A case report of Kluver–Bucy syndrome following herpes simplex encephalitis

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Kluver–Bucy syndrome (KBS) is a rare syndrome seen after lesions of temporal lobes of the brain. Herpes simplex virus 1 is the most common infectious agent causing KBS owing to predilection of the virus to selectively affecting the temporal lobes. In this paper, we present a rare case of KBS that developed after herpes simplex encephalitis.

Keywords:
herpes simplex encephalitis, herpes simplex virus 1, Kluver–Bucy syndrome

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Introduction

Kluver–Bucy syndrome (KBS) is a rare neuropsychiatric disorder characterized by hyperorality, hypermetamorphosis (excessive exploration of the environment), hypersexuality, bulimia, placidity, and visual agnosia. This results owing to lesions affecting bilateral temporal lobes, in particular the hippocampus and amygdala (Kar *et al.*, 2018). Central nervous system infections, head injury, and neurodegenerative diseases are the common causes. Among infective pathogens, herpes simplex virus 1 (HSV1) is the most common agent (Poduval *et al.*, 2005). We report a patient who developed KBS following herpes simplex encephalitis (HSE).

Case report

A 35-year-old man was referred for psychiatric consultation with 5-month history of irritability, impulsivity, excessive reactivity, inability to recognize objects of daily use, putting inedible objects in mouth, inappropriate sexual behavior, memory problems, and difficulty in falling asleep. He had history of fever, headache, vomiting, altered sensorium, and seizures 6 months back. His medical records during that time showed normal complete blood picture, erythrocyte sedimentation rate of 6 mm, and normal liver function tests. No bacterial growth was found in urine culture. Hepatitis-B surface antigen, hepatitis-C virus antibody, and leptospira antibody immunoglobulin M were negative. Screening for HIV was nonreactive. Reverse transcription PCR was negative for severe acute respiratory syndrome coronavirus 2. Cerebrospinal fluid (CSF) examination revealed lymphocytic leukocytosis with elevated proteins. CSF gram staining, Ziehl–Neelsen staining, and culture showed no microorganisms. CSF for PCR test was negative for Varicella zoster virus and herpes simplex type 2 and positive for HSV1. MRI of the brain revealed area of restriction diffusion with corresponding T2, FLAIR hyperintensity in bilateral medial temporal lobes, insular region, and basifrontal lobes. On contrast study, there was diffuse leptomeningeal enhancement. Electroencephalogram was abnormal showing background slowing with left frontotemporal intermittent epileptiform discharges. A diagnosis of HSE was made by the treating physician. He was treated with acyclovir 10 mg/kg thrice daily, levetiracetam 1 g twice daily, and other supportive medication for 14 days. The patient was discharged on levetiracetam 500 mg twice daily, clobazam 10 mg at bedtime, and donepezil 10 mg at bedtime. Later, risperidone 0.5 mg was added to control his behavioral problems. As patient's behavioral symptoms were worsening, he was referred to our hospital. The patient had no past history of psychological illness. Premorbidly, the patient was well adjusted. Based on the history, past investigations, and presenting symptoms, we diagnosed him with KBS. The family members of the patient refused for admission and also for neuroimaging. We sent for an electroencephalogram examination, which showed no abnormality. Other blood investigations were with in normal range. Levetiracetam was stopped, as it can worsen behavioral symptoms in few patients, and he was started on carbamazepine 600 mg, risperidone 2 mg, and trihexyphenidyl 2 mg. Zolpidem 10 mg was added for sleep. During follow-up visits, his irritability, impulsivity, insomnia, and hypersexuality significantly

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improved. There was minimal improvement in hyperorality and forgetfulness.

Discussion

In 1937, Heinrich Kluver and Paul Clancy Bucy described a dramatic behavioral syndrome in Rhesus monkeys that had undergone bilateral temporal lobectomy. The full KBS comprised six symptoms: hyperorality, placidity, hypermetamorphosis, dietary changes, altered sexual behavior, and visual agnosia. They developed within 2–3 weeks following the surgery (Kluver and Bucy, 1937). Hyperorality was manifested by a tendency to place nonfood objects in the mouth and to examine objects orally. Hypermetamorphosis, a word coined by Heinrich Neumann, was characterized by immediate motor response upon visual presentation of an object, regardless of its history or reward value.

The first human case of KBS was described by Hrayr Terzian and Giuseppe Ore in 1955 in a 19-year-old boy who underwent bilateral temporal lobectomy for seizure disorder (Terzian and Ore, 1955). The first reported case of complete KBS in human was in a 22-year-old male with bilateral temporal damage owing to HSE by Marlowe *et al*. (1975). Etiologies of human KBS include trauma, Pick's diseases, Alzheimer's disease, Reye's syndrome, ischemia or anoxia, stroke, porphyria, carbon monoxide poisoning, limbic encephalitis, and HSE (Stephen *et al*., 1997). HSE is the most common infection causing KBS due to predilection of the virus to selectively affect the temporal lobes. Overall, 90% of the HSE cases are caused by HSV1 (Poduval *et al*., 2005). HSE is a rare life-threatening condition that is thought to be caused by the transmission of HSV1 either from nasal cavity to the brain's temporal lobe or from a peripheral site on the face, along the trigeminal nerve axon to the brainstem. Despite its low incidence (1 in 500 000 individuals per year), HSE is the most common sporadic fatal encephalitis worldwide (Whitley, 2006). The diagnosis can be confirmed only by means of PCR or brain biopsy. Viral CSF cultures are rarely positive. MRI is a more sensitive tool than computed tomographic scans.

The complete KBS rarely occurs in humans. The diagnosis requires presence of at least three of the six symptoms mentioned earlier. Among humans, placidity, indiscriminate dietary behavior, and hyperorality are the most common manifestations, whereas hypersexuality is rarely seen. These patients often have accompanying aphasia, seizures, and memory issues (Stephen *et al*., 1997). Treatment of psychological manifestations of KBS is a big challenge. It is mainly symptomatic and supportive. Among drugs, carbamazepine has shown good results in past case reports (Stewart, 1985; AlJadani, 2019). It is used for controlling most of the symptoms of KBS. Leuprolide is a hormone used to reduce the sexual behavioral abnormalities, whereas antipsychotics, selective serotonin receptor inhibitors, and anticholinergics are also used sometimes (Jha and Patel, 2004).

Conclusion

Although KBS is not a life-threatening condition, it can significantly affect the quality of life of the patient and the caregivers to a greater extent. Neurobehavioral syndrome following lesions of temporal lobes should raise suspicion of this condition. More research is needed about the pathophysiology of the symptoms and pharmacological and nonpharmacological treatment methods.

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Conflicts of interest

There are no conflicts of interest.

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