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

Hong-Kyun Park, MD¹, Kyeong-Joon Kim, MD¹, Hye-Jin Moon, MD¹, Seon-Jeong Kim, MD², Chang-Ho Yun, MD, PhD², Seong-Ho Park, MD, PhD²

¹Department of Neurology, Seoul National University Hospital, Seoul National University College of Medicine, Seoul; ²Department of Neurology, Seoul National University Bundang Hospital, Seongnam, Korea

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, and amnesia (anterograde, retrograde or global). It has been reported in adult humans with structural lesions located in various locations of brain, bilateral or unilateral anterior temporal lobe, amygdala, 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°C 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/mm³ (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. 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
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 neuroimaging 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.

References

1. Klüver H, Bucy PC. Preliminary analysis of functions of the temporal
lobes in monkeys. 1939. J Neuropsychiatry Clin Neurosci 1997;9:606-20.
2. Pradhan S, Singh MN, Pandey N. Klüver Bucy syndrome in young
children. Clin Neurol Neurosurg 1998;100:254-8.
3. Ghika-Schmid F, Assal G, De Tribolet N, Regli F. Klüver-Bucy syndrome

www.kes.or.kr
after left anterior temporal resection. *Neuropsychologia* 1995;33:101-13.

4. Aronson LR, Cooper ML. Amygdaloid hypersexuality in male cats re-examined. *Physiol Behav* 1979;22:257-65.

5. Yoneoka Y, Takeda N, Inoue A, et al. Human Kluver-Bucy syndrome following acute subdural haematoma. *Acta Neurochir (Wien)* 2004;146:1267-70.

6. Lanska DJ, Currier RD, Cohen M, et al. Familial progressive subcortical gliosis. *Neurology* 1994;44:1633-43.

7. Fragassi NA, Longobardi T, Pellegrino MG, Di Salle F, Grossi D. Kluver-Bucy syndrome. A case report. *Acta Neurol (Napoli)* 1990;12:138-42.

8. Rossitch E Jr, Carrazana EJ, Ellenbogen R, Alexander E 3rd. Kluver-Bucy syndrome following recovery from transtentorial herniation. *Br J Neurosurg* 1989;3:503-6.

9. Yoo HU, Ku BD. Relapsing herpes simplex encephalitis resulting in Kluver-Bucy syndrome. *J Korean Neurol Assoc* 2008;26:397-400.

10. Lee SM, Han DK, Kim HD, Lee JS. Serial MRI, SPECT and FDG-PET findings in a case of herpes simplex encephalitis. *J Korean Epilepsy Soc* 2005;9:94-6.