Frontotemporal Brain Sagging Syndrome as a Treatable Cause Mimicking Frontotemporal Dementia: A Case Report

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Keywords
Apathy · Frontotemporal dementia · Intracranial hypotension · Blood patch

Abstract
Frontotemporal brain sagging syndrome (FBSS) is a progressive disorder characterized by symptoms similar to the behavioral variant of frontotemporal dementia (FTD), with a sagging appearance of the brain on imaging similar to that observed in spontaneous intracranial hypotension (SIH). The onset of behavioral and cognitive symptoms of FBSS is insidious and progressive, similar to those of FTD. Here, we report a case involving a 53-year-old man with progressive hypersomnia, apathy, forgetfulness, and personality changes but without headache or auditory symptoms. The combination of frontotemporal dysfunction, hypersomnia, and the appearance of a sagging brain on magnetic resonance imaging suggested a diagnosis of FBSS. Although a definite site of cerebrospinal fluid leakage could not be identified in our case, clinical symptoms and imaging findings were improved after an epidural blood patch. Considering FBSS as a differential diagnosis of FTD is important even in the absence of typical SIH symptoms, such as headache or auditory symptoms.
Introduction

Frontotemporal brain sagging syndrome (FBSS), a progressive disorder characterized by apathy, behavior, and cognitive dysfunction, hypersomnolence, and orthostatic headache, is diagnosed via a sagging appearance of the brain on imaging, similar to that observed in spontaneous intracranial hypotension (SIH) [1, 2]. Studies have shown that FBSS has an insidious onset, slow progressive course, and symptoms similar to that of a behavioral variant of frontotemporal dementia (FTD); however, it is a potentially treatable disease [1, 2]. Therefore, early diagnosis, treatment, and appropriate neuropsychological assessment for the evaluation of treatment efficacy are important. Here, we report a case of FBSS in which the clinical symptoms and imaging findings were improved after an epidural blood patch.

Case Report

A 53-year-old Japanese man with no previous medical history presented to our hospital for the evaluation of progressive apathy, forgetfulness, and personality changes. Approximately 3 years before evaluation, his family members noticed that he went out less frequently and had daytime hypersomnolence. After retiring from his job as a driver at the age of 51, he had become increasingly indifferent, less interested in the activities he had previously enjoyed, and easily provoked by normal conversations. Additionally, he frequently lost his temper with his family and exhibited symptoms of forgetfulness, highlighted by repetitive questioning and missing appointments. He did not report anxiety, depression, auditory symptoms, or headaches.

On examination, the patient was cooperative but exhibited mild disinhibition, such as speaking loudly in a quiet waiting room, joking, and showing garrulousness during neuropsychological tests. Addenbrooke’s Cognitive Examination III revealed an overall score of 87 (out of a total of 100), with subscale scores of 16/18 for attention and orientation, 20/26 for memory, 9/14 for fluency, 26/26 for language, and 16/16 for visuospatial skills. On the Wechsler Memory Scale-Revised, his general memory, verbal memory, visual memory, attention/concentration, and delayed recall scores were 75, 71, 96, 100, and 61, respectively. His frontal assessment battery (FAB) score was 15 out of 18, whereas his score for the Japanese version of the Apathy Scale [3] was 22 out of 42 (where a score of ≥16 reflects apathy [4]). The patient displayed no frontal release signs; intact cranial nerves; normal motor, reflex, sensory, and gait examinations; and no cerebellar signs. Blood work, including a complete blood cell count, electrolyte and liver enzyme levels, renal and thyroid function, and vitamin B1, vitamin B12, and folate levels were all normal. Results of syphilis screening and an HIV test were normal. Brain magnetic resonance imaging (MRI) showed severe sagging of the brain with a downward displacement of the cerebellar tonsils, downward drooping splenium, collapsed widened midbrain, and flattened ventral pons with effacement of the basal cisterns (Fig. 1). Gadolinium-enhanced images demonstrated intense pachymeningeal enhancement and subdural fluid collections with the enlargement of the venous sinus and pituitary gland (Fig. 1). The combination of frontotemporal dysfunction, hypersomnolence, and the appearance of a sagging brain on MRI suggested a diagnosis of FBSS.

