Neurological Sciences (2019) 40:875–877
https://doi.org/10.1007/s10072-018-3636-3

LETTER TO THE EDITOR

Sheehan syndrome mimicking dementia with Lewy bodies

Sang-Won Yoo[1] · Sung-Jin Park[1] · Joong-Seok Kim[1]

Received: 27 July 2018 / Accepted: 3 November 2018 / Published online: 13 November 2018

Dear Editor,

Sheehan syndrome refers to postpartum hypopituitarism caused by ischemic necrosis due to blood loss and hypovolemia during and after delivery [1]. The signs and symptoms of panhypopituitarism are diverse due to the involvement of various hormonal insufficiencies [1]. The most common hormones lost are growth hormone (GH), followed by gonadotropins, adrenocorticotropic hormone, and thyroid-stimulating hormone.

While secondary parkinsonism and neurobehavioral symptoms are well-recognized conditions associated with hormonal dysfunction, especially thyroid and adrenal gland dysfunction [2, 3]; clinical presentation as dementia with Lewy bodies (DLB) associated with Sheehan syndrome has not been reported.

We describe a patient with Sheehan syndrome who complained of vivid, recurrent hallucinations, fluctuating cognition, slurred speech, and bradykinesia.

A 62-year-old woman was admitted with vivid hallucinations, fluctuating cognition and alertness, slurred speech, and bradykinesia. Two years before her admission, the patient began to display intermittent sudden change of attention, waxing and waning episodes of abnormal behavior, and incoherent speech. Over the previous 2 months, she experienced recurrent visual hallucinations more than 10 times per day. Each event appeared abruptly and lasted for several minutes. The hallucinations were well formed, featuring people. She complained of seeing someone performing an exorcism. They occurred mostly at night but also during the day. She sometimes uttered something unintelligible to herself. She also had fluctuating alertness, manifesting as excessive daytime somnolence. She complained of depressed mood and anxiety. During the same period, the patient had slow movement and decreased agility in performing daily activities. She had no personal or family history of psychiatric or neurological diseases such as central nervous system infection, epilepsy, schizophrenia, depression, parkinsonism, or hepatic encephalopathy. She was never exposed to harmful environmental toxins.

Twenty-five years prior, she had experienced massive bleeding during childbirth and shortly thereafter experienced mental confusion due to hyponatremia. The patient was diagnosed with Sheehan syndrome. Her symptoms were mainly amenorrhea and pubic hair loss due to gonadotrophin disturbance. The patient received hormone replacement therapy, and her symptoms were well controlled for 10 years. Fifteen years prior, she acknowledged a harmful effect of long-term steroid treatment and discontinued hormone replacement without medical consultation. She intermittently took thyroxine and corticosteroid when she felt depressed and lacked motivation. She also complained of orthostatic intolerance and chronic constipation.

The patient was afebrile on arrival. On examination, she was oriented and scored a 21 out of 30 on the Korean version of the Mini-Mental State Examination test. A neuropsychological investigation showed disproportionate executive dysfunction, visuospatial and perceptual deficits relative to memory, and preserved language function. She also manifested multiple neurobehavioral symptoms and scored a 57 out of 144 on the Neuropsychiatric Inventory. The patient exhibited severe asymmetric akinetic rigidity on the left side. Postural reflex was also impaired. There were no signs of meningeal irritation and raised intracranial pressure.

Blood tests, including hepatic and renal functions, were unremarkable. However, her basic levels of thyroid hormone, GH, prolactin, follicle-stimulating hormone (FSH), luteinizing hormone (LH), and cortisol were below physiologic levels; free T4 0.27 ng/dL, insulin-like growth factor-1 10.55 ng/mL, prolactin 1.47 ng/mL, FSH 3.07 mIU/mL, LH 1.33 mIU/mL, and cortisol 1.21 μg/dL. A combined anterior pituitary stimulation test with a triple bolus injection of insulin, thyroid-releasing hormone, and gonadotropin-releasing hormone was performed

