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CLINICAL ARTICLE
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Observations on the Activation of Chronic Compensated Hydrocephalus in Adult Patients

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Objective: There is a broad spectrum of compensated hydrocephalus. Various terms such as long-standing overt ventriculomegaly in adult (LOVA) has been coined, however, even such terms leave diverse aspect of this condition out of account. We have experienced compensated hydrocephalus cases which were considered to be activated after a long time period of quiescent state, and tried to compare their clinical characteristics with the relatively well described entity of LOVA.
Methods: We conducted a retrospective review of 206 patients who underwent ventriculoperitoneal shunt (VPS) between February 2001 and May 2012. Of these, 6 patients had chronic compensated hydrocephalus. The clinical and radiological characteristics are evaluated.
Results: Definite triventriculomegaly was observed in two patients. Macrocephaly was observed in two cases, one with aqueductal stenosis (AS), the other with unknown status of aqueduct. All of the cases with triventriculomegaly were normocephalic. Spinal causes were thought as aggravating factor in two. Two endoscopic third ventriculostomy and eight VPS were performed in five patients. Four patients responded well but one took a very complicated course.
Conclusion: The relationships between macrocephaly, triventriculomegaly, and AS suggested in other studies were inconsistent. Blockage or narrowing of cerebrospinal fluid pathways were observed at various sites. Disturbances of spinal arachnoid pathways were related to the activation in some cases. Treatment is to be tailored individually considering various reigniting event. It is suggested that this entity is to be evaluated for better nomenclature reflecting diverse aspects of this condition. Further study is needed to elucidate underlying pathophysiology and effective management.
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KEY WORDS: Hydrocephalus · Long-standing overt ventriculomegaly in adult · Macrocephaly · Aqueductal stenosis.

Introduction

There is no nomenclature consensus in describing patients with chronic form of exceptionally enlarged ventricles and no overt sign of increased intracranial pressure (ICP). There is a broad spectrum of compensated hydrocephalus and the underlying cause of this form of chronic compensated hydrocephalus has been questioned. Little attention has given to this entity and not so many studies have been made so far. There are confusing terms used to describe these conditions such as the syndrome of hydrocephalus in young and middle-aged adults (SHYMA), late-onset idiopathic aqueductal stenosis (LIAS), long-standing overt ventriculomegaly (LOVA) of the adult, and late-onset aqueductal stenosis (AS).[3,5,13] Some authors proposed that LOVA must be considered as a special form of LIAS.[8]

Cerebrospinal fluid (CSF) dynamics may change over time between active and inactive states in these entities but the reason for late decompensation remains obscure. However, a new event in the central nervous system (CNS) may offset the compensatory mechanism in some patients. How to treat these patients is also a very controversial issue.[10] Ventriculoperitoneal shunt (VPS) with adjustable valve or endoscopic third ventriculostomy (ETV) is generally selected. The authors illustrate their experiences and review

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the literature.

Materials and Methods

The authors conducted a retrospective review of 206 patients who underwent VPS for hydrocephalus between February 2001 and May 2012. Of these, 6 patients had chronic compensated hydrocephalus. Chronic compensated hydrocephalus was defined as a case that has exceptionally enlarged ventricles with relatively well identified sulci and a significantly enlarged or destroyed sella turcica, and/or macrocephaly. The frontal horn size and biparietal diameter (BPD) were checked to figure out the Evan's ratio (ratio of frontal horn to maximal BPD) in all patients. Dimensional change & contour of sella turcica were examined using skull X-rays or scout film of computed tomography (CT). Macrocephaly was diagnosed when head circumference was measuring more than two standard deviations. The presence of triventriculomegaly and AS were checked on imaging studies. AS was defined as absence of the whole length of the aqueduct on midsagittal T1- and T2-weighted magnetic resonance imaging (MRI) images. Spine imaging was performed for patients who had symptoms or possibility of spine lesions.