MRI of the whole spine and computed tomography myelography revealed no cerebrospinal fluid (CSF) leakage. Although the neurological symptoms had reached a plateau phase after visiting our hospital, no improvement was noted. Given that the definite site of CSF leakage was unknown, an epidural blood patch was performed at the lumbar level. Autologous blood was injected into the epidural space at the L2–L3 level using a Tuohy needle. After infusing 29 mL of autologous blood, the patient complained of discomfort, which prompted the stoppage of the procedure. After the procedure, his symptoms, such as daytime hypersomnolence, apathy,
disinhibition, and forgetfulness gradually improved, allowing him to return to work. After 8
months of the epidural blood patch, his FAB score improved to 17, whereas his score on the
Japanese version of the Apathy Scale improved to 12. Follow-up brain MRI at 11 months after
the epidural blood patch showed improvement in the sagging appearance of the brainstem, less
venous sinus congestion, and less pachymeningeal enhancement (Fig. 2). The patient declined
to undergo further neuropsychological tests and repeated MRI.

Discussion/Conclusion

The patient presented to our institute with daytime somnolence, apathy, disinhibition,
and mild cognitive impairment but lacked symptoms of SIH, such as headache or auditory
symptoms. Although a definite site of CSF leakage could not be identified, clinical symptoms
and imaging findings had improved after an epidural blood patch.
Despite being a treatable disease, FBSS can present with FTD-like symptoms, including apathy and behavioral and cognitive symptoms, which suggested the importance of assessing the type of CSF leakage to determine the appropriate treatment strategy. Generally, FBSS has been attributed to a spontaneous leakage of CSF, with treatment options, depending on the type of CSF leakage [2, 5]. Therefore, the location and type of CSF leakage must be identified through testing, which includes whole spinal imaging. Similar to the current patient, a definite site of CSF leakage in patients with FBSS is often unidentified on multimodality spinal imaging [1, 2]. When no clear source of CSF leakage can be identified, treatment options include steroid administration and epidural blood patches [5, 6]. If neither of these is ineffective nor has only a transient effect, surgical procedures, such as dural reduction or placement of a wearable epidural infusion system, might be considered [5, 6]. Although digital subtraction myelography, not performed in our patient, has been reported to be useful for detecting CSF–venous fistulas when no CSF leakage is identifiable on conventional spinal imaging [7]. A case of FBSS with inferior vena cava obstruction that resulted in craniospinal hypovolemia via CSF–venous fistula has also been reported [8]. After an epidural blood patch, our patient demonstrated improvement in his clinical problems.
symptoms and imaging findings. However, given that the clinical nadir of neurological symptoms was reached and followed by a plateau phase before the treatment and that the improvements after treatment were exceedingly gradual, we cannot exclude the possibility that the latent improvement occurred spontaneously. Cases with spontaneous resolution of FBSS have been reported and spontaneous resolution of CSF leakage and “auto-blood patch” resulting from lumbar puncture or trauma have been postulated as mechanisms for spontaneous improvement in FBSS [9, 10].

The lack of a headache does not rule out the possibility of FBSS. The typical clinical course of FBSS is preceded by symptoms of SIH followed by symptoms of frontotemporal dysfunction [2]. However, our patient presented with no symptoms of SIH, such as a headache or auditory symptoms. Although the reason remains unclear, reports have shown that some patients with FBSS lack symptoms of SIH, similar to our case [2, 11].

FBSS can be characterized by its insidious onset, slowly progressive course, and symptoms similar to that of a behavioral variant of FTD. However, it is a potentially treatable disease. Thus, it is important to consider FBSS as a differential diagnosis of FTD despite the lack of typical SIH symptoms, such as headache or auditory symptoms.

Statement of Ethics

This study was conducted in line with the principles of the Declaration of Helsinki. Written informed consent was obtained from the patient for the submission and publication of this case report and accompanying images. This retrospective review of patient data did not require ethical approval in accordance with local guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

The authors did not receive any funding relevant to this study.

Author Contributions

Atsuhiko Sugiyama: reviewed the literature, obtained patient consent, and drafted the case report; Ado Tamiya: critically reviewed the manuscript; Ryo Otani: critically reviewed the manuscript; Hajime Yokota: critically reviewed the manuscript and performed radiological assessment; Hiroki Mukai: critically reviewed the manuscript and performed radiological assessment; Satoshi Kuwabara: critically reviewed and edited the manuscript and approved the final version for publication.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.
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