JETIR.ORG ISSN: 2349-5162 | ESTD Year : 2014 | Monthly Issue
JOURNAL OF EMERGING TECHNOLOGIES AND INNOVATIVE RESEARCH (JETIR)
An International Scholarly Open Access, Peer-reviewed, Refereed Journal

Klüver-Bucy syndrome: what is it?

Mr. Justin V Sebastian[1], Dr. Neenumol K Jose[2], Ms. Towar Shilshi Lamkang[3], Ms. Penina Langhu[4]

1. Associate Professor, Jagannath Gupta Institute of Nursing Sciences, India, Kolkata-700137
2. Associate Professor, Jagannath Gupta Institute of Nursing Sciences, India, Kolkata-700137
3. Ph.D Scholar, IES University, India, Bhopal-462044
4. Ph.D Scholar, IES University, India, Bhopal-462044

Abstract

Abnormalities in memory, social and sexual functioning, and idiosyncratic behaviours are hallmarks of Klüver-Bucy syndrome, a rare neurological condition of the brain that is linked to injury to both temporal lobes. Males and females are similarly afflicted by the extremely rare Klüver-Bucy syndrome. When the temporal lobes of the brain are injured, Klüver-Bucy syndrome might develop. The most prevalent cause of this is herpes simplex encephalitis. However, other infections and degenerative brain illnesses can also contribute to this symptom (a viral brain infection). Excessive oral tendencies, such as the impulse to put various objects into one's mouth, hypermetamorphosis (the drive to examine everything), memory loss, mood shifts, exaggerated sexual behaviour, apathy, placidity, distractibility, and agnosia are all possible signs of (difficulty identifying and processing visual information). It's also possible to observe a ravenous hunger that's difficult to satisfy. Dementia, or a loss of rational faculties, may also be accompanied by other symptoms.

Keywords: **Klüver-Bucy syndrome; Visual agnosia; Loss of normal fear and anger responses; Memory loss; Distractibility; Seizures, and Dementia**

Introduction

Memory loss and unusual behaviour may be symptoms of Klüver-Bucy syndrome (KBS), an extremely rare illness of the brain. It is a symptom of this sickness in some people to attempt to eat things that are clearly not food. Some people just have a very high sexual appetite. Seizures and dementia are among the more serious symptoms. Klüver-Bucy syndrome is brought on by harm to a significant part of the brain, the temporal lobes. This region of the brain is involved in the creation of new memories, the regulation of eating and sexual behaviour, and the interpretation of sensory and emotional input. Klüver-Bucy syndrome can be brought on by head trauma. Herpes simplex encephalitis is just one example of a disorder of the nervous system that can generate symptoms similar to those described. Individuals of various ages can be diagnosed with Klüver-Bucy syndrome. Unfortunately, there is currently no treatment available for this illness. However, medication can help lessen or even eliminate symptoms. Diagnosis at an early stage is crucial.

A historical perspective on Klüver-Bucy syndrome (KBS)

In 1888, two British experimental neurologists named Sanger Brown and Edward Albert Sharpey-Schäfer first recorded the clinical symptoms of KBS. They described the changes in monkey behaviour that occurred after the bilateral temporal lobes were removed. However, in 1939, neuropsychologist Heinrich Kluver and neurosurgeon Paul Clancy Bucy reported the full condition without knowing of the earlier description. Three weeks following a bilateral temporal lobectomy, a Rhesus monkey (called Aurora) exhibited a behavioural condition that was detailed in detail. Drs. Hrayr Terzian (1925–1988) and Giuseppe Ore initially identified KBS in a human patient in 1955, a 19-year-old who had had bilateral temporal lobectomy for seizures. Herpes simplex meningoencephalitis caused bilateral temporal injury in a 22-year-old male patient, who was the first person to be diagnosed with KBS.

Meaning

Klüver-Bucy syndrome is a very unusual form of mental retardation that occurs when both of the brain's temporal lobes sustain injury. In addition to inducing sexually inappropriate conduct, it also leads to people putting things in their mouths. Inability to detect objects visually (visual agnosia), diminished emotional responses (fear, rage), forgetfulness, irritability, convulsions, and dementia are all possible additional manifestations. Herpes encephalitis and trauma, both of which can cause brain damage, may be linked to the condition.