Results

There were 91 women and 115 men whose age ranged from 20 to 78 years (58.9±13.1). The common cause of hydrocephalus was subarachnoid hemorrhage (SAH), trauma, intracerebral hemorrhage in order. The gravity assisted valve (GAV) valve (Aesculap-Miethke, Tuttlingen/Postdam, Germany) was used in 82 patients (39.8%). Programmable valves were placed using ProGAV (Aesculap-Miethke, Tuttlingen/Postdam, Germany) in 38 (18.4%), Codman Hakim programmable valve (CHPV, Codman, Johnson & Johnson Co., Raynham, MA) in 37 (18%), and Strata valve (Medtronic Neurosurgery, Goleta, CA) in 12 patients (5.8%). Various pressure regulated valves such as Pudenz (Heyer-Schulte Pudenz Flushing valves, Integra™, Plainsboro, NJ) or Novus valves (Novus™ valve system, Integra™, Plainsboro, NJ) were used in 37 patients (18%).

The frontal horn size was 52.0±9.9 mm, BPD was 131.1± 11.4 mm, and the average of Evan's ratio was 0.42±0.30 (mean±standard deviation). Six cases of chronic form of marked ventriculomegaly including one nonoperative case were identified (Figure 1). And their mean frontal horn size was 61.6±8.2 mm, and mean BPD was 138.7±10.3 mm. The average Evan's ratio of these patients was 0.44±0.05.

Mean age was 47.8 years old (range: 23−66) and male to female ratio was 2 : 1. Mean follow-up period was 12 months (range: 2−24). Two ETV and eight VPS operation were performed in five patients. Four patients had good results but one patient took a very complicated clinical course and beyond remedy. The relationships between macrocephaly, triventriculomegaly, and AS were not constant. Definite AS was observed only in 1 case. Other 2 cases had narrowing distal to the aqueduct itself. Triventriculomegaly was observed in two patients. Macrocephaly was observed in two cases, one with AS, the other with unknown status of aqueduct. It was unable to evaluate the status of aqueduct because MRI had not been done in this case. All of the cases with triventriculomegaly were normocephalic. Spinal causes were thought as aggravating factor in two. Spinal causality was suspected in another one. Cases were summarized in Table 1.

Illustrative cases

Case 2

A 43-year-old male driver presented with chronic headache. After workup, a pressure regulated valve was placed. At the age of 46 and 48, he underwent operations at other hospital for spinal arachnoid cyst. At the age of 52, he collapsed during bathing and was brought to the emergency room (ER). CT scan showed ventriculomegaly and Evan's ratio was increased to 0.42. Spine MRI showed persistent long segmental (T10-L2) arachnoid cyst and severe arachnoid adhesion on magnetic resonance myelography (Figure 2A). ETV was thought to be inadequate to perform in this case. Shunt was revised with GAV. The patient's condition improved rapidly and postoperative Evan's ratio decreased to 0.37. Six months later, he lost his consciousness during driving and developed progressive mutism, urinary incontinence. CSF was aspirated by shunt puncture. There was no evidence of shunt infection and the patient's condition improved after aspiration. It was decided to change GAV to ProGAV with concerns about underdrainage of GAV that have been reported.[6,7] The mental status deteriorated unexpectedly first postoperative day and follow-up CT showed severe ventriculomegaly. Emergent external ventricular drainage (EVD) was placed on left side and then the patient's neurological condition recovered. It was planned to check distal catheter in the suspicion of problem in the peritoneal absorptive capability and found there was very severe adhesion in the peritoneal cavity. Omental adhesiolysis and repositioning of distal catheter were done. EVD was changed to low pressure regulated valve and the program-

TABLE 1. Summary of cases

Case No	Sex/ Age	PHx	Chronic ICP Sx	Macro-cephaly	IQ	Sellar enlargement	Triventri-culomegaly	AS	2nd hit	Operation	Follow-up (months)	mRS
1	M/29	Meningitis at 3 YO	None	Y	Subnormal	Y	±	NA	EDH	Craniotomy	24	0
2	M/43	None	Headache	N	Normal	Y	Y	Y	Spinal operation, omental adhesion	Pr-valve, spinal op, GAV, Pro-GAV+Pudenz, ETV+pro-GAV removal	5	4
3	F/61	None	None	N	Normal	Y	N	N	SAH, spinal Infection (?)	Embolization, pro-GAV, spinal op	16	1
4	F/65	None	None	N	Normal	Y	Y	±	Nontraumatic spinal hemorrhage	GAV	15	0
5	M/23	None	Headache, nausea	N	Normal	±	N	N	NA	ETV, GAV	2	0
6	M/66	None	None	Y	Normal	Y	N	N	NA	Pro-GAV	10	0

