

Normal-Pressure Hydrocephalus with Misleading Features of Irreversible Dementias: A Case Report

Michael Schwarzschild, MD, PhD, Guy Rordorf, MD, Kaaren Bekken, PhD, Ferdinando Buonanno, MD, and Jeremy D. Schmahmann, MD

ABSTRACT

An 85-year-old man presented with the clinical triad (gait instability, dementia, and bladder and bowel incontinence), the ventriculomegaly, and the normal CSF pressure that characterize normal-pressure hydrocephalus (NPH). Diagnostic uncertainty was raised by an unusually rapid onset and a lack of initial response to CSF tap tests. Additionally, periodic sharp waves on EEG suggested the possibility of Creutzfeldt-Jakob disease, and positron emission tomography (PET) demonstrated a pattern of cerebral hypometabolism typical of Alzheimer's disease. Nevertheless, the diagnosis of NPH was supported by delayed improvement following CSF tap tests, and it was confirmed by a dramatic clinical recovery after CSF shunting, resolution of the EEG and PET abnormalities, and a normal brain biopsy. NPH remains one of the few reversible causes of dementia, and the presence of its core features, regardless of rate of onset or ancillary test results, warrants careful consideration of therapeutic intervention. (*J Geriatr Psychiatry Neurol* 1997; 10:51-54).

The cardinal controversy surrounding idiopathic normal-pressure hydrocephalus (NPH) since its initial description 30 years ago,¹ has been how to determine which patients should undergo CSF shunting. Numerous ancillary studies [including the CSF tap test, cisternography, EEG, and positron emission tomography (PET)] have been investigated for their ability to predict shunt responsiveness, but their value has remained unproven.² We present a case of NPH that highlights the importance of the central clinicroadiographic features of NPH, and also illustrates the potential pitfalls of ancillary studies. In particular, the case demonstrates EEG and PET findings, not generally associated with NPH, that hindered its diagnosis and management.

CASE REPORT

This witty and vigorous 85-year-old semiretired mailman presented with rapidly progressive cognitive decline, gait disturbance, and incontinence. He was in excellent

health until January 5, 1994, when, after driving home, he was found confused on his front porch not knowing if he was at the right house. Three days later, he was found lost in his basement trying to find the upstairs bedroom. In addition to the initial spatial disorientation, he soon developed memory deficits, abulia, and poor judgment. As his condition progressed over the next 2 months, he became totally dependent on others for his care.

Unsteady walking was also noted during the first week of January, prompting his use of a cane. Within a week, he displayed hesitation and slowness in standing, and "got stuck" when walking. In February, the cane was replaced by a wheelchair, and by March, he was largely bed-bound.

Mild urinary incontinence developed at the end of January within 4 weeks of the onset of the patient's cognitive and gait difficulties. By mid-February, he had become completely incontinent of both urine and stool.

He had no significant past medical history, and specifically no history of antecedent viral syndrome, head trauma, meningitis, alcohol abuse, depression, or psychoactive medication. His family history was remarkable only for Alzheimer's disease in one of his six siblings.

In mid-March, a head CT showed marked ventriculomegaly with only mild sulcal enlargement (Fig. 1A), and he was admitted to our hospital with a concern for NPH. On examination, he appeared relatively youthful, but profoundly abulic sitting on an absorbent underpad in

Received August 8, 1996. Accepted for publication November 26, 1996.

From the Department of Neurology, Massachusetts General Hospital and Harvard Medical School, Boston, Massachusetts.

Reprint requests: Dr. Jeremy D. Schmahmann, Department of Neurology, Burnham 823, Massachusetts General Hospital, Boston, MA 02114.

