Normal-pressure hydrocephalus: a difficult diagnosis

Normal-pressure hydrocephalus was first described more than 40 years ago as a syndrome of cognitive impairment, gait disturbance, and urinary incontinence affecting the older adult. It is a syndrome of symptoms and signs, which should be diagnosed clinically with the aid of radiological imaging and diagnostic tests. Dementia caused by normal-pressure hydrocephalus is potentially reversible. We report a case of a patient in which this disorder was diagnosed clinically after radiological imaging, but with some uncertainty. We emphasise the need for increased awareness of the presentation of normal-pressure hydrocephalus and the associated diagnostic challenges.

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Normal-pressure hydrocephalus was first described by Hakim and Adams in 1965.[1] It is a progressive syndrome with an insidious onset—clinical presentation is diverse and often non-specific. The classic triad of cognitive impairment, gait disturbance, and urinary incontinence can be found in later stages of this syndrome. Normal-pressure hydrocephalus is a potentially treatable cause of dementia and general medical physicians should be aware of this diagnosis. We describe a case highlighting delay and difficulty in the diagnosis of a patient's deterioration in mobility and cognitive function.

Case report

An 84-year-old man with hypertension was referred to the geriatric outpatient clinic with a 7-month, gradual, progressive history of becoming "doddery" (ie, unsteady gait), a feeling of falling forwards, and memory problems. At examination he had difficulty walking, marked truncal ataxia, hypertonia, and hyper-reflexia in the lower limbs, but with no weakness. His movements were coordinated. Initially we thought he had early cervical myelopathy, and cervical spine X-rays were requested, which showed multilevel spondylitic changes.

The patient was reviewed 3 months later; he felt quite well and was fully independent, but his wife maintained that he was becoming more forgetful and had a tendency to become unsteady. At a further review, at 8 months, he was more unsteady. The ataxia was not thought to be significant, and another review was scheduled for 4 months later, 12 months after his original visit. At this assessment, he had not fallen, but said his legs were weak, although not any worse. He had further deterioration in memory. At this point a CT scan of the brain was requested on sucpiscion of normal-pressure hydrocephalus (figure 1). The scan showed that the ventricles were prominent and out of proportion with the sulci, and mild periventricular hypodensities were present. These radiological features suggested normal-pressure hydrocephalus.

Subsequent MRI of the brain was reported as showing: **"moderate dilation of all four ventricles, out of proportion to the size of the extra-axial cerebrospinal fluid spaces. Hyperintense foci seen in the periventricular white matter, pons, and in the basal ganglia region in keeping with chronic ischaemic changes due to microvascular disease (figure 2). The flow void in the cerebral aqueduct appears normal in size and flow, and is less likely to be a shunt-responsive normal-pressure hydrocephalus."** MRI of the cervical

Figure 1: CT of the brain showing prominent ventricles out of proportion with the sulci

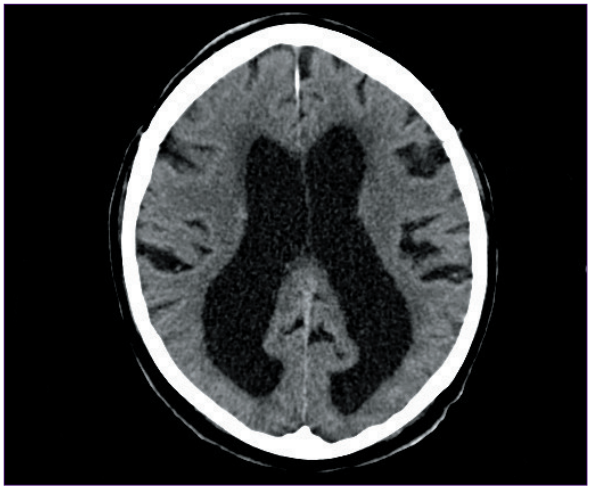
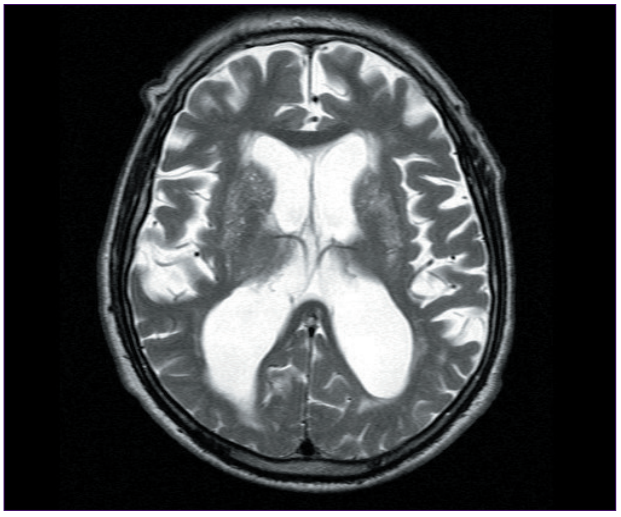


Figure 2: MRI of brain showing moderate dilatation of all four ventricles



spine showed multilevel disc degeneration with disc bulge, osteophytosis, arthritis of the facet joints, and hypertrophy of the ligamentum flavum causing cord compression at C5–6 with myelomalacia and central canal dilatation.