AS: aqueductal stenosis, EDH: epidural hematoma, ETV: endoscopic third ventriculostomy, F: female, GAV: gravity assisted valve, ICP: intracranial pressure, IQ: intelligence quotient, M: male, mRS: modified Rankin score, N: no, NA: not available, No: number, op: operation, PHx: past history, pr-valve: pressure regulated valve, ProGAV: programmable GAV, Sx: symptom, Y: yes, YO: years old, ±: indefinite, SAH : subarachnoid hemorrhage

mable valve pressure was down regulated, but it was useless for symptom improvement. MRI taken before ETV showed AS, sellar enlargement, upwardly stretched corpus callosum and periventricular edema (Figure 2B). As a last resort, ETV was performed and the ProGAV was removed (Figure 2C). Unexpectedly, third ventricular floor was very narrow and the membrane was relatively thick. Compliance loss due to repeated operation and repetitive deteriorations was considered to be the cause of incomplete recovery and very complicated clinical course. He showed apathy and developed facial palsy and tremor later.

Case 3

A 61-year-old woman with normal intelligence, normocephaly visited ER. A few months ago, she was diagnosed having an unruptured aneurysm and scheduled to embolization. At that time, brain MRI showed diffuse ventriculomegaly. Brain CT taken at ER showed SAH and coil embolization was performed. During intensive care, she becomes drowsy and right upper extremity weakness developed. Lumbar puncture (LP) showed elevated pressure (22 cmH_2O) and CSF drainage made full neurological recovery. A few days later, right hemiparesis with left lower extremity weakness developed. Imaging study showed no ischemic lesion. Postoperative day 16, EVD was placed. After EVD placement, upper extremity weakness was improved but paraparesis persisted. Due to septicemia, EVD was removed seven days later and intravenous antibiotics were used. After general ward transfer, lower extremity weakness persisted and cognitive impairment was observed (Mini-mental state examination 11/30). Lumbar drainage improved neurological condition temporarily. Postictus day 61, a VPS (ProGAV) was placed. After shunt placement the patient's neurological condition improved remarkably. She complained back pain during rehabilitation. Pyogenic spondylodiscitis due to septicemia was found on further examination. Continuous antibiotic irrigation was done and intravenous antibiotics were used. Diagnosis of spinal infection was somewhat delayed and it might be another aggravating factor of CSF absorption disturbance.

She made a near complete recovery and during follow-up, she developed Parkinsonism such as festinating gait, magnetic gait, and difficulty in turning around 14 months after shunt placement. Adjustment of shunt valve pressure made a remarkable improvement within two weeks.

Case 4

A 65-year-old woman visited ER complaining paraparesis, vomiting and voiding difficulty. She had neck pain for a

