Case Report
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Klüver-Bucy Syndrome with Isolated Bilateral Hippocampal Atrophy Following Status Epilepticus

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Klüver-Bucy syndrome may result from affection of various location of brain. We report a case of Klüver-Bucy syndrome associated with isolated bilateral hippocampal atrophy without any abnormal lesion in other areas following status epilepticus. A 31-year-old man who had no significant medical history presented with status epilepticus after encephalitis of unknown etiology. He had been recovered from status epilepticus three weeks later, but afterwards he developed Klüver-Bucy syndrome: hyperphagia, hypersexuality, hypermetamorphosis, anterograde amnesia and dysosmia. Initial brain MRI showed T2 hyperintensity and swelling of isolated bilateral hippocampus, especially CA1 region without any abnormal lesion in other areas. One month later, follow-up brain MRI showed isolated bilateral hippocampal atrophy. This is a meaningful case report because this case differs from other reports of Klüver-Bucy syndrome in humans in that the anatomic abnormalities revealed by MRI were very selective. We report this case because this case is very educative for above reason. Moreover, this report would give us additional information of the relationship between human behavior and limbic system. **(2012;2:10-12)**

Key words: Status epilepticus; Klüver-Bucy syndrome; Hippocampal atrophy

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Klüver-Bucy syndrome (KBS) is a constellation of cognitive dysfunction including the inability to recognize the emotional significance of objects, hypersexuality, altered emotional behavior (particularly placidity), hyperorality, hypermetamorphosis (the tendency to react to every visual stimulus) and amnesia (anterograde, retrograde or global).[1,2] It has been reported in adult humans with structural lesions located in various locations of brain, bilateral or unilateral anterior temporal lobe,[1,3] amygdala,[4] and so on. We report herein a case of patient with KBS related to isolated bilateral hippocampal atrophy (without any abnormal lesion in other areas) following status epilepticus.

Case Report

A 31-year-old man who had no significant medical history presented with status epilepticus. He had headache, fever of 39℃ and generalized tonic clonic seizure without recovery of consciousness. His vital sign was stable except fever. On neurological examination, there was no abnormality except stuporous mental status between frequent generalized tonic clonic seizures. Initial blood tests showed no leukocytosis and normal level of inflammatory markers (C-reactive protein and erythrocyte sedimentation rate). Cerebrospinal fluid (CSF) was clean and contained 108 leukocytes/mm^3 (83% polymorphonuclear leukocytes and 17% lymphocytes) with normal glucose (78 mg/dL) and protein (42 mg/dL). Serologic testing of blood and CSF was normal. Initial brain MRI showed T2 hyperintensity and swelling of isolated bilateral hippocampus, especially CA1 region without any abnormal lesion in other areas (Fig. 1A).

On the day of admission, we assumed that the patient had herpes encephalitis, and started maximum dosage of acyclovir (15 mg/kg three times a day) which is the hospital protocol for treatment of herpes encephalitis. Electroencephalography (EEG) showed frequent spike and waves on bilateral fronto-temporal areas dominantly on temporal areas, so we started antiepileptic drugs (initial phenytoin loading with subsequent add-on of levetiracetam, oxcarbazepine and topiramate) and midazolam-coma therapy. Further serologic tests including polymerase chain reaction (PCR) for herpes species were negative and no bacteria and viruses were grown. He recovered

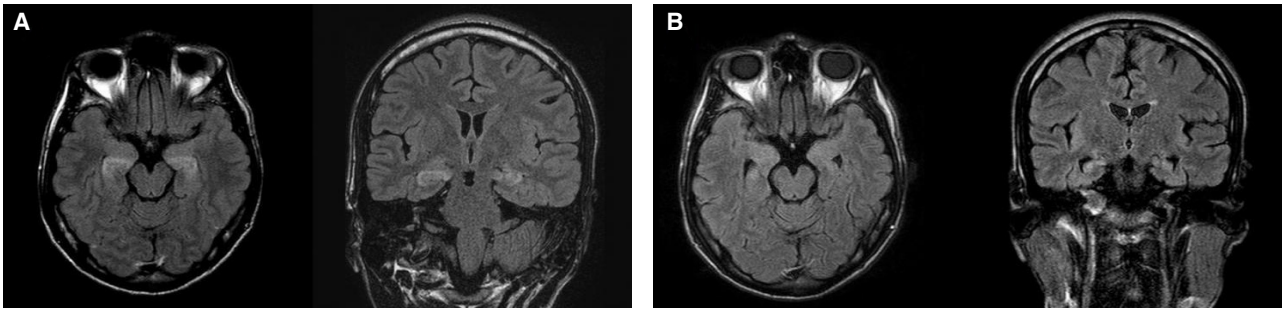

Figure 1. FLAIR images. (A) The initial axial and coronal MRI, three days after admission, show hyperintensities and swelling in the both hippocampi. (B) The follow-up axial and coronal MRI, which were performed forty-five days later, reveal atrophy in both hippocampi.

from status epilepticus three weeks later, but afterwards he developed KBS which is characterized by hyperphagia (he ate a large amount of food six to seven times a day), hypersexuality (he wanted to have a sex with the nurses, his ex-girlfriend and his mother), hypermetamorphosis, global amnesia (memory registration was normal, but recall was impaired) and dysosmia. On the 46th day of hospitalization, the score of his Korean-version of mini-mental status examination was 20/30 (orientation to time- 3/5, orientation to place- 2/5, immediate recall- 3/3, delayed recall- 0/3, attention and calculation- 3/5). Since we considered that his symptoms occurred due to secondary temporal lobe epilepsy, we performed an EEG, but his EEG showed no definite epileptiform discharges. Forty-five days later, follow-up brain MRI showed isolated bilateral hippocampal atrophy (Fig. 1B). Finally, we concluded that he had a KBS, and the patient was discharged on the 58th day of hospitalization and his cognitive dysfunction is persistent with minimal improvement for six months follow-up.

Discussion

This report describes a patient presenting as KBS after status epilepticus associated with MRI-documented structural lesions exclusively located in bilateral hippocampus. Unfortunately, we could not reveal the etiology of this patient.

After recovery from status epilepticus, many of the patients show postictal psychiatric phenomenon for a while. Typically, the delayed postictal psychosis lasts between 12 hours and 7 days, but occasionally psychiatric symptoms may persist for up to 3 months. However, the constellation of symptoms including hypersexuality, global amnesia, dysosmia, and hypermetamorphosis in this patient was compatible with KBS, and it lasts for a long time.

The lesions associated with KBS are assumed to include to affect not only the anterior temporal structures,[1-3] such as amygdala,[4] uncus and hippocampus, but also extratemporal structures such as cingulate or orbitofrontal cortex.[5] KBS may occur due to various etiology such as of dementia,[6] large territorial infarction,[7] uncal herniation,[8] or herpes encephalitis.[9] Lee *et al.*[10] reported that the FDG-PET may represent functional anatomy through reduced glucose metabolism over the temporal lobe in the patient with herpes simplex encephalitis. We wanted to observe the glucose metabolism of the temporal lobe and amygdala, but we could not perform the FDG-PET because of his low economic status.

This case suggests the possibility that the lesion limited to bilateral hippocampal area may result in the KBS. However, we cannot exclude the microscopic or functional involvement of other areas beyond hippocampus such as amygdala that could not documented by MRI. The functional neuroimagings such as FDG-PET or SPECT may give us additional information of the relationship between human behavior and limbic system.

Acknowledgement

We explained to the patient and his family that we will report his case and received consent from them. The authors have no financial conflicts of interest.

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