✉ Joong-Seok Kim
neuronet@catholic.ac.kr

[1] Department of Neurology, College of Medicine, The Catholic University of Korea, Seoul St. Mary's Hospital, 222 Banpo-daero, Seocho-gu, Seoul 06591, Republic of Korea

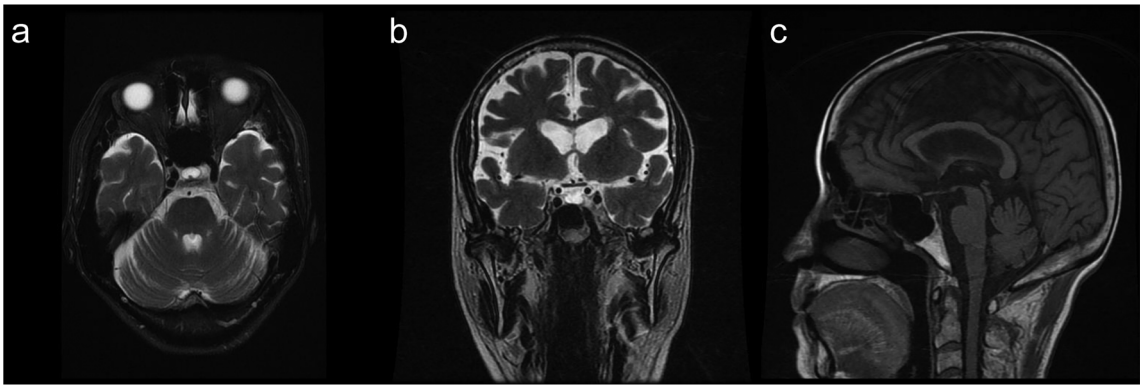

Fig. 1 Axial (**a**) and coronal (**b**) T2-weighted and sagittal (**c**) T1-weighted magnetic resonance images of the brain showed an empty sella turcica

and revealed that LH, FSH, prolactin, cortisol, and GH did not rise above intended level or showed any increasing trends when measured every 30 min for two hours. The results were compatible with panhypopituitarism.

Electroencephalography did not reveal any epileptic activities, excluding epileptic encephalopathy. Brain magnetic imaging revealed an empty sella turcica (Fig. 1), which can be found in chronic Sheehan syndrome. There were no structural changes that might provoke neurobehavioral manifestations. To exclude other neurodegenerative pathologies, we performed positron emission tomography using ^{18}F-N-(3-fluoropropyl)-2beta-carbon ethoxy-3beta-(4-iodophenyl) nortropane and ^{18}F-florbetaben, which showed normal presynaptic dopamine uptake and lack of amyloid accumulation in cortices, respectively (Fig. 2).

After prescribing 100 mcg of thyroxine and 15 mg of hydrocortisone, her fluctuating psychosis and cognition, vivid hallucination, parkinsonism, and orthostatic intolerance were resolved.

The patient had a fluctuating cognitive impairment and recurrent, well-formed visual hallucinations, parkinsonism, and other features, such as depression, orthostatic intolerance, and constipation. These prominent features, in combination with other suggestive and supportive abnormalities, suggested a clinical diagnosis of DLB.

In this patient, we checked accompanying brain pathologies with dopamine transporter and amyloid scans. In addition, we clinically excluded other etiologies such as Hashimoto's encephalopathy and other autoimmune encephalitis based on chronicity and fluctuation of her symptoms. Patient's cognitive and neurobehavioral symptoms were completely responded to hormones replacement. Resolutions of her symptoms after simple hormones replacement excused further unnecessary expensive investigations and we can confirm that many of her symptoms and signs could be assigned to a lack of physiologic hormones [1]. Parkinsonism [2, 3] and psychosis [4, 5] due to panhypopituitarism have been reported, and her seemingly dysautonomic symptoms of orthostatic

