

Klüver–Bucy Syndrome Induced by Bilateral Basal Ganglion Lesion

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Abstract

Previous literature showed several etiologies result in Klüver–Bucy syndrome. The most common involved region is the bilateral mesial temporal lobes. In this article, we present the first case of Klüver–Bucy syndrome that occurred after CNS infection with bilateral basal ganglion lesion, which linked the sensory, motor and reward information by cortico-basal ganglia circuitry.

Keywords:

Klüver–Bucy syndrome, Basal ganglion, Infection, Cortico-basal ganglia circuitry, Temporal lobe

Introduction

Klüver–Bucy syndrome is a rare neuropsychiatric disorder and consists of placidity, docility, visual agnosia, dietary changes, hypersexuality and hyperorality which initially observed in a rhesus monkey after bilateral temporal lobotomy. Many etiologies have been reported in human; include CNS infection, cerebrovascular events, tumors, and various neurodegenerative disorders. The most common involved region is the bilateral mesial temporal lobes [1]. In this article, we present the first case of Klüver–Bucy syndrome in the world that occurred after CNS infection with bilateral basal ganglion lesion.

Case Report

This 11-year-old boy was admitted to acute psychiatric ward due to his disrupted behavioral and emotional symptoms, which presented for nearly two years after CNS infection. Tracing back his history, he contracted an acute CNS infection with presentation of fever, generalized tonic-clonic seizure and conscious disturbance

at the age of nine. The infection pathogen was proved to be mycoplasma by serological testing of the presence of mycoplasma IgM. After antibiotics, anticonvulsant and supportive care, he survived with general weakness and aphasia at first. He gradually recovered from the weakness and responsiveness also improved. However, he started to have various abnormal behaviors progressively six months later. The aberrant behaviors included hyperorality (tendency to touch objects by mouth and kiss others), hypermetamorphosis (strong tendency to react to visual stimulus), dietary changes (eat stool), loss of previously existing fears (increased aggressive behavior), and hypersexuality (masturbate in public). At school, he can't focus on lectures. The annoying behavior included increasing attentiveness to visual stimuli around him and a need to touch most of the things entered his sight. He frequently kissed other female classmates and used the dirty words towards young lady teacher. Brain MRI revealed lesion over bilateral basal ganglion (**Figure 1**) and no positive finding over medial temporal area (**Figure 2**).

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When we saw the patient at the age of 11, he still had above aggressive and impulsive behavior. The intellectual functioning test was done which showed full-scale IQ score was 67, what placed him below average. We treated him with risperidone 2mg/d and valproate 600mg/d; however, the effects were limited.

Discussion

This case meets the criteria of Klüver–Bucy syndrome: which includes changes in dietary habits, excessive oral tendencies, hypermetamorphosis, placidity and altered sexual behavior.

The previous reports showed that the Klüver–Bucy syndrome mostly occur after bilateral temporal lobe damage. Extratemporal involvement can be seen in up to 20-30% of the cases [2-4]. The most affected extratemporal lesions are seen in the frontal and parietal cortices, the occipital lobe, and brain stem. Our patient showed his main lesion in bilateral basal ganglion with preservation of bilateral medial temporal structures. Compelling evidence suggests that basal ganglion is involved in mediating human behaviors via so-called cortico-basal ganglia circuitry, the cortices include prefrontal cortex, limbic, and sensorimotor cortex [5,6]. It

