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Case report

Senile brain atrophy and hydrocephalus

—— A case of treatable dementia, idiopathic normal pressure hydrocephalus ——

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ABSTRACT

A 76y male suffering from progressive dementia (Hasegawa's dementia scale = 6), urinary incontinence, and gate disturbance received CSF shunting. The clinical findings of MRI, CT-cisternography, and a CSF-tap test were not typical for idiopathic normal pressure hydrocephalus (iNPH); rather, the MRI showed brain atrophy. The decision to perform shunting surgery was made due to the clinical manifestation of progressive dementia, and fortunately, it was successful. Senile brain atrophy does not rule out hydrocephalus. The indication of CSF shunting for senile iNPH is outlined.

Key words: Treatable dementia, Normal pressure hydrocephalus, Idiopathic

INTRODUCTION

Idiopathic normal pressure hydrocephalus (iNPH) represents a treatable form of dementia (Martin U, 1989). Recent estimates of the incidence of this condition are in the region of 2.9% of in elderly subjects of >65y (Hiraoka K, 2008). Thus >30000 dementia senile >65y people are estimated to be treatable yearly in Japan. As dementia is an increasing demographic problem, treatable forms such as iNPH have become a central issue in geronto-neurology. However, when brain atrophy accompanied with marked dilatation of ventricles is shown on an aged person's brain computed tomography (CT) scan and/or magnetic resonance imaging (MRI), it is sometimes very difficult to make differential diagnoses between brain atrophy and iNPH in order to determine the surgical indication for shunting. Progressive dementia with marked dilatation of the ventricular system might be a reliable manifestation for CSF-shunting, although adjunctive investigations such as CT-cisternography and the CSF-tap test are not typical for iNPH. We emphasize the importance of progressive dementia when nominating senile patients for cerebro-spinal fluid (CSF) shunting.

CASE PRESENTATION

A 76y male suffering from progressive month by month dementia (Hasegawa's dementia scale = 6 on admission), urinary incontinence, and gait disturbance of the petit-pas gait type received CSF shunting. Ventricular dilatation (Evans ratio>3) with marked peri ventricular lucency (PVL) and a slack subdural space with tightness of high parietal interhemispheric fissure were marked on admission MRI (Figure 1). However, these findings are not typical and controversial for iNPH; and rather, brain atrophy was detected. No history of head injury and a normal MR Angiograph (MRA) were confirmed. His CSF-dynamics were investigated by CT-cisternography revealing almost no delay in CSF washout and/or ventricular reflex of intrathecally injected contrast material (Isovist™) (Figure 2). A 30ml CSF sample (daily tap test) was not definitive for iNPH. The concentrations of Vitamins B_{12} and B6 were normal, and no hypothyroidism was revealed on a

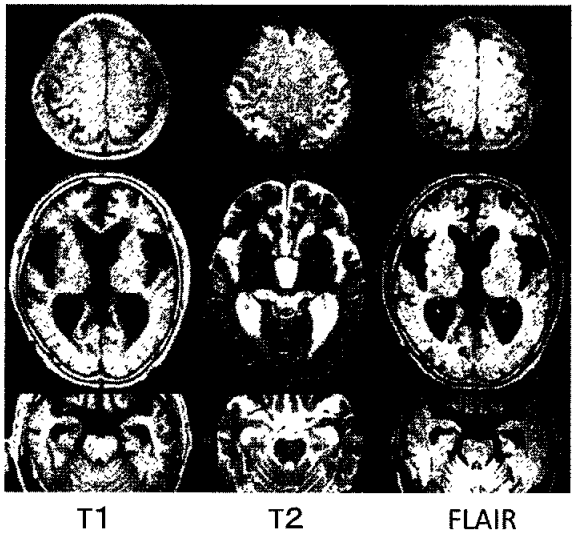

Fig. 1 Axial views of magnetic resonance imaging (MRI) just before CSF shunting, T1-weighted MRI (*left*), T2-weighted MRI (*middle*), and fluid-attenuated inversion recovery (FLAIR) MRI (*right*). Ventricular dilatation (Evans ratio>3) with peri ventricular lucency (PVL) and a slack subdural space, and trapped CSF were observed in the Sylvian fissure. These findings were less diagnostic for iNPH, and rather they were viewed as brain atrophy or a mixed configuration of iNPH and brain atrophy

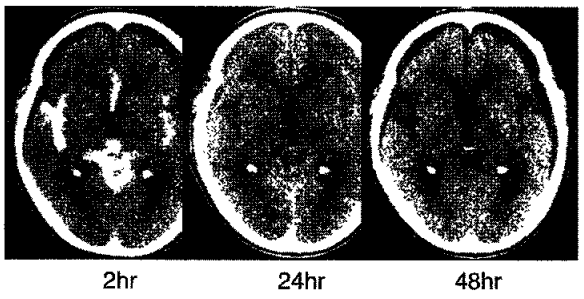

Fig. 2 CT-cisternography. 3 serial views, 2, 24, and 48 hrs after intrathecal injection of contrast material (Isovist). The absence of washout delay or contrast material reflex in delayed phase at 48 hrs excluded possible iNPH. Incidentally, a CT scan revealed hygloma and shrinkage of ventricles (a form of external hydrocephalus) and a possible mechanism for this is described in text

blood examination. As a result, a Hakim™ programmable V-P shunt system was successfully installed. No any post-operative complications were observed and the patient demonstrated a good operative course.

