—Case Reports—

Isolated Prosopagnosia Caused by Damage to the Right Inferior Longitudinal Fasciculus: A Case Report

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Prosopagnosia is a cognitive disorder in which facial recognition is severely impaired despite normal vision and intelligence. Prosopagnosia was first reported in the 1800s, but its cause remains unclear. Although other neurological symptoms are often present, some patients have pure prosopagnosia. The bilateral occipital lobes are believed to be associated with symptoms. Recent brain imaging techniques have identified the right fusiform gyrus (rFG), located at the junction of the right occipital temporal lobe, as the affected region. In this report, we present a case of associative prosopagnosia with no concomitant symptoms in a 76-year-old man. Brain magnetic resonance imaging detected a subcortical hemorrhage in the right temporal lobe. Using tractography based on diffusion tensor imaging, we visualized atrophy of the right inferior longitudinal fasciculus (ILF). This is the first time tractography has been used to show a clear association between associative prosopagnosia and ILF damage projecting from the rFG. (J Nippon Med Sch 2025; 92: 220–224)

Key words: prosopagnosia, face recognition, right fusiform gyrus, inferior longitudinal fasciculus

Introduction

Prosopagnosia, also known as face blindness, is derived from the Greek *prosopon*, meaning face, and *gnosis*, meaning knowledge. It is a condition in which one cannot recognize faces despite having normal vision, intelligence, and object recognition. Prosopagnosia can be acquired or developmental and may result from stroke[1], trauma[2], or resection of the temporal occipital lobe in the minor hemisphere[3,4]. It is classified as apperceptive, associative, and amnesic[5]. Although the foci of prosopagnosia are gradually being elucidated, the definite foci of the subtypes are unclear. Here, we report a case of associative prosopagnosia due to subcortical hemorrhage in the right temporal lobe of a 76-year-old man. The case findings suggest a relationship between associative prosopagnosia and the right inferior longitudinal fasciculus (ILF).

Case Presentation

The patient was a 76-year-old man with a history of type 2 diabetes and angina. One day, he was suddenly unable to identify familiar faces. Although he was able to recognize gender, facial expressions, eyes, and noses, he could only identify people by their voices. He also reported that the faces of celebrities he saw on TV were different from those he remembered. Four months later, there was no change in his condition and he was referred to the neurology department of our hospital, at which time he reported being unable to recognize the face of the cardiologist who had been treating him for years.

The results of a physical examination were normal. A neurological examination showed that he was right-handed (LQ+100), and there were no abnormalities associated with the cranial nerves, including his eyesight and visual field. He exhibited a reduction in the tendon re-

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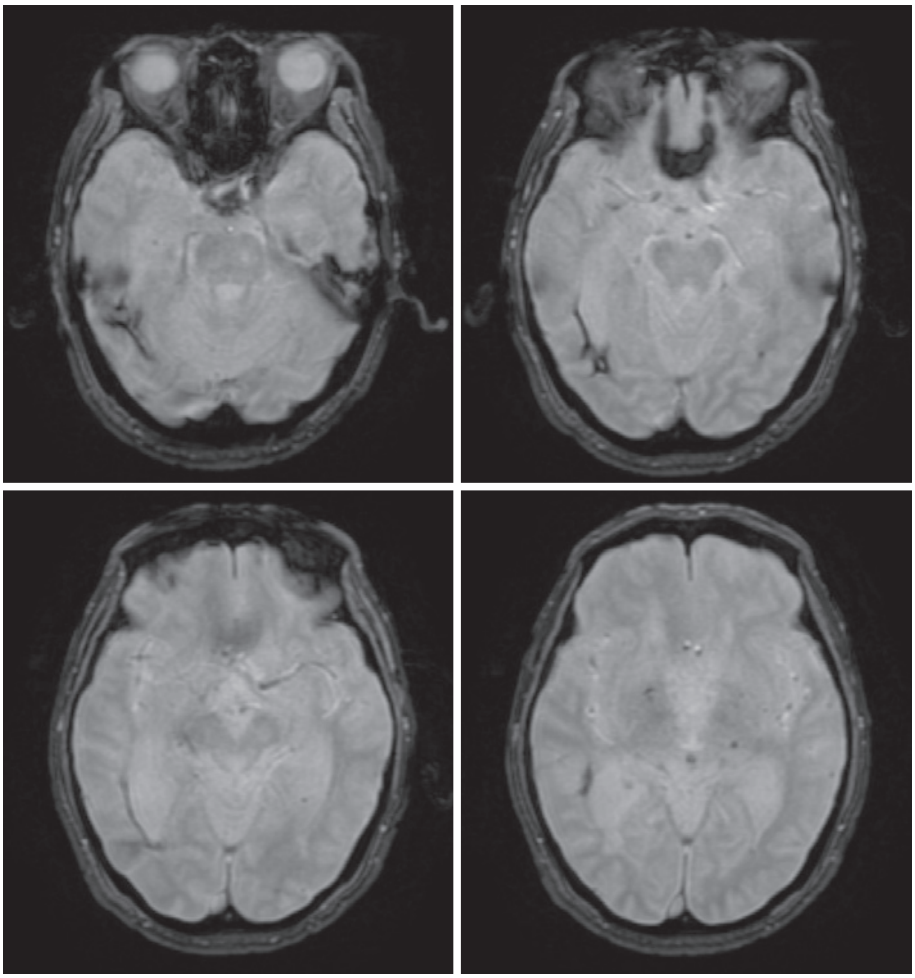