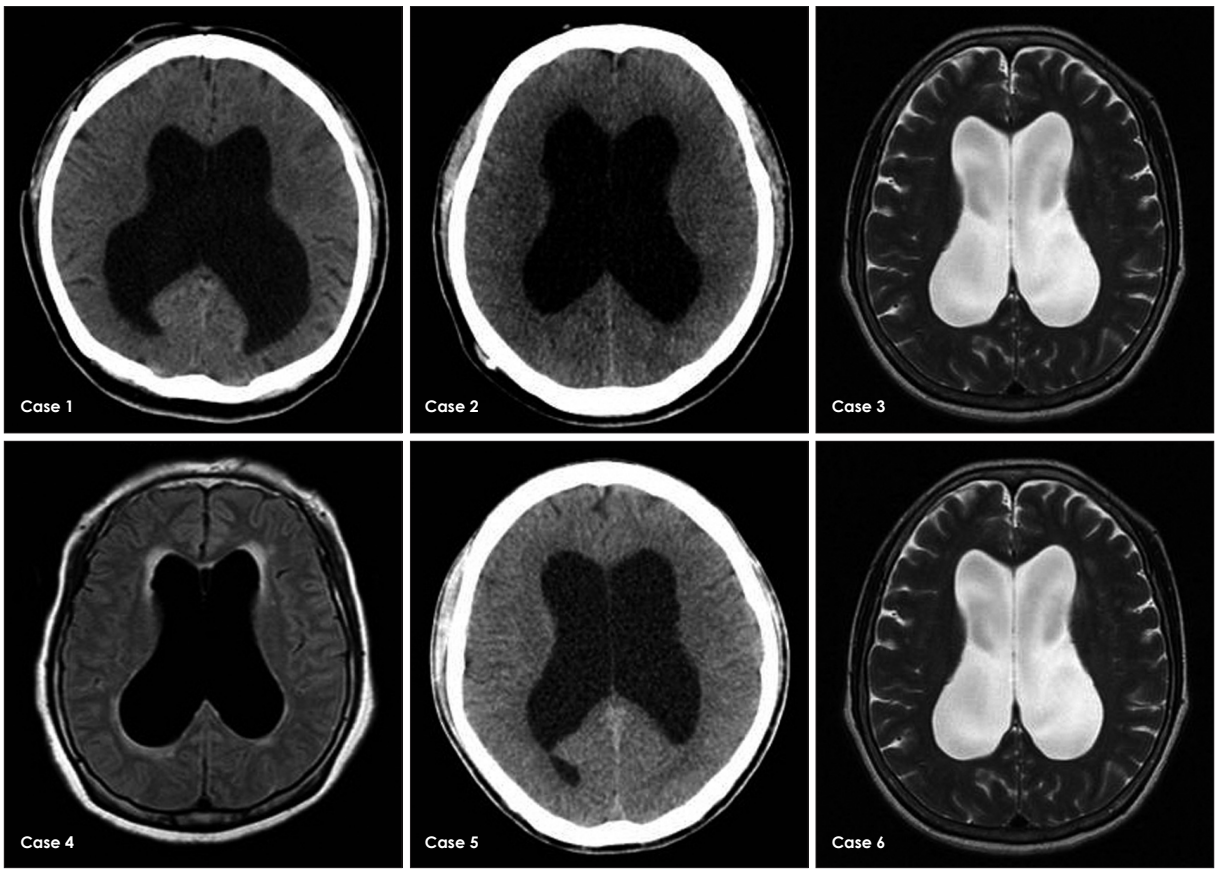

FIGURE 1. CT/MRI axial scans demonstrating ventriculomegaly with relatively well- identified sulci. Periventricular edema is observed in case 4 which suggest acute aggravation of quiescent long-standing hydrocephlalic process. CT: computed tomography, MRI: magnetic resonance imaging.

