Delayed Detection of Hydrocephalus following Mildly Traumatic Subarachnoid Hemorrhage in Corticobasal Degeneration: A Case Report

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Keywords
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Abstract
A 65-year-old woman presented with slowly progressive aphasia with gait disturbance associated with parkinsonism. She experienced a fall that resulted in a brain trauma. Brain imaging revealed a small amount of subarachnoid hemorrhage (SAH) with intraventricular bleeding. Despite conservative therapy, gait disturbance and hyporesponsiveness gradually deteriorated following that brain trauma. One month later, she was transferred to our hospital, and magnetic resonance imaging revealed prominent communicating hydrocephalus. A ventriculoperitoneal shunt and brain biopsy were performed. Neurosurgical intervention did not improve the patient’s neurological condition. Clinical-pathological analysis confirmed the diagnosis of corticobasal degeneration (CBD) as an underlying disease relating to parkinsonism and aphasia. In patients with parkinsonism with high risks of falling, attention should be paid to neurological deterioration due to traumatic SAH-related hydrocephalus. Particularly, in
patients with aphasia such as in those with CBD, delayed detection of posttraumatic complications might cause poor responsiveness to surgical intervention.

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Introduction

Due to extension of the life expectancy, neurodegenerative diseases presenting with parkinsonism have become a global concern. Therefore, raising awareness of complications of these diseases is needed. Corticobasal degeneration (CBD) is a rare neurodegenerative disease classified as a tauopathy [1]. Patients with CBD show various neurological symptoms such as parkinsonism, apraxia, aphasia, and cognitive impairment [1]. Similar to patients with other parkinsonisms, such as Parkinson's disease, progressive supranuclear palsy, and multisystem atrophy, patients with CBD are at a high risk of falling [2, 3], which may cause brain trauma.

Patients with brain trauma often experience traumatic subarachnoid hemorrhage (SAH) that occasionally induces hydrocephalus, deteriorating neurological functions [4, 5]. Patients with CBD might be at a high risk of traumatic SAH-induced hydrocephalus due to frequent falls. However, to the best of our knowledge, no studies have investigated traumatic SAH-related hydrocephalus in patients with parkinsonism, including CBD. Herein, we describe the first case of a patient with CBD who developed hydrocephalus following a fall with mildly traumatic SAH and exhibited subacute neurological deterioration. This case highlights caution for traumatic SAH-related hydrocephalus in patients with parkinsonism with a high risk of falling and a risk of delayed detection of posttraumatic complications, especially in those with aphasia such as CBD.

Case Presentation

A 65-year-old Japanese woman developed difficulty in outputting words when she was 60 years old. Motor aphasic symptoms gradually progressed. The patient showed difficulty in understanding conversations at the age of 64 years. She visited a clinic where haloperidol and biperiden were administered. Around the same time, she developed gait disturbance. Three months after starting her course of treatment, she experienced a fall that resulted in brain trauma. The following day, the patient experienced a repeated fall. Two days after the second fall, the patient was admitted to a hospital for gait disturbance and repeated falls. On neurological examination, her Glasgow Coma Scale (GCS) score was E4VAM6. She showed signs of parkinsonism such as akinesia, axial and left-side predominant limb rigidity, and postural disturbance. Brain computed tomography revealed mild intraventricular hemorrhage with subdural hygroma (shown in Fig. 1a). The susceptibility-weighted image of brain magnetic resonance imaging detected mild SAH with intraventricular hemorrhage (shown in Fig. 1b, c). Ventricular size was normal (shown in Fig. 1d). No aneurysm was detected on magnetic resonance angiography. These findings of brain imaging suggested that her brain trauma was mild. However, gait disturbance and hyporesponsiveness gradually exacerbated despite the discontinuation of antipsychotic drugs and initiation of rehabilitation. One month later, she was transferred to our hospital.

On admission, a physical examination revealed no abnormal findings. On neurological examination, the patient showed severe motor aphasia, and her GCS score was E4VAM5.
She was unable to sufficiently comply with verbal instructions. The patient showed mild limitation of vertical eye movement. Paralysis was not apparent. Parkinsonism, such as limb and trunk muscle stiffness and akinesia, was detected. Pyramidal signs such as hyperreflexia and pathological reflexes with a positive grasp reflex were apparent. She was unable to stand and walk. Blood test results revealed no abnormalities, including blood count, electrolytes, glucose levels, and thyroid function. Tests for whole-blood tuberculosis-specific interferon-gamma assay and serum cryptococcal antigen were negative. The appearance of cerebrospinal fluid (CSF) was clear, with an opening pressure of 135 mm H2O. Mildly elevated levels of CSF protein (54 mg/dL) and cell count (10 cells/μL with 100% mononuclear cells) were detected along with normal glucose levels (69 mg/dL). Cultures of bacteria, fungi, and acid-fast bacilli in the CSF were negative. Additionally, CSF cytology was classified as class I. Electroencephalogram showed diffuse slow waves without paroxysmal activity. Brain magnetic resonance imaging revealed prominent communicating hydrocephalus (shown in Fig. 1e). Conservative treatment with oral L-DOPA at a maximum dose of 450 mg/day did not improve the patient's neurological condition. A ventriculoperitoneal (VP) shunt was performed on day 23 after admission to our hospital. To make a histopathological diagnosis of underlying diseases causing aphasia and parkinsonism, brain biopsy from the right parietal lobe at a site inserting a VP shunt tube was performed after obtaining an informed consent from her family. The size of the ventricle