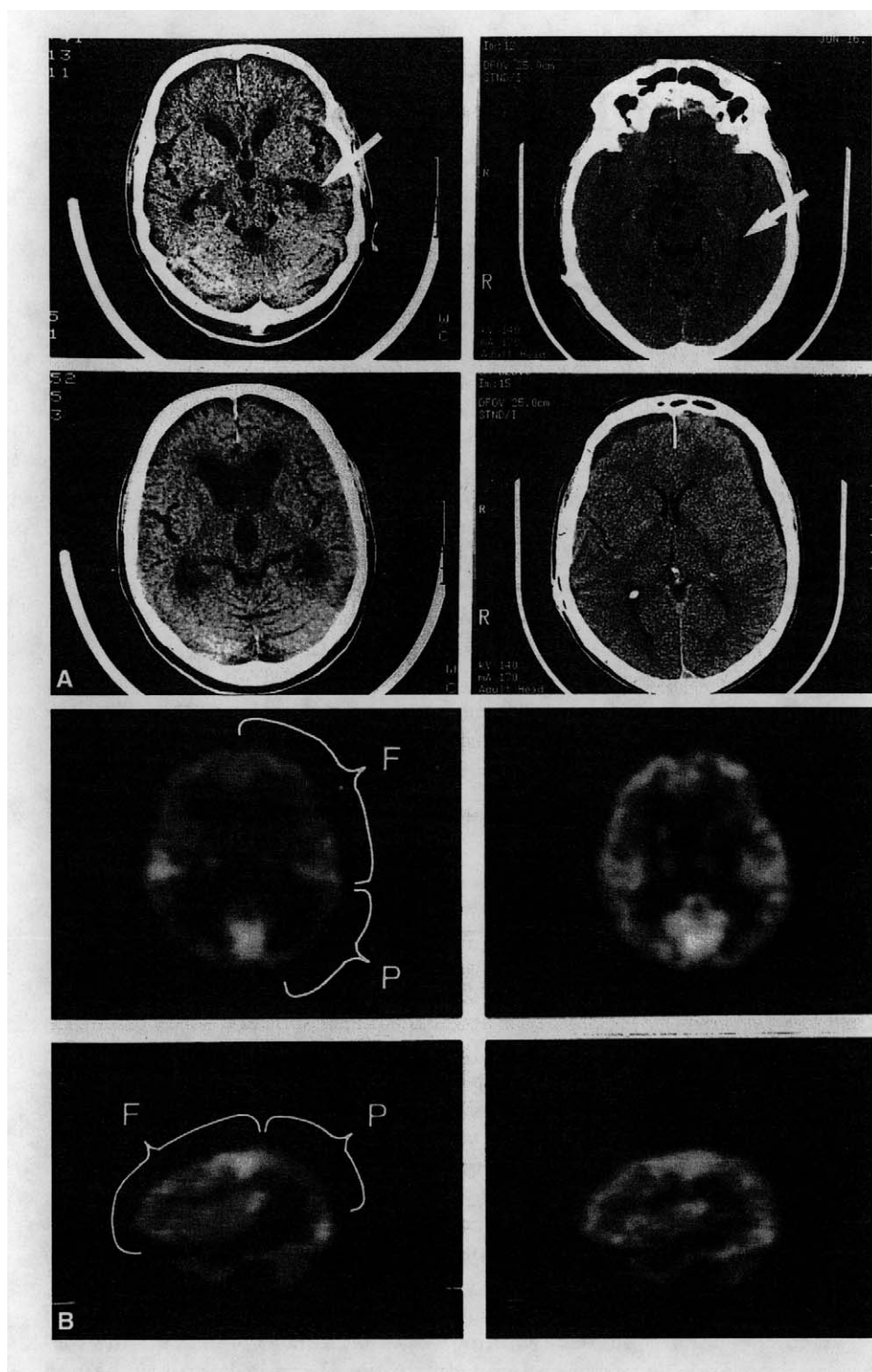


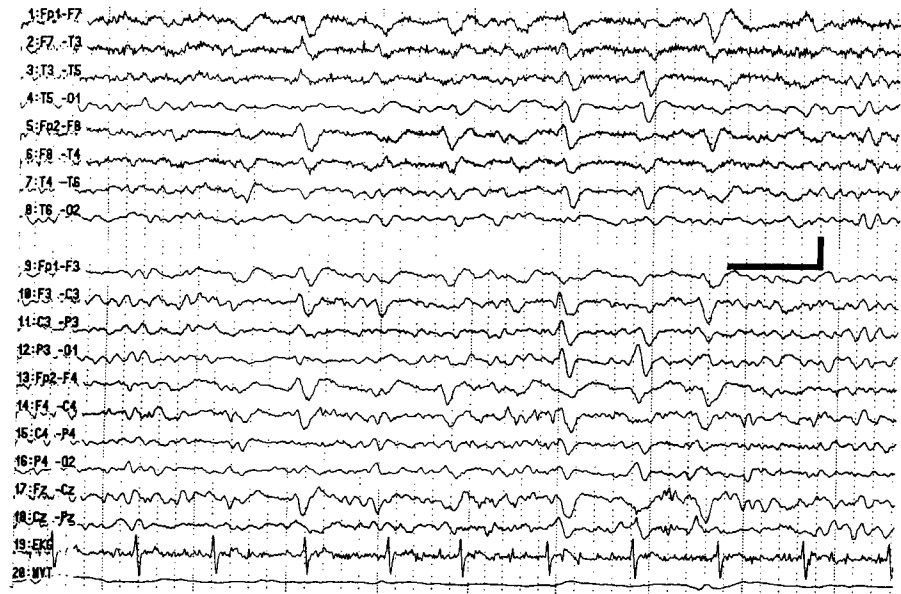
Figure 1. Anatomic and functional neuroimaging before and after ventriculo-peritoneal shunting. (A) Noncontrast axial head CT images at the level of the interpeduncular fossa (upper images) and the level of the frontal horns of the lateral ventricles (lower images) at presentation (left) and 3 weeks after shunting (right). The ventricular spans between the caudate nuclei at the level of the frontal horns measure 34 mm before shunting and 9 mm after shunting. Lower axial images not shown here demonstrated dilation of the 4th as well as the 3rd and lateral ventricles on the initial CT, with diminution after shunting. (B) Axial (upper) and parasagittal (lower) FDG-PET scans of the brain 1 week before (left) and 2 months after (right) ventriculo-peritoneal shunting. The focal hypointensity seen initially in the anterior frontal lobes (F) and posterior parietal lobes (P) was no longer apparent after shunting. In the axial images, the ratio of FDG uptake (measured in mCi/mL) between the parietal cortex and the peri-Rolandic cortex increased 36% after shunting. Similarly, in the parasagittal images, the ratio between parietal cortex and cerebellum increased 20% after shunting.