Does Klüver-Bucy syndrome go by any other names?

Other names for Klüver-Bucy syndrome include:

- Bilateral temporal lobe disorder.
- Post-encephalitic Klüver-Bucy syndrome.

- Post-traumatic Klüver-Bucy syndrome.

Epidemiology

Studies on human KBS are mostly limited to case studies and reports. Therefore, determining an accurate prevalence is challenging.

Etiology

KBS has been linked to a variety of diseases. From infectious diseases like shigellosis to psychological issues like methamphetamine withdrawal, the list of ailments for which there are numerous case reports is long. It is unknown, however, exactly how KBS developed under these circumstances.

- Herpes simplex encephalitis (HSE)
- Stroke (temporal lobe infarction - usually bilateral)
- Listeria meningoencephalitis
- Traumatic brain injury
- Central nervous system tuberculosis
- Primary cerebral Whipple disease
- Alzheimer disease
- Pick disease
- Hypoglycemia
- Acute sporadic porphyria
- Huntington disease
- Juvenile neuronal lipofuscinosis
- Toxoplasmosis
- Epilepsy
- Parkinson disease
- Heat stroke
- Shigellosis
- Methamphetamine withdrawal
- Systemic lupus erythematosus
- Anoxic-ischemic encephalopathy
- Neurocysticercosis
- Non-Hodgkin lymphoma
- Mycoplasmal bronchitis.
- Methotrexate leukoencephalopathy
- Subdural hygroma
- Susac syndrome - associated with partial KBS
- Anti-NMDAR encephalitis
- Exposure to cannabis

Pathophysiology of Klüver-Bucy syndrome

When either the temporal neocortex or the amygdala are destroyed on both sides, KBS clinical symptoms become noticeable. Because anterior temporal lobe impairment is typically less severe in humans compared to that following entire temporal lobe resection in monkeys, the full syndrome is rarely encountered in humans. There is an ongoing debate on the precise anatomical basis of KBS. Disturbances in the temporal regions of limbic networks, which interact with various cortical and subcortical circuits to regulate emotional behaviour and affect, are thought to be at the root of KBS. Lesion of both Ammon's horns on both sides is a prerequisite for KBS, as is involvement of the medial temporal lobe. Even though most people think that KBS is caused by problems in both temporal lobes, the amygdala, uncus, hippocampus, orbitofrontal and cingulate gyri, and insular cortex all have something to do with it.

Clinical Features

There is a lack of consensus in the literature when it comes to describing clinical manifestations. Common components are as follows:

- Amnesia: It is a type of memory loss in which the person can't remember things that happened recently or in the past. It can be either anterograde (forgetting what happened during the amnesic episode) or retrograde (forgetting what happened before it).
- Tameness: Calmness, also known as docility, is characterized by a decreased "flight or fight" response. Overeating and eating things that aren't food are both signs of hyperphagia, which means too much eating.
- Hyperorality: "The urge or compulsion to put things in one's mouth for a closer inspection"
- Hypersexuality: This resulted in an increased desire to have sex and an increased propensity to seek sex with inappropriate objects.
- Visual agnosia: Disorientation when it comes to recognizing members of one's own family or familiar objects.

Here are some examples of inconsistent presentations:

- Hypermetamorphorsis; "an uncontrollable want to take in and respond to everything in view"
- Having a diminished or nonexistent emotional reaction

Diagnosis of Klüver-Bucy syndrome

Klüver-Bucy syndrome is diagnosed by doctors after a thorough physical examination and other diagnostic procedures. Exams like a CBC and a CT scan might fall into this category.

- CT scan.
- Electroencephalogram (EEG).
- MRI.

A proper diagnosis at an early stage is crucial for successful treatment. Occasionally, Klüver-Bucy syndrome is misdiagnosed as autism. A proper diagnosis can only be made after an optimal course of treatment has been determined.