The patient was referred for neurosurgical opinion and possible intervention to improve balance. The neurosurgeon identified that the patient had difficulty in walking and had short-term memory loss. He had no urinary difficulties or symptoms indicative of myelopathy in the upper limbs. He had signs of mild myelopathy—with global hyper-reflexia, but negative Hoffman's sign, no clinically significant truncal ataxia, and Rhomberg's sign was negative. These findings contrasted with those in the geriatric clinic. From these radiological and clinical findings, the patient was diagnosed as having a form of multi-infarct dementia, and his vascular dilatation was not out of proportion with the enlargement of the extra-axial cerebrospinal spaces.

Therefore, the patient did not have normal-pressure hydrocephalus causing memory loss, which was supported by the absence of ataxia and urinary problems. The mild cervical myelopathy was not responsible for his walking difficulties. MRI of the lumbar spine was suggested to investigate possible lumbar canal stenosis because the patient's leg symptoms were postulated to be due to claudication. However, the patient declined this investigation and was discharged back to primary care.

Discussion

We know that hydrocephalus is an abnormal enlargement of the ventricles due to accumulation of cerebrospinal fluid resulting from disturbed flow, absorption, or secretion of cerebrospinal fluid. Normal-pressure hydrocephalus results from an abnormality in absorption of cerebrospinal fluid. No obstruction in flow of cerebrospinal fluid exists, and it is in physiological range, but a pressure gradient exists between the brain and ventricles resulting in a chronic hydrocephalus, with subsequent encroachment and damage to the surrounding brain parenchyma.

Gait disturbance

Gait disturbance is usually the first symptom in normal-pressure hydrocephalus, since the fibres of the corticospinal tract supplying motor function to the legs pass close to the lateral ventricles in the corona radiata.[4] Gait is wide based, apraxic, and described as magnetic as patients take small steps and have reduced floor clearance.[5] As described in the patient above, hypertonia and hyper-reflexia maybe found with absence of weakness or incordination.[6] After successful shunting, disturbance of gait is the first symptom to improve.

Urinary incontinence

Urinary incontinence is usually seen in advanced cases of normal-pressure hydrocephalus. The first urological symptoms are those of increased urinary frequency and urgency. Urodynamic studies will demonstrate bladder detrusor muscle instability. The patient may develop incontinence of urine and faeces but some patients with normal-pressure hydrocephalus may never develop urinary symptoms. Some elderly patients will have urinary and faecal incontinence due to other comorbid states, which can lead to misdiagnosis.

Cognitive impairment

Normal-pressure hydrocephalus is estimated to cause 5% of all dementias.[7,8,9] The dementia is subcortical, which helps to differentiate it from disease such as Alzheimer's disease. The cognitive impairment is characterised by inertia, forgetfulness, and poor executive function.[10] Cognitive impairment is not the most overt symptom or presenting feature of normal-pressure hydrocephalus.[11]

Diagnostic tests

Normal-pressure hydrocephalus can be difficult to diagnose because the presenting symptoms can be non-specific, but the possibility of this diagnosis must be remembered. Neuroimaging with CT and MRI will show the presence of ventricular dilatation and disproportionate ventriculo-sulcal dilatation. Elderly patients can naturally show evidence of ventriculomegaly on imaging, and patients with cerebral microvascular disease, such as the patient described, will also show such changes.

In the early stages of normal-pressure hydrocephalus, this affects mainly the frontal and temporal horns with preservation of the surrounding brain tissue. An estimation of pressure of the cerebrospinal fluid, and assessment of clinical response to drainage of cerebrospinal fluid can be done.

Conclusion

Normal-pressure hydrocephalus is a condition with presentation akin to other conditions causing mobility, memory, and urinary difficulties. As discussed, our case had diagnostic ambiguity since this patient presented mobility and memory difficulties that were attributed to cervical myelopathy, normal-pressure hydrocephalus (since incontinence can begin late), multi-infarct cognitive impairment, and finally possible lumbar canal stenosis. This diagnostic uncertainty was not resolved after radiological imaging because the multiple opinions of the findings differed.

Normal-pressure hydrocephalus can be difficult to diagnose, or may be misdiagnosed for months to years as other conditions such as Parkinson's disease or Alzheimer's disease. Doctors should remain vigilant to the possibility that normal-pressure hydrocephalus is causing a patient's deterioration in mobility, memory, and urinary function, and strive to make a careful timely diagnosis.

We have no conflict of interest.

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