Fig. 1 MRI T2*WI images showing the subcortical hemorrhage in the right temporal lobe.

flex. The motor, sensory, and coordination examinations were normal. Blood, urine, and electroencephalography test results were normal. Brain magnetic resonance imaging (GE Healthcare, 1.5 T) revealed a subcortical hemorrhage in the right temporal lobe (**Fig. 1**). The results of cranial magnetic resonance angiography were normal.

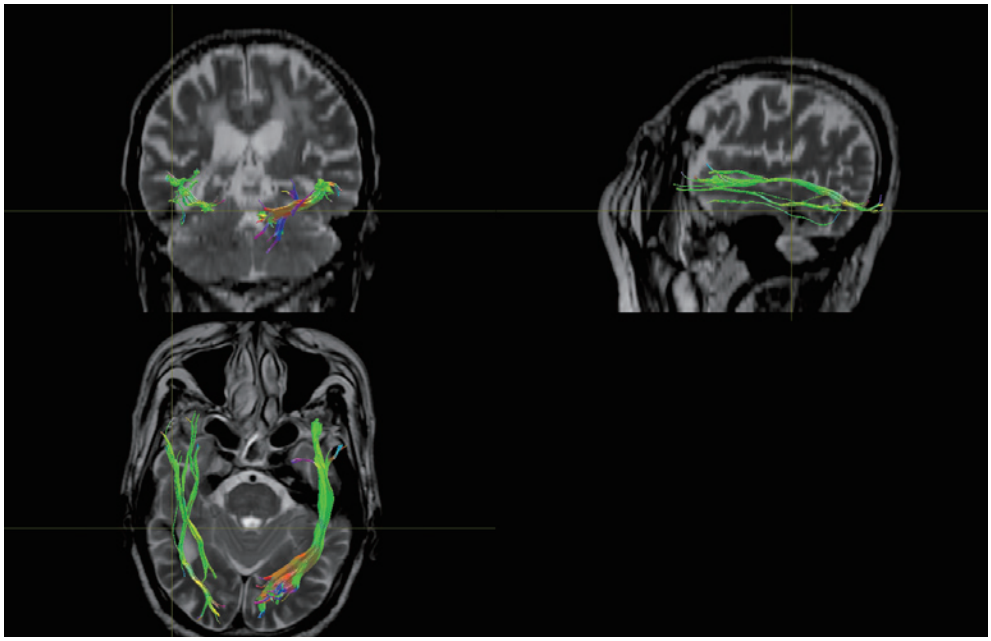
Neuropsychological examination showed no ideomotor apraxia, ideational apraxia, dressing apraxia, or object, color, finger, or lateral agnosia. He scored 28/30 on the Mini-Mental State Examination, 38/48 (79.1%, normal range >50%) on the Japanese and Caucasian Facial Expressions of Emotion, and 13/21 (62%) on the Facial Emotion Identification tests[6]. To evaluate facial recognition, we presented the faces of 15 celebrities with whom the patient was familiar. Although he successfully identified the sex of all 15 celebrities, he could only recognize eight. Moreover, he could not provide any information that would identify the people he was unable to recognize. For face recognition, we used the Visual Perception Test for Agnosia Famous Face Test version 2 (VPTA-FFT ver. 2)[7]. Twelve subjects (famous faces) were presented for each item, and the results are shown in **Table 1**. As for the face pattern, the patient was able to give a delayed response for one subject in "naming" and two in "instruction." Regarding the head pattern, although the number of correct answers increased, the deviation from the average for a healthy group was remarkable. The same results were seen for "recognition of name."