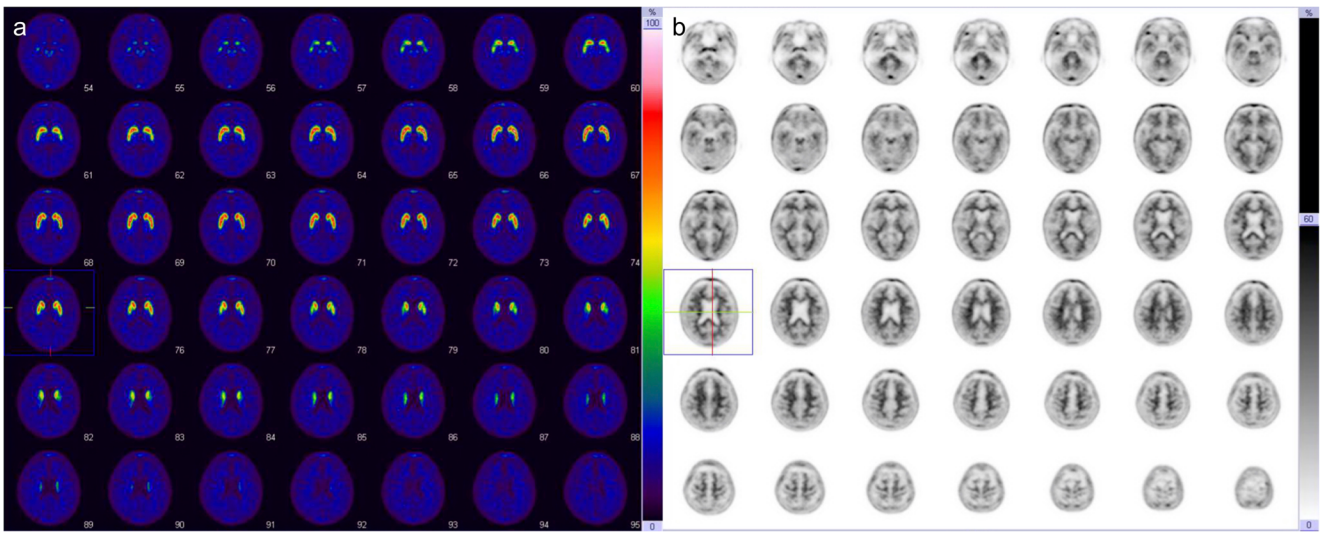

Fig. 2 Positron emission tomography using ^{18}F-N-(3-fluoropropyl)-2beta-carbon ethoxy-3beta-(4-iodophenyl) nortropane (**a**) and ^{18}F-florbetaben (**b**) demonstrated normal presynaptic dopaminergic transporter uptake and no deposition of the radioligand in the bilateral cerebral cortex, indicating no beta-amyloid deposition

intolerance and constipation can be explained by osmotic dysregulation and hypothyroidism.

Neurobehavioral and cognitive disturbances are known symptoms of panhypopituitarism, regardless of sex [5, 6]. Psychiatric symptoms of panhypopituitarism encompass hypo- and hyperactive neurobehaviors and have no specific pattern [4–6]. The exact mechanism of this manifestation is still poorly understood, but disturbed physiologic interactions between pituitary hormones and dominant neurotransmitters such as dopamine, serotonin, γ-aminobutyric acid, and glutamate have been suggested to be the cause [4].

The hypothesis that most intrigued us in explaining our patient's DLB-like symptoms was the role of thyroid hormone, which modulates important neurotransmitters [7]. Our patient's improvement after continuous stable thyroxine supplementation further substantiates its role.

Our case differs from previous reports in that her progressive cognitive impairment was concurrent with fluctuating attention, vivid hallucinations, and parkinsonism that mimicked DLB. To our knowledge, no report of DLB-like panhypopituitarism has been published. This is the first report that extensively searched for neurodegenerative pathologies and excluded synucleinopathy and amyloidopathy that could confound the clinical effects of hypopituitarism.

Funding This research was supported by Basic Science Research Program through the National Research Foundation of Korea (NRF) funded by the Ministry of Science, ICT, and Future Planning (NRF-2017R1D1A1B06028086).

Compliance with ethical standards

Ethics statement The institutional review board at St. Mary's Hospital approved this case report.

Conflict of interest The authors declare that they have no conflicts of interest.

Patient consent The patient has consented to the submission of the case report to the journal.

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