A Case of Dementia with Atrophy of the Ambient Gyrus

Key words: ambient gyrus, argyrophilic grain dementia, easy excitability, amnesia, SPECT

Argyrophilic grain dementia (AGD) or dementia with grains was defined by Braak and Braak as a form of senile dementia that only demonstrates argyrophilic grains as the morphological cause of dementia (1, 2). Argyrophilic grains are immunoreactive with anti-tau antibodies, and AGD may be classified as a form of tauopathy. Multiple studies support the differentiation of AGD from Alzheimer’s disease (AD). Despite genotypic and immunohistochemical differences, the clinical presentation of AGD largely overlaps that of AD, such that the clinical distinction between the two is difficult.

In a study from a community-based geriatric hospital, characteristic cases of AGD demonstrated severe atrophy of the ambient gyrus (the junction between the temporal lobe and amygdala) with spongiosis, neuronal loss, and gliosis, as well as many grains, pretangles, coiled bodies, and tau-immunoreactive astrocytes (3). The temporal neocortex and hippocampus are relatively spared in AGD, in contrast to those in Alzheimer’s disease. That study suggests that selective severe involvement of the ambient gyrus may explain the clinical manifestations of a limbic-type dementia in AGD. I recently encountered a man with amnesia, easy excitability, hyperreflexia of the lower extremities, difficulty in calculation, disorientation, predominant atrophy of ambient gyri and dilatation of the inferior horns of lateral ventricles.

A 76-year-old man had a 10-year history of dementia. About 10 years earlier, he became angry only at home when his wife with a hearing difficulty would ask him to repeat his comment once more. He was easily excitable at home but remained gentle in front of others. About 5 years previously, he forgot that he had lit a cigarette. Also, he yelled loudly “Deruna (Don’t appear)” when a person he disliked appeared on television. About 3 years ago, he presented with weakness of the left lower extremity. On March 19, he complained of low back pain. Thereafter, he presented with anorexia and only ate pudding and yogurt. By April 1, he only drank liquids. On April 11, he was admitted to this hospital.

When he was 19 years old, he had pleuritis. Both parents had died but the causes of death were unknown. His brothers, sisters, sons and daughter were healthy. He was an ex-smoker. He drank one cup of double shochu daily and he had worked in an iron foundry.

Physical examination was unremarkable. Neurological examination demonstrated mild hyperreflexia of the bilateral lower extremities. His orientation was poor and he could not calculate easy subtraction problems. He showed easy excitability and became excited when examined by pinprick sensory test and Babinski’s sign.

Mini-Mental State Examination was 16/30. Chest Xp showed calcification of the left pleura. Chest CT showed bilateral thickening of pleura, regional thickening and calcification of the left pleura. Fiberoptic gastric endoscope demonstrated active stage gastric ulcer. MRI showed atrophy of the ambient gyri, dilatation of the bilateral inferior horn of the lateral ventricles and mild cerebral atrophy (Fig. 1A, B). IMP-SPECT showed a decreased blood flow of the bilateral frontal and temporal lobe. Three-dimensional stereotactic surface projection (3D-SSP) method. The hot area (red color) indicated decreased blood flow. Decreased blood flow was shown in the frontal and temporal lobe and preserved blood flow was shown in the posterior cingulate cortex and temporo-parietal area.

Figure 1. A, B: Coronal section of MRI T1-weighted image (TR 300 TE 20) revealed severe atrophy of the bilateral ambient gyri (arrows: medial wall of the inferior horn of the lateral ventricles) and dilated bilateral inferior horn of the lateral ventricles. C: IMP-SPECT. Three-dimensional stereotactic surface projection (3D-SSP) method. The hot area (red color) indicated decreased blood flow. Decreased blood flow was shown in the frontal and temporal lobe and preserved blood flow was shown in the posterior cingulate cortex and temporo-parietal area.
surface projection (3D-SSP) demonstrated decreased blood flow of the bilateral frontal and temporal lobe and preserved blood flow of the parietotemporal lobe and posterior cingulate gyri (Fig. 1C). He was treated by a proton pump inhibitor and fluids. Gradually, he regained his appetite. Thereafter, his general condition improved and he was discharged.

It has been previously reported that cases of AGD demonstrate severe atrophy of the ambient gyrus (3). The coronal section of MRI in the present case showed severe atrophy of the ambient gyrus and this finding was quite similar to the macroscopic findings of AGD previously reported (3). SPECT in this case showed decreased blood flow in the frontotemporal region and preserved blood flow in the parietotemporal area and posterior cingulate area. Alzheimer’s disease shows decreased blood flow in the posterior cingulate cortex and parietotemporal area (4). Dementia with Lewy bodies shows decreased occipital perfusion (5). The SPECT findings in the present case differed from those of Alzheimer’s disease and dementia with Lewy bodies. There was no hyper-oral behavior, bulimia or language disturbance such as semantic dementia (or Gogi aphasia). Since severe atrophy was restricted to the temporal lobe, Pick’s disease was unlikely. This patient presented with amnesia, easy excitability, hyperreflexia of the lower extremities, difficulty in calculation and disorientations while MRI showed severe atrophy of the ambient gyrus and preserved posterior cingulate cortex, the parietotemporal area and occipital lobe. The possibility of either Alzheimer’s disease or dementia with Lewy bodies was low, but there is a possibility that the diagnosis in this patient was argyrophilic grain dementia.

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