Treatment

- Patients need close supervision not only to prevent bulimia and the resulting obesity but also to stop the unconstrained and inappropriate sexual activity that has reportedly led to at least one patient's conviction for a crime.
- Changes in mood or behaviour after HSE treatment could mean a relapse, which would mean taking aciclovir for a longer time.
- Carbamazepine has been demonstrated to be effective, but selective serotonin reuptake inhibitors (SSRIs) may be superior.

Prognosis

However, recovery from HSE can take a long time, and cognitive and behavioural problems might range from very severe to very modest. Memory loss fits with the idea that structures in the medial temporal lobes are in charge of how memories are formed and stored.

Complications

The patient's hyperorality and hypermetamorphosis make him more likely to try to put anything he finds in his mouth. As a result of his hypersexuality, he may approach others with the intention of having sexual relations, which could result in legal action if the diagnosis is not known. Bulimia can cause you to gain weight, lose electrolytes, and not take care of your teeth as well as you should.

Reference

1. Kar SK, Das A, Pandey S, Gupta B. Kluver-Bucy Syndrome in an Adolescent Girl: A Sequel of Encephalitis. J Pediatr Neurosci. 2018 Oct-Dec;13(4):523-524.
2. Vannemreddy PSSV, Stone JL. Sanger Brown and Edward Schäfer before Heinrich Klüver and Paul Bucy: their observations on bilateral temporal lobe ablations. Neurosurg Focus. 2017 Sep;43(3):E2.
3. Klüver H, Bucy PC. Preliminary analysis of functions of the temporal lobes in monkeys. 1939. J Neuropsychiatry Clin Neurosci. 1997 Fall;9(4):606-20.
4. TERZIAN H, ORE GD. Syndrome of Klüver and Bucy; reproduced in man by bilateral removal of the temporal lobes. Neurology. 1955 Jun;5(6):373-80.
5. Brigo F. Hrayr Terzian (1925-1988): a life between experimental neurophysiology and clinical neurology. Neurol Sci. 2021 Sep;42(9):3939-3942.
6. Marlowe WB, Mancall EL, Thomas JJ. Complete Klüver-Bucy syndrome in man. Cortex. 1975 Mar;11(1):53-9.
7. Lanska DJ. The Klüver-Bucy Syndrome. Front Neurol Neurosci. 2018;41:77-89.
8. Olson DA. Klüver-Bucy syndrome as a result of minor head trauma. South Med J. 2003 Mar;96(3):323.
9. Salloway S, Malloy P; Cummings. The Neuropsychiatry of Limbic and Subcortical Disorders. American Psychiatric Pub.1997. p. 125.
10. Ozawa H, Sasaki M; Sugai K; Hashimoto T; Matsuda H, Takashima S, Uno A, Okawa T. "Single-Photon Emission CT and MR Findings in Klüver-Bucy" (PDF). American journal of neuroradiology (Oak Brook, IL,: American Society of Neuroradiology), 1997, 18 (3): 540–542.
11. Afifi, Adel K., Bergman R. Functional Neuroanatomy: Text and Atlas. McGraw-Hill Professional. 2005, p. 299.
12. Jha S, Patel R; Kluver-Bucy syndrome -- an experience with six cases. Neurol India. 2004 Sep52(3):369-71.

13. Devinsky J, Sacks O, Devinsky O; Kluver-Bucy syndrome, hypersexuality, and the law. Neurocase. 2010 Apr16(2):140-5. Epub 2009 Nov 18.
14. D Ku B, Sang Yoon S; Relapsing herpes simplex encephalitis resulting in kluver-bucy syndrome. Intern Med. 201150(7):763-6. Epub 2011 Apr 1.
15. Pascual-Castroviejo I, Pascual-Pascual SI, Viano J; Kluver-Bucy syndrome. Seven year follow-up of one patient. Neurologia. 2008 Mar23(2):114-8.
16. Raschilas F, Wolff M, Delatour F, et al; Outcome of and prognostic factors for herpes simplex encephalitis in adult patients: results of a multicenter study. Clin Infect Dis. 2002 Aug 135(3):254-60. Epub 2002 Jul 10.
17. Jha S, Patel R, Yadav RK, et al; Clinical spectrum, pitfalls in diagnosis and therapeutic implications in herpes simplex encephalitis. J Assoc Physicians India. 2004 Jan52:24-6.