A case of semantic dementia

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Abstract

We are reporting a female case of semantic dementia. Characteristic of this type of dementia, she could not recognize some objects such as a pencil, a ruler or her dominant hand. When shown a pencil, she would ask "What is a pencil?" Her brain MRI showed atrophy of bilateral temporal lobes, especially anterior poles. SPECT also showed abnormally decreased blood flow in temporal lobes and frontal lobes. She has been taking galantamine for three years without any side effects. Her ability to speak fluently and the number of objects that she cannot recognize have been almost consistent for three years. Galantamine might be beneficial towards speaking fluency and object recognition.

Keywords: semantic dementia, semantic PPA, FTD, Alzheimer's diseases, galantamine

Care Report

The patient is a seventy-four year old housewife. The chief complaints were forgetfulness and irritability. Her family history and past history showed nothing in particular. She showed forgetfulness, she bought the same things again and again, forgot how to make some dishes and could not recognize the names of some things. Her husband noticed a change in her temper and in her psychological state. Her neurological status showed nothing in particular but showed decreased MMSE score (15/30; 1:1/5, 2:1/5, 3:3/3, 4:1/5, 5:0/3, 6:2/2, 7:1/1, 8:3/3, 9:1/1, 10:1/1, 11:1/1). Her ability to communicate fluently with her husband was not impaired, however she lost the ability to recognize or identify items such as a pencil, ruler or dominant hand. This inability to identify objects was seen in our clinic. When she was shown a pencil, she claimed that she had never seen a pencil before and asked what it was, and how it was used. She suddenly stood up and had to be prevented from leaving. After our clinic, she was given a WAIS test by a speech therapist. She could not finish the test because she became angry and suddenly left the examination room. When presented with a question that she was unable to answer, she would repeatedly claim that being unable to answer the questions was natural, given her age.She asked me what disturbance she had in her brain. She did not have insight into her cognitive condition. No abnormality was seen in her blood test, chest X-ray and ECG.

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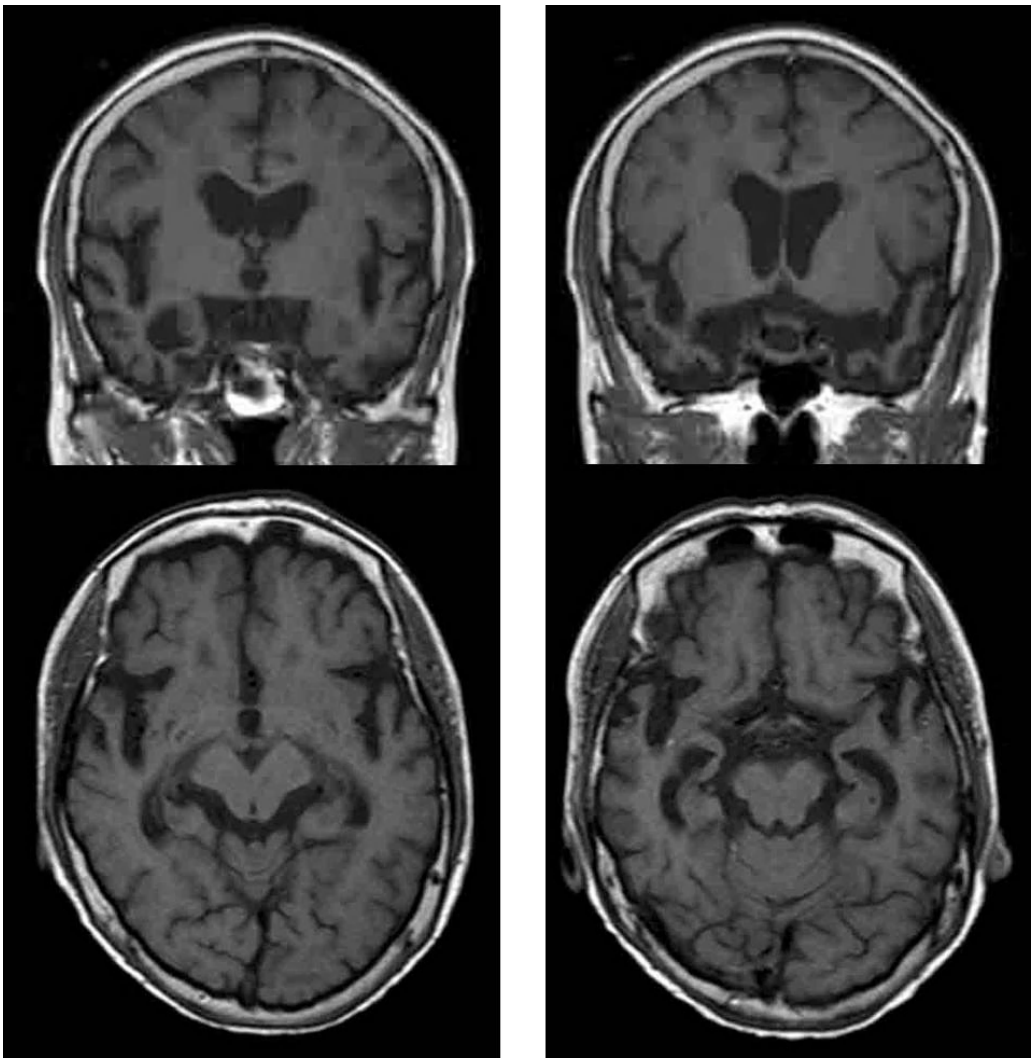