a wheelchair. He did not speak spontaneously and took 20 seconds to answer a simple question. He was disoriented to time and place; he knew his name, but not those of his children. He repeated well but did not remember words for more than 10 seconds. Long-term memory was essentially absent. Saccadic eye movements were mildly slowed. Despite moderate paratonic rigidity, his strength was normal throughout. Tests of coordination of arms and legs were also normal when evaluated with the patient seated or lying. There was no

myoclonus. Grasp, suck, and root reflexes were easily elicited. Deep tendon reflexes were 1+ and symmetric, and the plantar responses were flexor. He displayed marked retropulsion and required two people to help him stand. He could not initiate gait, but when tugged forward, he occasionally took slow short steps over a slightly widened base.

He underwent two lumbar punctures 3 days apart with large-volume (30–40 mL) CSF removal (CSF tap test³). The opening pressure was 180 mm for both. All CSF

Figure 2. EEG on presentation showing a typical periodic sharp-wave complex. The lengths of the horizontal and vertical bars represent 1 second and 100 μ V, respectively. Such complexes typically lasted 10 to 15 seconds and occurred every 4 minutes in the two EEGs performed during the initial hospitalization. The predominant background rhythm with the patient awake was theta at 5 to 7 Hz.



and other standard laboratory studies for reversible dementias were normal. Three EEGs consistently showed not only generalized slowing, but also brief runs of periodic sharp-wave activity (Fig. 2), considered characteristic of Creutzfeldt-Jakob disease (CJD)⁴ in the absence of evidence for metabolic or inflammatory encephalopathy.

With the lack of clinical response to the CSF tap tests during this 2-week hospitalization and a new concern for CJD, the patient was discharged to a skilled nursing facility with a plan to reevaluate him should his clinical status stabilize or improve. Over the next month there, his family and nursing staff reported a slight improvement in his speech and gait. At times, he was able to walk with a walker and staff assistance. He remained incontinent, however, and unable to bathe or dress himself.

In early May, he was re-admitted to our hospital. He demonstrated more spontaneous speech but also more confabulation, perseveration, and emotional lability. He remained markedly disoriented and largely abulic. A follow-up EEG no longer showed any sharp-wave activity, although background slowing persisted. FDG-positron emission tomography (PET) demonstrated symmetric focal hypometabolism in the temporoparietal and frontal regions (Fig. 1B), a pattern typical of Alzheimer's disease.⁵

Nevertheless, based on the possibility of a delayed clinical response to the earlier CSF tap tests and the resolution of periodic sharp waves, the decision was made in accordance with the family's wishes to perform ventriculoperitoneal shunting. On May 25, 1994, a Radionics medium-pressure shunt valve (set to keep CSF pressure between 80 and 120 mm) was inserted into the anterior horn of the right lateral ventricle. A right frontal-cortical and white-matter biopsy, performed during the shunt placement, was entirely normal. Specifically, no spongiform changes, senile plaques, or neurofibrillary

tangles were seen (on Bielschowsky as well as hematoxylin-eosin staining).

