Transient phonemic paraphasia by bilateral hippocampus lesion in a case of limbic encephalitis

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Abstract

Although the hippocampus has not typically been identified as part of the language and aphasia circuit, recent evidence suggests that the hippocampus is closely related to naming, word priming, and anomia aphasia. A 59-year-old woman with limbic encephalitis of possible autoimmune etiology, after recovery of consciousness, presented with severe memory impairment in both anterograde and retrograde modalities, episodes of fear, hallucination and convolution, and transient fluent, phonemic paraphasia, together with small sharp waves diffusely by EEG. Brain MRI revealed bilateral symmetric, discrete lesions in the body to the infundibulum of the hippocampus.

The transient phonemic paraphasia noted in our patient may have been a result of primary damage in the hippocampus and its fiber connection to the Wernicke’s area or secondary partial status epilepticus that might have originated in the hippocampus.

Introduction

Although the hippocampus has not typically been identified as part of the neuronal circuit for language and aphasia, recent evidence suggests that the hippocampus is closely related to naming, word priming, and anomia aphasia.1,2 We present herein a case that presented with limbic encephalitis (LE),3 bilateral symmetric, discrete lesions within the hippocampus, as well as transient phonemic paraphasia, a combination that has not been reported previously.

Case Report

A 59-year-old, previously healthy, right-handed woman developed high fever (39°C) and vomiting. Five days later, she became somnolent and began to have generalized convulsions that brought her to our hospital. On the day of admission to our hospital, she was somnolent and began to have generalized convulsion, and transient fluent, phonemic paraphasia; e.g., omotai (heavy) > amatai, shinja, nebusoku (sleepless) > nobusoku, tsukatte morau (let someone to use it) > takette morau, passport > kusagari. Her speech was fluent, and word/sentence recognition was preserved. This phenomenon lasted for half a day and then disappeared, but was repeatedly observed by the co-medical staff for a week. We were not able to perform a detailed aphasia battery during a period of a week because of non-availability of a clinical psychologist. On the 44th day, EEG showed small sharp waves in performed, Frontal Assessment Battery (FAB) score 14/18. She had no speech difficulty or aphasia at all at that time.

On the 18th day after admission, brain MRI results revealed bilateral, symmetrical high signal lesions localized at the body to the infundibulum of the hippocampus, which slightly extended to the parahippocampal and the amygdala on diffusion-weighted images and fluid attenuated inversion recovery (FLAIR) images. Therefore, non-herpetic, autoimmune limbic encephalitis (LE) became the tentative diagnosis. The patient was then started on 3 courses of 1,000 mg/day intravenous methylprednisolone for three days.

After these treatments, her hallucination and seizures disappeared gradually, and she became able to eat meals, although her memory disturbances persisted. On the 31st day after admission, EEG showed 6Hz theta waves in the frontal lobe bilaterally, and no spikes were observed. On the 39th day, brain MRI still revealed bilateral, symmetrical high signal lesions at the body to the infundibulum of the hippocampus, which slightly extended to the parahippocampal gyrus and the amygdala only by FLAIR images (Figure 1). The CSF findings returned to normal. On the 41st day, she began to show signs of transient fluent, phonemic paraphasia; e.g., omotai (heavy) > amatai, saha (mackerel) > sabako, jinja (shrine) > chinja, nebusoku (sleepless) > nobusoku, tsukatte morau (let someone to use it) > takette morau, passport > kusagari. Her speech was fluent, and word/sentence recognition was preserved. This phenomenon lasted for half a day and then disappeared, but was repeatedly observed by the co-medical staff for a week. We were not able to perform a detailed aphasia battery during a period of a week because of non-availability of a clinical psychologist. On the 44th day, EEG showed small sharp waves in
Discussion

The diagnosis of LE in this case was based on: clinical features of acute onset of high fever, disturbance of consciousness, and epilepsy; and after recovery of consciousness, severe memory impairment, fear, and hallucination; the CSF finding of pleocytosis and increased protein content; bilateral symmetric lesions in the hippocampus by an MRI scan.

Our patient was unique in that she developed, during a course of LE, transient phonemic paraphasia. The paraphasias appeared to be very transient, lasting for approximately half a day, observed for one week. The hippocampus has not typically been identified as part of the language and aphasia circuit. However, recent evidence suggests that the hippocampus is closely related to naming, word priming, and anomia aphasia. Among these, Urbach et al. have described the results of the Wada test, in which sodium amytal and SPECT tracer 18F-hexamethylpropyleneamine oxime (HMPAO) were injected into the posterior cerebral artery, with 4 of the 14 injected subjects revealing transient (lasting 1-3 min.) fluent anomia aphasia, together with contralateral hemianopia.1 Paralyzed brain areas as shown by HMPAO were the parahippocampal gyrus, the hippocampus, and the occipital lobe. Similarly, Jernigan et al. performed MRI volumetry in normal volunteers and found that the hippocampal volume contributed independently to increased naming latency and decreased word priming by a speech test.2 These findings might be a reflection of anatomical evidence that the hippocampus has fiber connections to the Wernicke’s area.5

Another factor related to the transient phonemic paraphasia in our patient was the focal epilepsy. This is because our patient had seizures twice at the end of a one-week episode of phonemic paraphasia. EEG in our patient at the times of transient paraphasia also showed sharp waves in the frontal, temporal, and parietal lobes bilaterally. Although partial status epilepticus can present with multiple, discrete MRI lesions in the brain, MRI lesions in our patient were localized at the body to the infundibulum of the hippocampus. Previously, aphasic status epilepticus has been recognized in complex partial seizures (temporal lobe epilepsy).7 Dong et al. reported 4 aphasic patients in whom this condition lasted from two days to three months. In those patients, MRI findings were normal; but focal slow waves by an electroencephalography and focal hypermetabolism by 18F-fluorodeoxyglucose (FDG) positron emission tomography (PET) were shown in the temporal lobe. Taken these findings into account, the transient phonemic paraphasia noted in our patient may have been a result of primary damage in the hippocampus and its fiber connection to the Wernicke’s area or secondary partial status epilepticus that might have originated in the hippocampus and the temporal lateral cortex.

References