Figure 1. Brain MRI images
Upper is coronal image and lower is axial image Bilateral temporal lobes especially anterior poles showed markedly atrophy and para-hippocampal areas also showed atrophy.

Her brain MRI showed marked atrophy in bilateral temporal lobes, especially in anterior poles and also bilateral parahippocampal areas (Figure 1). Her brain blood flow SPECT showed apparently decreased blood flow rate in bilateral frontal and temporal lobes (Figure 2). I diagnosed her as having the semantic variant PPA of frontotemporal dementia. I prescribed her galantamine 4mg twice a day, after each morning and evening meal. After four weeks, the doses of the drug were increased to 8mg twice a day.

Clinical course of the patient

When the patient was examined three years later, we found that her memory had continued to decline, and she had become unable to recognize the faces of people she did not see every day, such as neighbors. Her irritability gradually disappeared and her speech remained stable. The number of things that she cannot recognize is almost stable. She shows a stubborn attitude in our clinic and continues to blame her age as the reason for her memory loss.

Discussion

Semantic dementia has been classified as

semantic variant PPA (primary progressive aphasia) of FrontoTemporal Dementia (FTD) in 2011 [1]. The hallmark of semantic PPA is difficulty identifying or recognizing familiar objects. For example, when a patient is shown a pencil, he can neither name it nor can he recognize the word when it is provided. The patient characteristically asks "What is a pencil?" when it comes up in conversation or during testing. This happens for rare words at first and common nouns during later stages. Fluent spontaneous speech is retained. Some patients have problems recognizing familiar objects and faces. In later stages, patients with semantic PPA may show behavioral abnormalities of FTD [2]. This patient has showed difficulty in identifying some familiar things, like a pencil or a ruler. When she was asked which of her hands was her dominant hand, she could not understand the question and asked to have it explained to her. She could speak fluently and we were able to communicate easily. Her ability to recognize faces has continued to decline, and she can no longer recognize her neighbors. She continues to present a stubborn attitude while in our clinic. MRI and blood flow SPECT examinations showed typical changes as semantic PPA, which was marked atrophy of anterior poles of temporal lobes [1] (Figure 1 and 2).As with all forms of