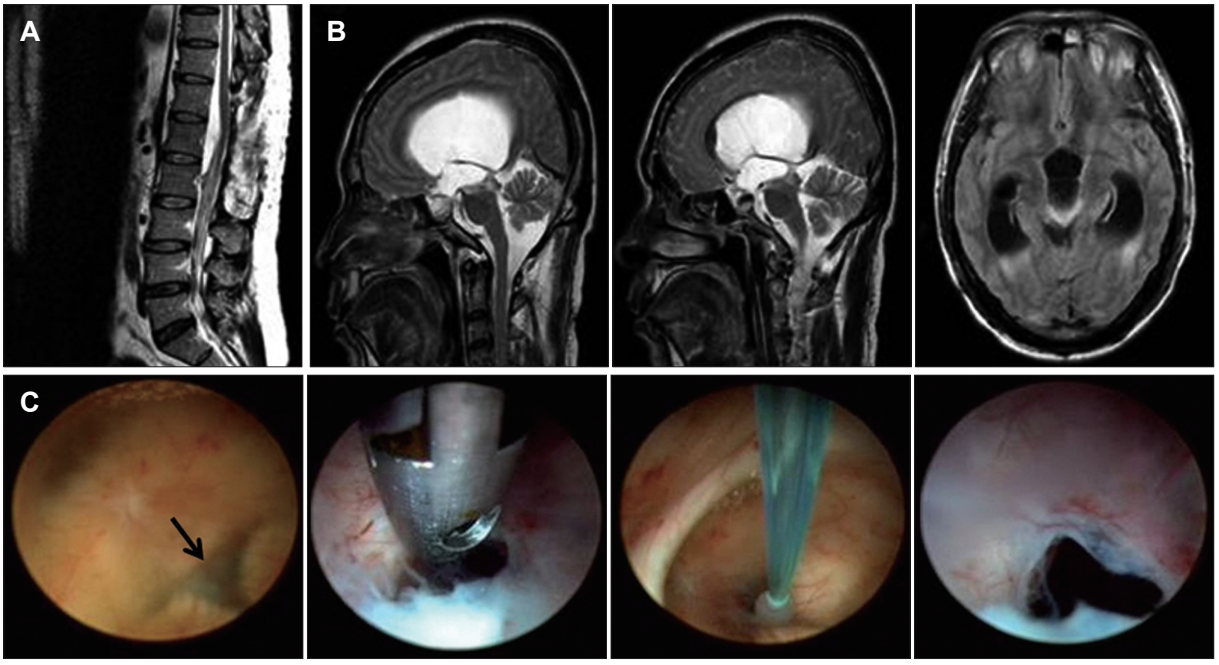

FIGURE 2. T2-weighted sagittal MRI showing persistent T10-L2 segmental arachnoid cyst (A), MRI taken before ETV show AS, sellar enlargement, upwardly stretched corpus callosum and periventricular edema (B), ETV was performed and the fenestration was enlarged using a 3-French Fogarty balloon catheter. Unexpectedly, third ventricular floor was very narrow and the membrane was thick (C, arrow). AS: aqueductal stenosis, ETV: endoscopic third ventriculostomy, MRI: magnetic resonance imaging.

month and temporary hearing disturbance. Brain MRI showed triventricular hydrocephalus with periventricular interstitial edema and abrupt narrowing distal to the aqueduct (Figure 3A). Thoracic spine MRI showed long segmental (from T2 to T10) epidural hematoma (EDH) (Figure 3B). She had no history of trauma. Spinal angiography showed no vascular lesion. LP showed elevated red blood cell count and protein. Opening pressure was 20 cmH_2O. Due to spinal lesion, a VPS (GAV) was selected as a treatment modality. Postoperatively, headache disappeared, voiding difficulty and gait disturbance recovered to normal within a few weeks.

Discussion

The underlying cause of chronic compensated hydrocephalus has been questioned and is not clear yet. Oi et al.[13] attributed it to AS in all patients which begun before their cranial sutures fused during infancy. Diagnostic criteria include severe ventriculomegaly in adults that is associated with macrocephalus measuring more than two standard deviations in head circumference and/or neuroradiological evidence of a significantly expanded or destroyed sella turcica. But their 22 cases included data of pre-MRI era and it was difficult to convince all the cases were caused by AS taking into account their 3D-CT use, and they also included four of 20 cases who were not macrocephalic. Their criteria were not clear and they restrict etiology only to AS in spite of various possible causes.

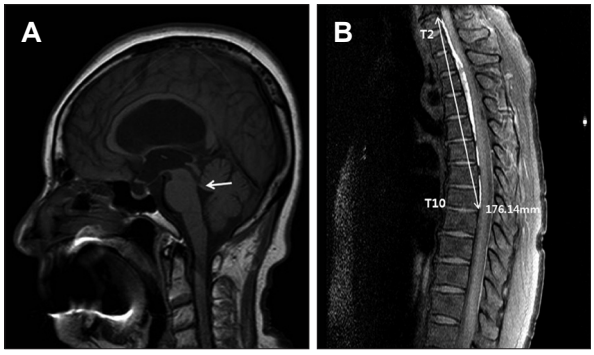

FIGURE 3. Preoperative MRI demonstrating triventriculomegaly, empty sella and abrupt narrowing distal to the aqueduct (A, arrow). Thoracic spine MRI revealed about 18 cm length epidural lesion (B). MRI: magnetic resonance imaging.