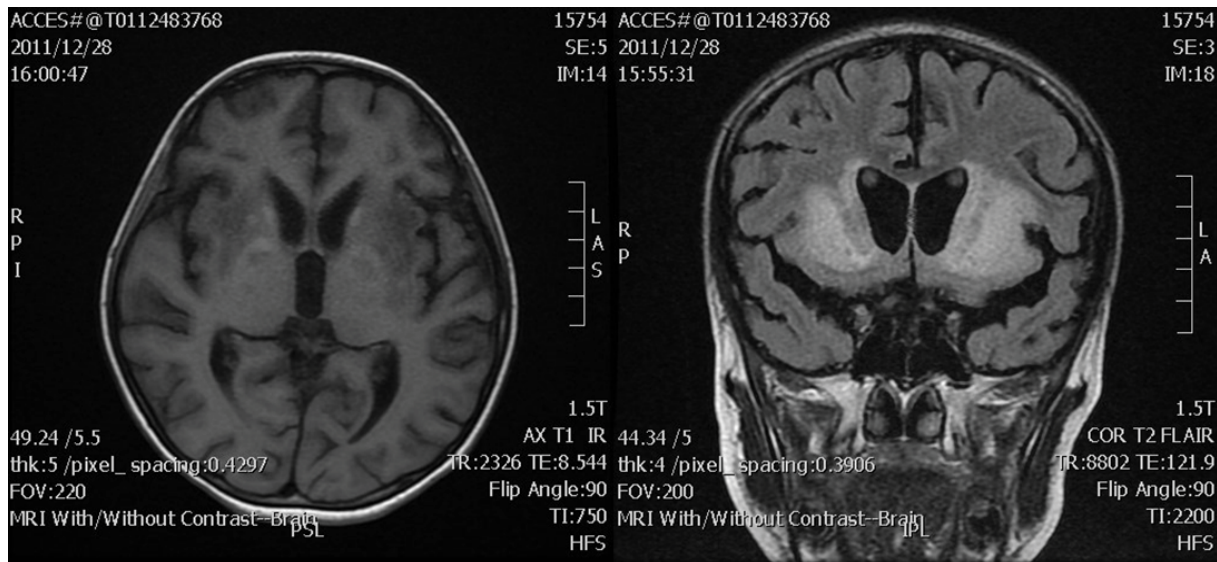


Figure 1: Increased signal intensity on T2WI near both basal ganglia, both caudate nuclei and right frontal subcortical areas.

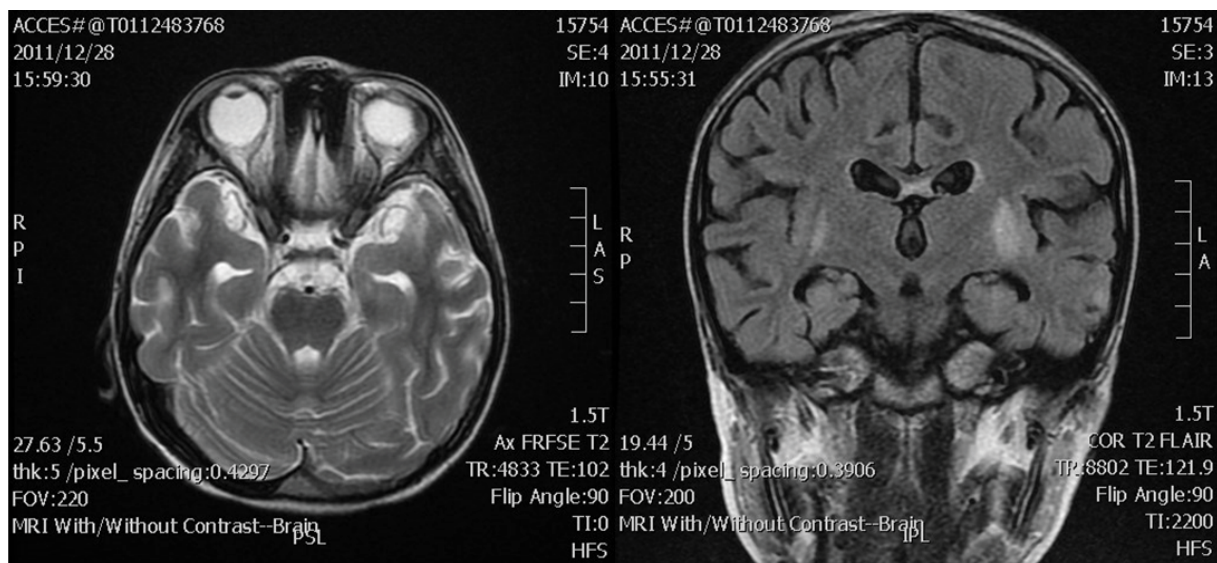


Figure 2: No degeneration change over bilateral medial temporal area.

plays an important role in linking the sensory, motor and reward information. The reward system mechanism may in charge of human motor control. If the cortico-basal ganglia circuitry disrupt, could contribute to a variety of emotional, motor and cognitive symptoms associated with psychiatric disorders [7].

Until now, there is no specific medication for Klüver–Bucy syndrome; several reports showed that the annoying behavior might respond to the treatment carbamazepine, risperidone and

selective serotonin reuptake inhibitors (SSRIs). Some experts use in combination with clonidine, methylphenidate and lamotrigine [8,9]. Actually, the drug history of the patient demonstrated that he did respond to these medications.

Conclusion

In conclusion, the case of our patient suggested that bilateral basal ganglion might be an extratemporal lesion for Klüver–Bucy syndrome. The optimal treatment for this distressful condition needs further investigation.

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