FTD, there is no cure for semantic PPA, and in most cases its progression cannot be slowed. Although no medications have been proven effective specifically in FTD, many clinicians look to the medications and treatment approaches targeting behavioral disturbances as necessary. For instance, some FTD patients benefit from selective serotonin reuptake inhibitors [2] (SSRIs, used in treating obsessive-compulsive behaviors and irritability). Kertesz et al. did a clinical trial of galantamine for the patients with bvFTD (behavioral variety) and PPA. Galantamine is not effective in the behavioral variety of FTD, but a trend of efficacy is shown in the aphasic subgroup, which may be clinically significant. Galantamine appeared safe in FTD/PPA [3]. Galantamine has two actions, one is as anti-acetylcholine esterase inhibitor and the other is as allosteric potentiating ligand [4]. The latter action can stimulate allosteric acetylcholine receptors of monoamine neurons and can make the neurons release monoamines, like noradrenaline, dopamine or serotonin [4,5]. We prescribed galantamine to her in expectation of its anti-choline esterase action and the other action as allosteric potentiating ligand. After only three years of using the drug, her irritability gradually improved and other behavioral and social difficulties have not become evident. She can still speak fluently and her inability to recognize objects has not worsened. We do not have the evidence that galantamine may be a beneficial drug for semantic PPA patients. In this case, we cannot exclude the possibility that her clinical course would have been the same even without the drug. The clinical trial for many patients with semantic PPA should be done in order to have good evidence of the efficacy of the drug. She made excuses for her inability to answer questions. This kind of behavior is often seen in Alzheimer's disease [6]. Although the results of her MRI and SPECT showed that she apparently had FTD, we could not exclude the possibility that she could be suffering from Alzheimer's disease. Her clinical phenotype is apparently semantic PPA of FTD but neuropathological diagnosis remains unknown. We reported a case clinically diagnosed as having semantic PPA. Galantamine might have been beneficial in improving her irritability and to retain the ability of recognizing objects and of fluent speaking.

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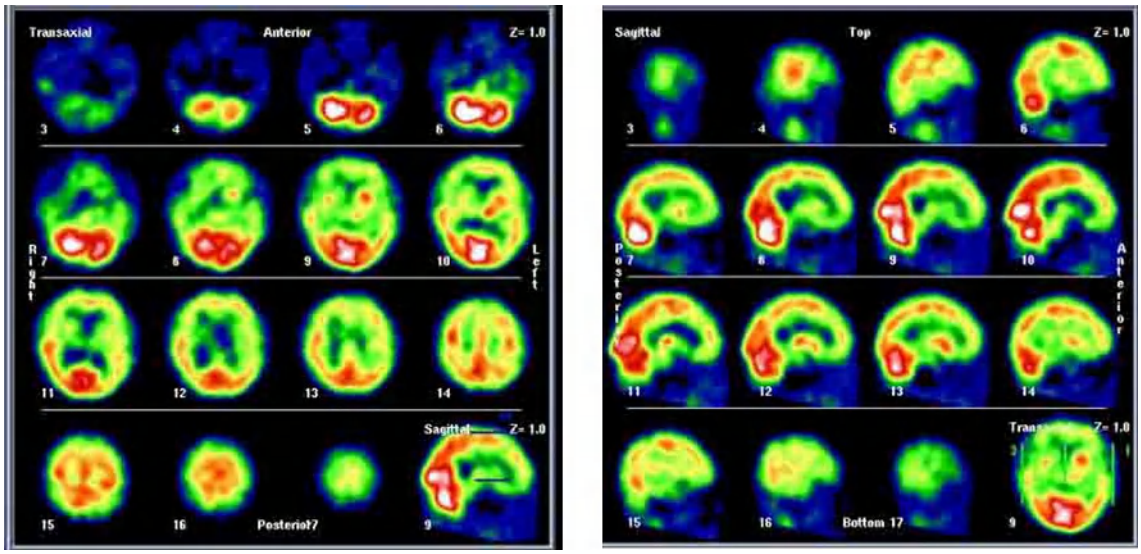

Figure 2. Brain blood flow SPECT
Bilateral frontal lobes and temporal lobes showed low blood flow rate.