Despite the lack of clear cut criteria of normal or enlarged fourth ventricle, triventriculomegaly is usually accepted as a sign of AS and many reports are based on this.[4,5,15] But, triventriculomegaly does not always mean that the underlying cause is AS.[16] Knol et al.[9] suggested a greater enlargement of the third than of the fourth ventricle is compatible with a CSF block not only intraventricular but also in the subarachnoid space. Nugent et al.[12] reported even communicating hydrocephalus may cause a secondary occlusion of aqueduct. AS cannot explain all the triventriculomegaly and its diagnosis should be decided quite deliberately.

Additionally, AS is sometimes difficult to diagnose by advanced technology such as anatomic MRI and phase-contrast flow study. Fukuhara and Luciano[5] reported nine patients among 26 patients had a positive flow in the aqueduct despite triventriculomegaly. With the result of seven successful ETV's out of these nine patients, it is suggested that positive flow does not necessarily exclude AS.

Kiefer et al.[8] refer to Oi's article saying that LOVA pa-

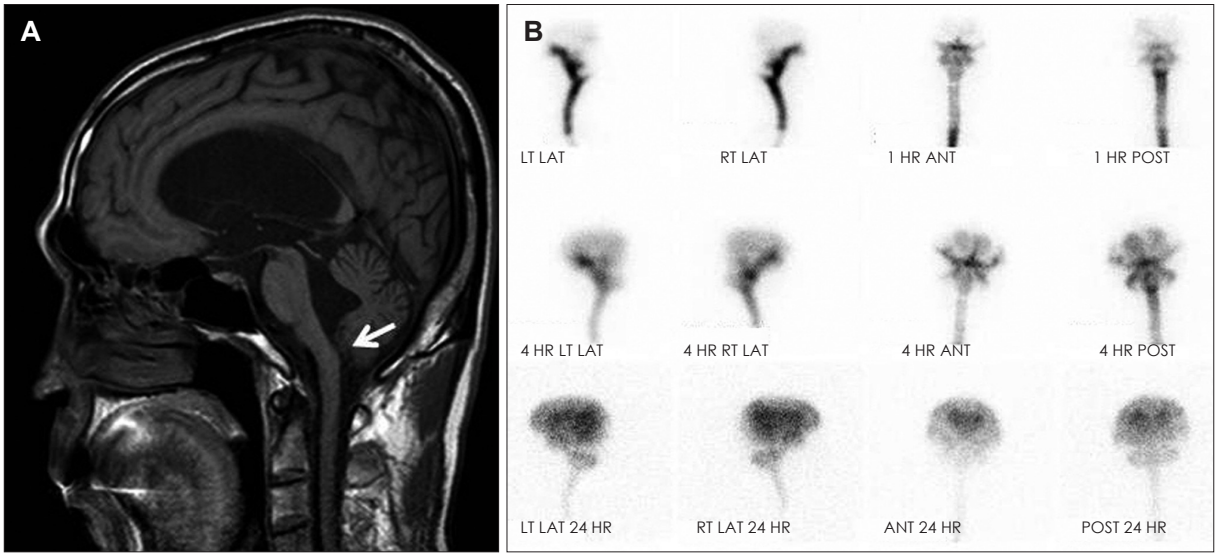

FIGURE 4. MRI showing ventriculomegaly and abrupt narrowing at the foramen of Magendie level (arrow) suggesting obstructive hydrocephalus (A). Radioisotope cisternography shows communicating type hydrocephalus (B).

tients are usually macrocephlic and diagnosis needs clinical or radiological signs of a long-standing increase in ICP. As Oi's study include three normocephalic and one unknown status patients as stated above, the authors questioned macrocephaly may not an absolutely necessary condition to diagnose this entity and tried to find the real incidence in this population. In similar context, Wilson and Williams[17] reported that a significantly larger proportion of patients with idiopathic normal pressure hydrocephalus (iNPH) have a head circumference greater than the 90th or 97th percentile than normal population. But they concluded that compensated congenital hydrocephalus is just one etiology of iNPH but not all.