Three days after shunting, the patient became remarkably more alert and talkative. He was now continent of urine and stool, and requested assistance to use the bathroom. Six days after shunting, he was oriented to hospital and walked independently. After further recovery at a rehabilitation hospital, he returned home where his cognitive function continued to improve. At follow-up 2 years after shunting, he is enjoying full independence, living alone and cooking, shopping, and managing his finances on his own. He has regained his memory for recent and remote events, except for the

Table 1. Neuropsychiatric Test Scores Before and After Ventriculoperitoneal Shunting

	Preshunting	Post-shunting	
		1 mo	5 mo
Attention	12	33	34
MDRS			
Attention (37)	12	33	34
Memory (25)	1	9	18
Orientation (9)	0	3	9
Initiation (37)	1	16	26
Construction (6)	0	4	6
Conceptualization (39)	1	23	30
Total (144)	15	85	114
Boston Naming Test (60)	0	21	25

Cognitive function was assessed using the Mattis Dementia Rating Scale (MDRS) and the Boston Naming Test at presentation 2 months prior to ventriculoperitoneal shunting (Preshunting); and 1 and 5 months following the procedure (Post-shunting). Parentheses contain the highest possible scores for the two tests as well as for the MDRS subtests. The mean total score on the MDRS for healthy control subjects matched in age and education to our patient are 141.⁶ By contrast, a score of less than 90 correlates with severe dementia.⁶ Both the MDRS and the Boston Naming Test were administered by the same neuropsychologist on all dates.

period of his profound dementia. He continues to use a cane for safety because of mild residual unsteadiness of gait. He is no longer incontinent.

Follow-up neuropsychiatric evaluations substantiated the cognitive gains with continued improvement in composite scores on the MDRS and the Boston Naming Test for at least 5 months after shunting (Table 1). Subtest assessment indicated the global nature of the dementia and its reversal. Follow-up head CT scans demonstrated marked resolution of ventriculomegaly and the presence of bilateral subdural hygromas (Fig. 1A). A repeat PET study showed resolution of the focal hypometabolism (Fig. 1B).

DISCUSSION

NPH presents a rare opportunity to reverse a specific form of dementia. Despite the characteristic clinical triad and radiographic presentation of NPH in our patient, we were initially dissuaded from recommending CSF shunting because of the narrow benefit-to-risk ratio for this procedure² and several atypical features. First, the rapid development of severe symptoms in less than 1 month contrasted with the usual time course in NPH: generally over months to years, with an insidious onset of gait disturbance usually preceding cognitive and urinary difficulties.⁷

Second, repeated CSF tap tests did not lead to prompt improvement in symptoms. Such negative CSF tap tests have been correlated with less favorable outcomes in patients who went on to surgical shunting, although exceptions do occur.³

Third, the evidence for alternative etiologies of NPH symptoms undermines prospects for a beneficial response to shunting.⁸ Although the EEGs performed on the patient displayed nonspecific generalized slowing of background rhythms commonly observed in NPH patients,⁹ they also initially contained periodic sharp-wave complexes. This pattern, considered an electrophysiologic hallmark of CJD, is occasionally seen in metabolic and inflammatory encephalopathies, but has not been previously reported in NPH. Furthermore, FDG-PET imaging demonstrated a symmetric multifocal pattern of cerebral hypometabolism typical of Alzheimer's disease. In fact, it has recently been suggested⁵ that this Alzheimer's disease pattern on PET is helpful in excluding a diagnosis of NPH, which in a small series was associated instead with a generalized nonfocal pattern of hypometabolism.

Kaye et al,¹⁰ however, have described a case of NPH with shunt-responsive PET findings of asymmetric multifocal hypometabolism particularly involving the

temporoparietal and frontal regions, and thus questioned the use of PET in evaluating NPH. Our similar finding of a dramatic reversal of clinical and focal PET abnormalities after shunting, as well as a normal frontal-brain biopsy in our patient supports their contention, arguing further against a role for PET in diagnosing NPH.

Clinical and electrophysiologic improvements were noted more than a month after CSF tap tests in our patient, raising the possibility of a delayed benefit of lumbar puncture, weakening the evidence for an irreversible dementia, and thus prompting surgical decompression of the cerebral ventricles. The abrupt and dramatic recovery, taken together with the triad of symptoms on presentation and the absence of consistent evidence for an alternative etiology, confirmed the primary diagnosis of NPH. This case, while anecdotal, nonetheless illustrates the importance of focusing on the core features of NPH in considering a shunting procedure. Until their predictive value can be significantly increased, ancillary studies in NPH will remain of limited value in these difficult management decisions.

Acknowledgment

We thank Dr. Keith Chiappa for his helpful discussions.

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