

Persistent Klüver-Bucy Syndrome after Bilateral Temporal Lobe Infarction

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Abstract- Klüver-Bucy (KBS) syndrome is a rare and complicated neurobehavioral syndrome in humans resulting from damage of bilateral anterior temporal portion, especially the amygdala. It can be seen in association with a variety of etiologies. Stroke is a rarely reported. Here we present a 50-year-old right handed man who developed persistent KBS after cardioembolic stroke involving bilateral lateral temporal lobes. He exhibited all clinical features of KBS including visual agnosia, hypersexuality, placidity, hyperorality and hypermetamorphosis. The anatomical basis of pathophysiology, clinical course and possible treatment are discussed.

Key Words: Klüver-Bucy syndrome, Temporal lobe infarction, Disconnection syndrome

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INTRODUCTION

Klüver-Bucy syndrome (KBS) was first described on the basis of experimental bilateral temporal lobectomy in monkeys⁽¹⁾. It is a rare neurobehavioral syndrome with a constellation of neurological symptoms including hyperorality (a strong tendency to examine all objects orally, putting objects into mouth, licking, biting, chewing, touching with lips, and bulimia), indiscriminate hypersexuality, visual agnosia (a psychic blindness of inability to recognize objects without a loss of gross visual discrimination), placidity with loss of normal fear and anger, and hypermetamorphosis (strong tendency to react to visual stimulus)⁽¹⁾. The first human KBS was described by Terzian et al. in 1955 in an intractable

epileptic who underwent bilateral temporal lobectomy⁽²⁾. Subsequently, this syndrome was reported in several neurological disorders that had destruction or dysfunction of bilateral mesial temporal lobes. The common etiologies of KBS reported in humans include herpes simplex encephalitis, toxoplasmosis, tuberculous meningitis, head trauma, hypoxia, hypoglycemia, heat stroke, Parkinson's disease, Alzheimer's disease and Huntington's disease⁽³⁻⁷⁾.

Full-blown KBS in human is rare and the diagnosis can be made based on the presence of at least three of the aforementioned symptoms⁽³⁾. Lilly et al.⁽³⁾ also pointed out the differences between humans and nonhuman primates is a more complex behavioral syndrome inevitably associated with aphasia, amnesia, dementia

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and seizure. Most cases of human KBS demonstrate a transient profile, but there are still few patients having persistent partial KBS with poor prognosis⁽⁴⁻⁵⁾. From the review of the literature, only one case of KBS was related to bilateral thalamic infarction⁽⁸⁾. Here we describe a full-blown and persistent KBS caused by bilateral lateral temporal lobe infarction of cardioembolic type.

CASE REPORT

A 50-year-old right-handed man, a heavy smoker, was first admitted in October 2005 for ischemic infarction over right temporal, parietal and insula areas with occlusion of right middle cerebral artery (MCA) distal to

the M1 portion. Stroke survey revealed left atrial appendage thrombosis without any valvular heart disease or arrhythmia. Work-ups for related vasculopathy, coagulation profile, autoimmune diseases including antiphospholipid syndrome and biochemistries were all unremarkable. Anticoagulant was used for secondary stroke prevention. He had mild sequelae as evidenced by hyperloquacity and elevated mood.

Unfortunately, he was found somnolent and confused at home prior to this admission in June 2006. Neurological examination revealed that the patient's muscle power and cranial nerves function were intact, but showed no response to any aural or visual stimulation without eye contact. He became alert about 2 days