We used tractography based on diffusion tensor imaging to identify the responsible foci and visualize the association fibers. For diffusion MRI tractography, we acquired 30 volumes with b = 1,000 s/mm^2 and one volume without diffusion weighting. Diffusion MRI data were processed using FSL[8] and MRTrix3[9]. After estimating the orientational distribution function with constrained spherical deconvolution[10], right inferior longitudinal fasciculus (ILF) tractograms were generated using the deterministic tracking algorithm implemented in MRTrix3, with the seed and target placed in accordance with Wakana et al.[11]. Fractional anisotropy (FA) and the

Table 1 Scores on the Visual Perception Test for Agnosia Famous Face Test, version 2

	Patient	Average of healthy group (age >70 years)*
Face pattern		
Naming	23	10.476
Instruction	22	4.5264
Recognition of name	4	1.614
Head pattern		
Naming	19	0.55392
Instruction	12	0.3684
Recognition of name	3	0.15

*normative values from the manual



	FA*	ADC ($10^{-3}mm^2/sec$)**
Right ILF	0.25	1.2
Left ILF	0.31	0.99

*Fractional anisotropy

**Apparent Diffusion Coefficient

Fig. 2 Tractography images showing atrophy of the right inferior longitudinal fasciculus (ILF).

apparent diffusion coefficient (ADC) were computed from the same data by using the standard weighted least squares method[12]. The ILF tractograms were then used as regions of interest to extract tract-specific FA and ADC values. Using tractography, we were able to visualize atrophy in the ILF, a long-range white matter pathway that connects the occipital and temporal-occipital areas of the brain to the anterior-temporal areas (**Fig. 2**).

This study was approved by the Ethics Committee of Toho University Omori Medical Center (Date of approval: April 30, 2020).

Discussion

In this report, we describe a patient who was unable to recognize faces. He had no visual impairment, cognitive dysfunction, or other agnosia. While trying to recognize faces, he was able to distinguish between facial expressions and gender but could accurately identify people

only by their voices. He also complained that his memory of faces did not match the faces he saw. Therefore, we suspected associative prosopagnosia. Typically, when we look at a face, we tend to focus on its inner features, such as the eyes, nose, and mouth. However, prosopagnosia patients tend to look at external features such as the ears and hair[13,14]. We believe this explains why our patient scored better on the "head pattern" recognition in the VPTA-FFA ver. 2.

Functional magnetic resonance imaging (fMRI) has helped identify the foci responsible for face recognition. Regions in the ventral occipital-temporal cortex form the core of the face-selective network and include the occipital and fusiform facial areas. The anterior temporal lobe (ATL) is also considered an extended face network[15]. The ILF is a long white pattern pathway that connects these areas and is assumed to be involved in facial recognition[15]. The ILF tends to lateralize to the right and is suspected of playing a role in visual processing and memory[16,17]. Latini et al.[17] reported that the ILF consists of the fusiform, dorsolateral occipital, lingual, and minor cuneal branches.

Although there are three known prosopagnosia subtypes, the foci for each remain unclear. In associative prosopagnosia, as in the present patient, the ATL is believed to be the responsible lesion[18–21]. In our patient, a subcortical hemorrhage in the right temporal lobe was detected, and tractography confirmed atrophy of the ILF. These findings and previous reports strongly suggest impairment of the fusiform branch, which connects the fusiform gyrus to the anterior temporal regions, and the dorsolateral occipital branch, which connects the superior, middle, and inferior occipital gyri to the anterior temporal regions of the ILF. A limitation of this study is that we did not use the Cambridge Facial Memory Test.

In conclusion, this case showed an association with damage of the ILF projecting from the rFG, thus providing new insights on the cause of isolated associative prosopagnosia.

The patient provided written informed consent for publication of this report.

Data Availability Statement: All data generated or analyzed during this study are included in this article.

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Conflict of Interest: The authors have no conflicts of interest.

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