A CT scan 1 month after CSF shunting revealed normalization of ventricular size as shown in Figure 3. As for higher brain

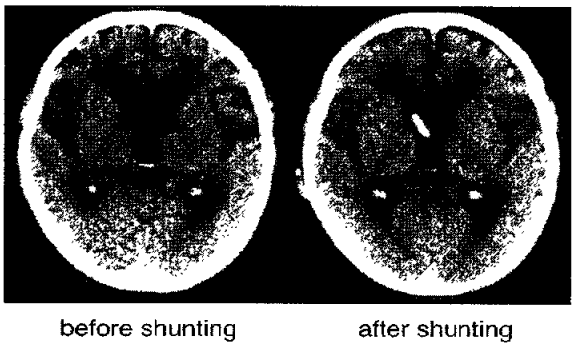

Fig. 3 CT scans before (*left*) and 1 month after (*right*) CSF shunting revealed normalization of ventricular size without hygloma. A ventricular catheter (Hakim™ programmable V-P shunt system (Codman, MA, USA)) was installed into the right ventricle through its anterior horn. The final pressure of the shunting system was 90mmCSF

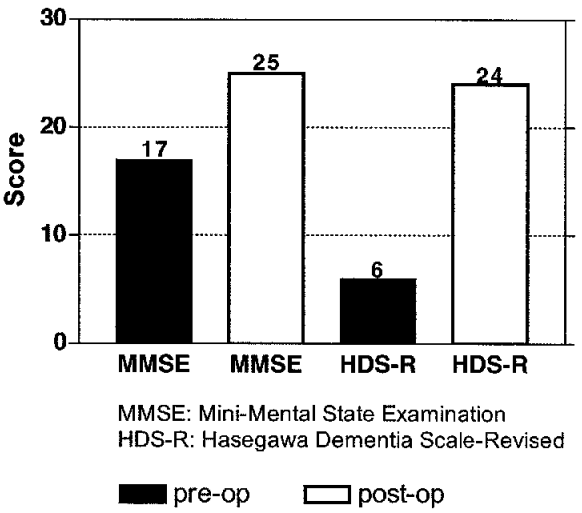

Fig. 4 Scores of the Mini-Mental State Examination (MMSE) and Hasegawa's Japanese Dementia Scale – revised version (HDS-R) performed on 1 month before and/or 17 days after CSF shunting. Cognitive impairment (>24 in both examinations) disappeared after the shunting

function, Figure 4 shows that Hasegawa's Japanese Dementia Scale (HDS-R) and Mini Mental State Examination (MMSE) were performed 1 month before and/or 17 days after CSF shunting. The patient's cognitive function was considerably improved, and his progressive dementia completely disappeared after CSF-shunting. The patient gradually regained micturition desire, and his urinary incontinence disappeared within a few days.

DISCUSSION

In case of senile dementia whose CT and MRI are not typical or controversial for iNPH, only typical symptoms manifesting progressive dementia might indicate CSF-shunting. Finding normal CT-cisternography and brain atrophy with slack subdural space (or pan-dilated subarachnoid space) does not rule out hydrocephalus in aged persons especially when high parietal interhemispheric fissure is relatively tight as shown in this case. Indeed, this case report cautiously notes that the relationship between brain atrophy and hydrocephalus is very complicated and that sometimes brain atrophy masks the presence of hydrocephalus. The most predictable symptom for successful shunting was manifestation of progressive dementia. Generally, however, surgical indication for patients aged >80 should be made carefully for CSF shunting surgery after deeply considering their potential complications, present status, and anesthesiological risk assessment. The type of shunting surgery such as ventriculo-peritoneal shunt, ventriculo-atrial shunt, and lumbo-peritoneal shunt, etc, should be chosen carefully on a case by case basis to minimize surgical invasiveness. In order to make a differential diagnosis of senile dementia, hydrodynamic studies (i.e. CSF flow resistance) and blood chemical examinations (i.e. serum VB_{12} and folate levels, thyroid function test, and VDRL for syphilis, etc) should be performed during the diagnostic process for iNPH; however, the therapeutic effect of high dose administration of Vitamin B against Alzheimer's is controversial (Aisen PS, 2008).