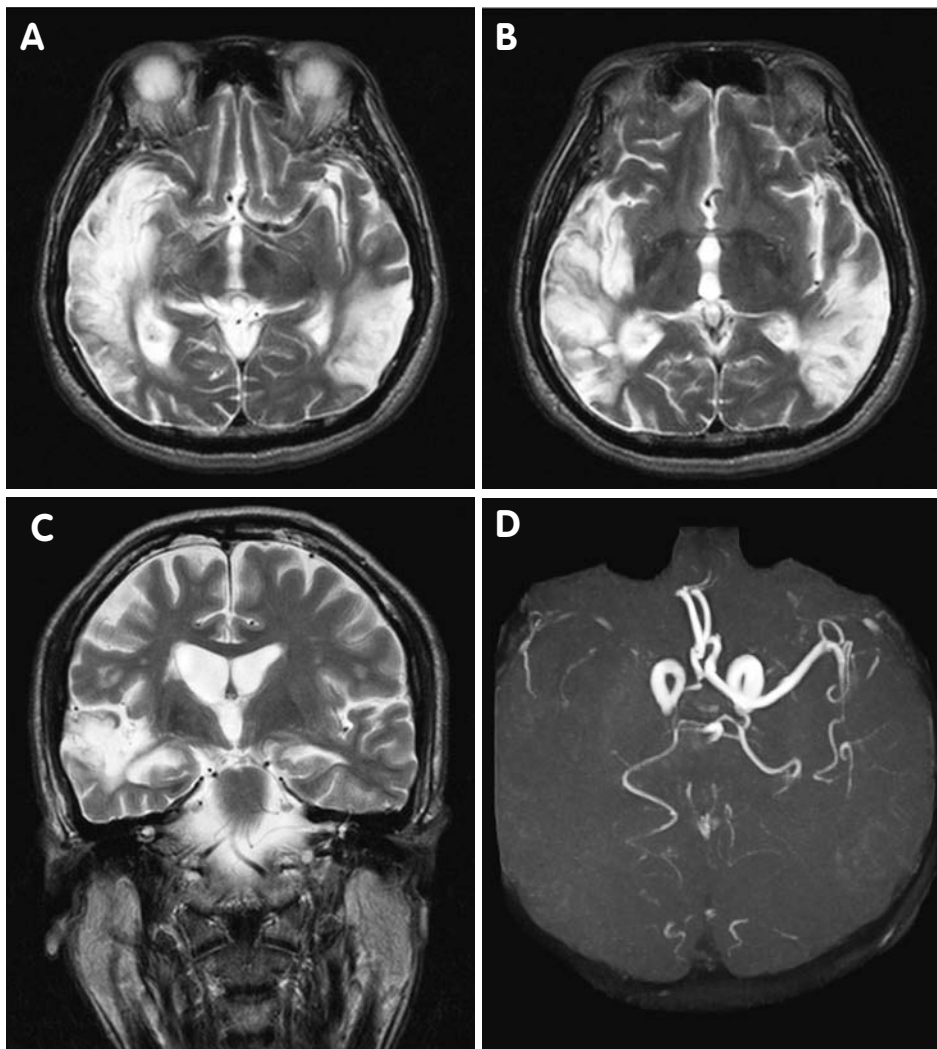


Figure. (A-B) The axial T2 weighted MR image shows that the infarct areas involving medial temporal lobe is more extensive on right side and more lateral on left side. (C) The coronal MRI shows that right amygdala is involved on right. (D) MRA shows the occlusion of right MCA and the temporal branch of left MCA.

later but could not recognize anyone, including his family or friends. But he always had a tendency to explore any object by repeatedly playing with it, or manipulating it if he did see them. He did not comprehend, repeat or name, nor response to written language or attempt to communicate in writing. He liked to touch everything nearby with his mouth, such as curtains, bedrails and other people's hands. When eating, he ate food without using his hands or utensils. He also liked to hug females such as nurses and friends nearby and to kiss them. He showed little affect and was easily distracted usually by shifting his attention. He wandered restlessly and, if restrained, became agitated but soon calmed down when his attention was diverted.

The EEG showed diffuse slow waves predominating over the bitemporal areas. CT scan of brain on the second day of admission showed a right temporal lobe infarction. A MRI two months later showed a new left temporal lobe infarction (Fig. A-D).

Initially, we used methylphenidate 20 mg/day for his somnolent state. He became more uncontrollable for oral tendency and hypersexuality. We discontinued methylphenidate and tried carbamazepine 600 mg/day without significant improvement. Antipsychotic agents (Quetiapine and Haloperidol) were used to control episodes of aggression and bizarre behavior. He could occasionally greet the caregivers, but would greet strangers, and sometimes followed them like a pet following a master even 6 months after stroke. Interestingly, he still had persistent visual agnosia, placidity, aphasia, but less hypersexuality and hyperorality 1 year later.

DISCUSSION

This case is an example of complete KBS, resulting from a disrupted of connection between temporal cortices and amygdala on both sides due to strokes. As in the initial description by Klüver and Bucy in 1939⁽¹⁾ in monkeys, bilateral extensive temporal lobectomy (amygdala, hippocampus, and adjacent cortical structures) was the anatomical pathomechanism. However, the lesions reported in humans are not as extensive and also not as

consistently localized as in animal experiments. Therefore, abnormal behavioral deficits in humans would not be fully developed or more complicated and accompanied with aphasia and amnesia such as in our case.

The exact anatomical basis in humans to produce KBS is still controversial. Most cases result from bilateral extensive destruction of the temporal lobe, including not only amygdala and uncus, but also hippocampus, cingulate gyrus, orbitofrontal, insular, and temporal cortices^(3-5,9). However, unilateral lesion may also cause similar symptoms. Some case reports reveal that a partial KBS or hypersexuality has developed after a left temporal lobectomy in right handed persons⁽¹⁰⁻¹¹⁾ and in a case of acute subdural hematoma compressing right amygdala⁽¹²⁾. Some cases result from epileptic discharges from the temporal lobe (especially left-sided) and respond well to carbamazepine⁽¹³⁻¹⁵⁾.

Geschwind et al.⁽¹⁶⁾ first viewed KBS as a disconnection syndrome produced by interruption of visual input to limbic circuit. Muller et al. described a KBS occurring after bilateral thalamic infarction which destroyed pathways connecting the dorsomedial thalamus with prefrontal cortex and other limbic area that are essential for memory and emotional regulation⁽⁸⁾. In our case, the infarcts were more extensive at the right temporal lobe with partial involvement of the mesial temporal area and amygdala, and less extensive and more lateral at the left temporal lobe and might disrupt sensory input to the medial temporal limbic system.

The clinical course of KBS varies among the reports. It is possible that KBS patients due to epileptic seizures, infection or post-infectious, and traumatic brain injury would be reversible if recognized early and treated appropriately^(3,5,14-15). Though some cases have good response to carbamazepine or antipsychotics, management of KBS is still a challenge.

CONCLUSION

Stroke is a rare cause for KBS. We report a persistent KBS following recurrent cardioembolic strokes involving bilateral temporal lobe. The prognosis seemed

to be poor in such case.

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