Our cases make it clear that macrocephaly may not be an essential condition for diagnosis of LOVA and diagnostic criteria of this condition should be clarified more. With this finding and the past history of meningitis at three years old also suggest the onset age cannot be limited to infancy before closure of calvarial suture.

The term of LOVA, rather than SHYMA of LIAS, is used in this paper: there were patients in old age, and presence of AS is inconsistent in our series. But when using the term LOVA in a more widened meaning, not in the sense that the Oi et al.[13] previously defined but verbatim without mention of causality, AS as a sole causality and macrocephly as a diagnostic criteria should be reevaluated. As previously stated, definite AS was observed only in 1 case and other 2 cases had narrowings distal to the aqueduct itself in this study (Figure 3A, 4).

Sellar turcica widening means previous hydrocephalic process which was arrested or radiological evidence of long-standing increased ICP. Our cases showed sellar widening in all as expected.

Others reported chronic headache as a symptom of long-standing increased ICP.[4,15] But, there were only two symptomatic cases of long-standing increased ICP, and other cases were asymptomatic before activation.

CSF dynamics may change over time between active and inactive states but the reason for activation of once-arrested hydrocephalus remains unclear. A two-hit disease theory of Bradley may be applied to this condition.[1] In terms of case 1, EDH as a second hit does not cause activation because it might not cause the CSF blockage. The chronic increased ICP symptom was tried to be cured by shunt and effective for a long time in case 2, but further progression was supposed to be caused by CSF absorption disturbance due to omental adhesion and spinal problem with loss of compliance. SAH caused activation in case 3 and spinal hemorrhage activated the process in case 4, and shunt operation restablized the hydrocephalic process. The cause of aggravation in case 5 was not obvious. Considering case 2, case 4, and possibly case 3 the spinal lesion should be suspected as a possible disturbing source and evaluated accurately.

It remains to be determined how to monitor and decide when to operate patients with incidental, compensated hydrocephalus to prevent irreversible neurological deterioration while simultaneously avoiding premature or even unnecessary treatment. Opinions are divided on the matter of treatment. VPS with adjustable valve or ETV is generally selected. Some cases are very difficult to treat because of compliance loss or other reasons.[14,15] Some authors recommend third ventriculostomy as a treatment of choice.[5] But, the result of ETV was not satisfactory in other series.[15] If the initiating event causes blockage in the once-reestablished pathways such as subarachnoid space hemorrhage, it may be inappropriate to contemplate ETV in these patients. In these cases, VPS would be an appropriate selection. Inappropriate selection of treatment modality without deliberate consideration of relevant factors may lead to surgical failure or even deterioration.

The limitation of this study is small number of patients firstly. Secondly, detailed evaluation of cognitive function suggested by some authors was done only in two patients.[2,6,15] Thirdly, relevant studies were not sufficiently done such as cine phase-contrast MRI despite there is insufficient evidence to support the value in predicting response to shunting.

Conclusion

The relationships between macrocephaly, triventriculomegaly, and AS suggested in other studies were not consistent in this study. Dimensional and configurational changes of sella turcica were observed constantly. In contrast to the attributing AS itself as a sole causality, blockage or narrowing of CSF pathways were observed at various sites. Disturbances of spinal arachnoid pathways were related to the activation in some cases. A new event in the CNS may initiate activation of quiescent hydrocephalic process in some patients, therefore selection of treatment modality is to be tailored individually considering these factors. It is suggested that this entity is to be evaluated for better nomenclature reflecting diverse aspects of this condition. Further study is needed to elucidate underlying pathophysiology and effective management.

■ The authors have no financial conflicts of interest.

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