve maximum benefit with minimum risk. The most significant risk is that of collection of extra-axial fluid collections of CSF or blood due to over-shunting. Programmable shunts have also been developed and are in use. Other procedures of diverting CSF from ventricles is endoscopic third vetriculostomy.

Idiopathic NPH can be difficult to treat. Those patients who are most likely to benefit have a significant gait disturbance and a limited degree of dementia [9]. Use of lumbar puncture to assess improvement and pressure prior to shunting has been reported [10]. Factors that do not appear to affect the outcome are patient's age, duration of symptoms and extent of ventricular dilatation. Overall as many as 50% of NPH patients, who undergo shunting, experience improvement [4,6,9]. In terms of symptoms, improvement is commonly seen in gait and urinary incontinence, but not as consistently in cognitive impairment, particularly in short term memory.

Key Points

- The symptomatology of NPH in an elderly patient can be easily overlooked as it may appear to be part of aging.
- Many patients of NPH do not come to medical attention and remain undiagnosed.
- It is important to diagnose NPH since it is a potentially treatable cause of dementia and timely neurosurgical intervention improves quality of life.

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References

1. Hakim S, Adams RD. The special clinical problem of symptomatic hydrocephalus with normal cerebrospinal fluid pressure. J Neurosci 1965;2:307-27.
2. Adams RD, Fisher CM, Hakim S, Ojemann RG, Sweet WH. Symptomatic occult hydrocephalus with "normal" cerebrospinal pressures: a treatable syndrome. N Engl J Med 1965;273:117-26.
3. Bradley WG, Scalzo D, Queralt J, Nitz WN, Atkinson DJ, Wong P. Normal-pressure hydrocephalus: evaluation with cerebrospinal fluid flow measurements at MR imaging. Radiology 1996;198:523-9.
4. Symon L, Dorsch NWC. Use of long-term intracranial pressure measurement to assess hydrocephalic patients prior to shunt surgery. J Neurosurg 1975;42:258-73.
5. Vessal K, Sperber EE, James AE Jr. Chronic communicating hydrocephalus with normal CSF pressures: a cisternographic-pathologic correlation. Ann Radiol (Paris) 1974;17:785-93.
6. Chang CC, Kuwana N, Ito S, Ikegami T. Prediction of effectiveness of shunting in patients with normal pressure hydrocephalus by cerebral blood flow measurement and computed tomography cisternography. Neurol Med Chir (Tokyo) 1999;39:841-6.
7. Bergstrand G, Oxenstierna G, Flyckt L. Radionuclide cisternography and computed tomography in 30 healthy volunteers. Neuroradiology 1986; 28:154-60.
8. Granado JM, Diaz F, Alday R. Evaluation of brain spect in the diagnosis and prognosis of the normal pressure hydrocephalus syndrome. Acta Neurochir 1991; 112: 88-91.
9. Poca MA, Mataró M, Matarín M, Arikan F, Junqué C, Sahuquillo J. Good outcome in patients with normal-pressure hydrocephalus and factors indicating poor prognosis. J Neurosurg 2005; 103:455-63.
10. Bradley WG. Normal Pressure Hydrocephalus: New Concepts on Etiology and Diagnosis. AJNR Am J Neurorad 2000; 21:1586-90.