This case did not fulfill the diagnostic guidelines for possible iNPH (Marmarou A, 2005). However, fortunately, the dementia of our patient disappeared after ventriculo-peritoneal shunting. Our only motivation to embark on the surgical intervention was based on his "progressive dementia". Brain atrophy accompanied by progressive dementia in aged persons should be carefully differentiated between Alzheimer's dementia, and chronic subdural hematoma, and other types of dementia. There are also many differential diagnoses for progressive dementia such as AZ, vascular dementia (lacunar infarction), DLB, Parkinson's disease, PSNP, etc. (Holodny AI, 1998, Iddon JL, 1999). The progressiveness of dementia might be the most predictive factor for the effect of shunting. During careful clinical observation, our patient's cognitive function deteriorated month by month. On the other hand, if the senile dementia is not progressive even though MRI findings are typical for iNPH, shunting might not be effective. The gait disturbance of shown of normal pressure hydrocephalus typically involves petit-pas gait, magnet gait, or broad-based gait; however, sometimes it is difficult to distinguish from Parkinson's disease (Stolze H, 2001) as some cases only show brain atrophy on MRI.

Although the guidelines recommend a CSF tap test to rule out iNPH, repeated removal of CSF failed to confirm the progressive dementia as possible iNPH in our case. Such cases should be carefully diagnosed by advanced examinations such as continuous drainage of the CSF. However, in aged persons, it is contraindicated for safety reasons. Furthermore, cerebral blood flow (CBF) dynamic studies as a non-invasive study for the aged persons for iNPH is also controversial, as CBF studies using ^{133}Xe inhalation in iNPH brains support the notion that increases in CBF probably do not account for clinical improvement in normal pressure hydrocephalus (Graff-Radfird NR, 1987). Thus, at present, the definite diagnosis of iNPH can only be made after shunting even in cases such as ours which was diagnosed as sub-possible iNPH according to the guidelines for iNPH.

It is rare to observe a form of external hydrocephalus during the progress of iNPH as it temporarily disappears or ventricular enlargement with hygloma is incidentally observed on CT cisternography (Fig. 2). As one of possible mechanisms of this, sudden alterations in CSF dynamics might occur via spontaneous CSF shunting through an arachnoid membrane perforation (a form of external hydrocephalus or masked iNPH). This condition can develop during the progress of iNPH as described below. A schematic drawing of a possible mechanism of non-obstructive hydrocephalus is shown in Fig. 5. In typical iNPH, the following additional findings are highly diagnostic: (1) a high frontal interhemispheric space and (2)

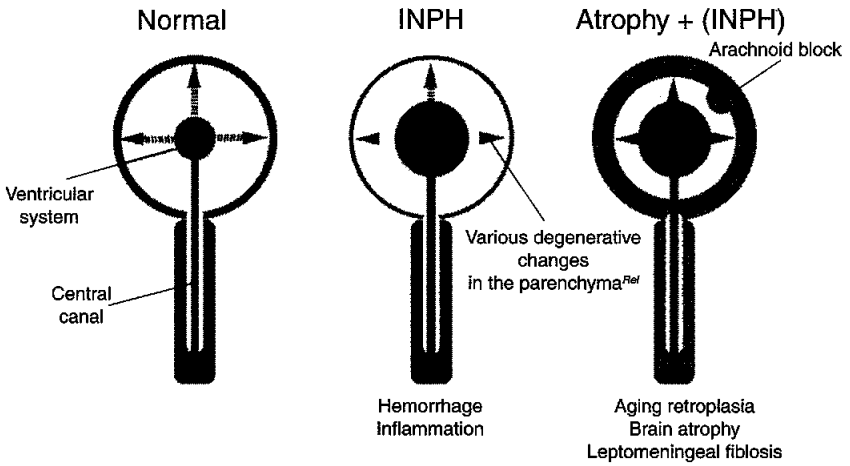

Fig. 5 Schematic drawing of the mechanism of non-obstructive hydrocephalus. In typical iNPH, the following additional findings are highly diagnostic: (1) a high frontal interhemispheric space and (2) arachnoid trapping. However, in aged iNPH patients with cerebral atrophy, their findings are not diagnostic. In such cases, a definitive diagnosis can only be made after shunting and/or continuous CSF drainage. The possible cause of iNPH is not of vascular origin, but rather is related to parenchymal aging process such as adhesion, inflammatory reaction, and leptomeningeal fibrosis

arachnoid trapping (or dilated subarachnoid space). However, in aged iNPH with cerebral atrophy, these findings are not diagnostic. In such cases, a definitive diagnosis can be made by shunting and/or continuous CSF drainage. In normal parenchyma, the CSF dynamics are balanced to keep the size of ventricles normal. However, CSF flow resistance explains ventricular dilatation such as iNPH (Bech RA, 1999). Perhaps these findings might be a possible cause of iNPH. However, the cause of iNPH is probably not of vascular origin, but rather is related to parenchymal aging process such as adhesion, inflammatory reaction, and leptomeningeal fibrosis. However, at the present time the pathogenesis of iNPH is controversial (Ishikawa M, 2008), so making a differential diagnosis between senile brain atrophy and iNPH should be done carefully after clinical observation of the progressiveness of the dementia. In order to keep the brain young, CSF dynamics are an essential aspect together with the macro aging process of vascular and parenchymal degeneration.

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