Fig. 1. Brain images. Brain CT 3 days after brain trauma. Arrowheads show mild intraventricular hemorrhage (a). SWI of brain MRI 5 days after brain trauma reveals mild SAH with intraventricular hemorrhage (b). Arrows show a small amount of SAH around the Sylvian fissure (c). FLAIR sequence image detects normal ventricular size (d). FLAIR sequence image 33 days after brain trauma shows a prominent enlargement of the lateral ventricles and narrowing of the subarachnoid space (e). Brain CT 20 days after VP shunt (f). CT, computed tomography; FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging; SAH, subarachnoid hemorrhage; SWI, susceptibility-weighted image; T2WI, T2-weighted imaging; VP, ventriculoperitoneal.
was reduced following the operation (shown in Fig. 1f). However, the patient’s neurological condition did not change remarkably. Brain pathology showed ballooned neurons on hematoxylin-eosin staining (shown in Fig. 2a). Neuropil threads (shown in Fig. 2b) and coiled bodies (shown in Fig. 2c) were detected by Gallyas staining. Phosphorylated-tau immunostaining showed positivity for the neuropil threads and the cytoplasm of some neurons (shown in Fig. 2d). No astrocytic plaques were detected in the biopsy specimens. These pathological findings confirmed the diagnosis of CBD as an underlying disease presenting with aphasia and parkinsonism. She was transferred to another hospital and moved to a long-term care facility. Six months after the VP shunt placement, her neurological condition did not improve.

**Discussion/Conclusion**

In our patient, gait disturbance and hyporesponsiveness exacerbated following brain trauma. In addition, she was not able to stand and walk within 4 months after the onset of gait disturbances. Furthermore, her neurological findings did not improve after more than a half year despite discontinuation of antipsychotic drugs. These are signs of atypical clinical course of neurodegenerative diseases and drug-induced parkinsonism only. In our patient, traumatic SAH-related hydrocephalus would be mainly associated with subacute neurological deterioration following a fall causing brain trauma.

Posttraumatic hydrocephalus is an important complication in patients with brain trauma [5]. Compared with patients without traumatic SAH, those with traumatic SAH have a higher
risk of developing posttraumatic hydrocephalus [5]. The pathophysiology of hydrocephalus following SAH is suggested to be ascribed to blood clots disturbing CSF flow and inflammation-mediated adhesion interfering with CSF absorption at the arachnoid granulation [6]. A rabbit model of SAH showed an increased number of water vesicles at the choroid plexus in the early phase following autologous blood injection in the cisterna magna. Consequently, this might increase CSF production and thereby might induce hydrocephalus [7, 8]. Hydrocephalus usually develops within 3 months of traumatic SAH [4, 5]. Tian et al. [4] reported that increasing age, intraventricular hemorrhage, diffuse distribution, or thickness more than 5 mm of traumatic SAH were risk factors for developing hydrocephalus in patients with traumatic SAH. Among these risk factors, intraventricular hemorrhage showed the highest odds ratio value of traumatic SAH-related hydrocephalus. Similar to these reports, hydrocephalus developed within 1 month after the onset of traumatic SAH with intraventricular hemorrhage in our patient. On the other hand, traumatic SAH in our patient was focal and mild, which caused delayed detection of traumatic SAH-related hydrocephalus.

Patients with CBD frequently experience recurrent falls [2, 3], which may cause traumatic complications that induce neurological deterioration. In our study, traumatic SAH-related hydrocephalus deteriorated the patient’s neurological symptoms, and the VP shunt did not improve her condition. In patients with traumatic SAH-related hydrocephalus, whether underlying parkinsonism is a risk factor for poor responsiveness to VP shunt remains unclear. In contrast, underlying neurodegenerative disorders with parkinsonism and dementia are negative prognostic factors for outcomes in patients with subdural hematoma, a common complication of brain trauma [9]. Underlying parkinsonism might also be a risk factor for poor response to VP shunts in patients with traumatic SAH-related hydrocephalus. Furthermore, the relatively long duration between the onset of traumatic SAH-related hydrocephalus and neurosurgical intervention in our patient may be linked to the poor responsiveness to the VP shunt. Underlying parkinsonism and aphasia may cause delayed detection of brain trauma-related complications, including traumatic SAH-induced hydrocephalus.

In conclusion, herein, we describe a patient with CBD who had deteriorated neurological function with hydrocephalus following mildly traumatic SAH with intraventricular hemorrhage. In patients with parkinsonism, with high risk of repeated falls, we need to pay attention to brain trauma to prevent neurological exacerbation of traumatic SAH-related hydrocephalus even in patients with mild traumatic SAH. Particularly, in patients with CBD, not only parkinsonism but also aphasia may cause delayed detection of brain trauma-related complications.

Statement of Ethics

Ethical approval is not required for this study in accordance with local guidelines. Written informed consent was obtained from the patient’s next of kin for publication of the details of their medical case and any accompanying images because the patient showed aphasia.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Yusuke Mochizuki contributed to collecting data. Yusuke Mochizuki and Minori Kodaira contributed to drafting the manuscript. Mitsunori Yamada and Midori Sato contributed to analyzing pathological findings. Tomoki Kaneko contributed to analyzing radiological findings. Yusuke Mochizuki, Minori Kodaira, Yasufumi Kondo, Mitsunori Yamada, Masafumi Kuroiwa, Tomoki Kaneko, Midori Sato, and Yoshiki Sekijima contributed to interpreting data and revising the manuscript.

Data Availability Statement

All data of this study are included in this article. Further inquiries can be directed to the corresponding author.

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