

Case Report

Kluver- Bucey syndrome - An experience with six cases

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The Kluver-Bucey syndrome (KBS) is a neurobehavioral syndrome and can be seen in association with a variety of neurological disorders. Case records of 6 patients with KBS seen during a period of 5 years in a university hospital were reviewed. During the study period 6 patients with KBS, aged between 4 and 14 years, were seen. Hyperorality, hypersexuality, and abnormal behavior were the most common manifestations. Of the 6 patients, 5 had recurrent unprovoked seizures. The associated neurological disorders included anoxia-ischemic encephalopathy (2), herpes simplex encephalitis (1), neurocysticercosis (NCC) (1), traumatic brain injury with gliosis (1 case) and tuberculous meningitis (1 case). Prognosis was poor in all the patients except in the patient with NCC.

Key Words: Kluver-Bucey syndrome, herpes, anoxia-ischemic encephalopathy, neurocysticercosis, traumatic brain injury, tuberculous meningitis

Introduction

The first case of Kluver-Bucey Syndrome (KBS) was reported in a 22-year-old male patient with bilateral temporal damage due to herpes simplex meningoencephalitis.¹ The syndrome is characterized by psychic blindness (inability to recognize familiar objects), hypermetamorphosis (strong tendency to react to visual stimulus), increased oral exploration, placidity, indiscriminate hypersexuality and change in dietary habits. Though the clinical characteristics of KBS are similar in both rhesus monkeys (in whom it was first described)² and humans, some features like aphasia, amnesia, dementia, and seizures are probably exclusive to humans.^{3,4} The syndrome has been described in association with many disorders of the central nervous system like Alzheimer's disease, juvenile neuronal lipofuscinosis, Huntington's disease, herpes simplex encephalitis (HSE), toxoplasmosis, traumatic brain injury, hypoglycemia, acute intermittent porphyria, traumatic brain

injury, tuberculous meningitis, heat stroke and Shigellosis.^{5,6,7}

We report the clinical characteristics of 6 patients with KBS associated with different etiologies including neurocysticercosis and tuberculous meningitis.

Case Reports

The case records of patients with KBS seen in a university hospital over a period of 5 years were reviewed. The data collected included demographic data, clinical and radiological characteristics, electroencephalographic findings and the possible associated neurological disorders (Table 1). Hyperorality, hypersexuality, and abnormal behavior were the most common manifestations. Of the 6 patients, 5 had recurrent unprovoked seizures. The associated neurological disorders included anoxia-ischemic encephalopathy (2 cases), herpes simplex encephalitis (1 case) (Figure 1), neuro-cysticercosis (NCC) (1 case), traumatic brain injury with gliosis (1 case), and tuberculous meningitis (1 case). With the exception of the patient with NCC, the outcome was poor in the other patients.

The clinical characteristics of the patients with KBS associated with neurocysticercosis and tuberculous meningitis are presented in detail. Both the causes are very unusual.



Figure 1: Cranial MRI demonstrating the hyper intensities (T2) in Temporal and Parietal lobes, suggesting HSE.

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Table 1: Outline of clinical features of the 6 patients of Kluver Bucy Syndrome

Age/sex	Etiology	Visual agnosia	Orality	Hypermetamorphosis	Behavior	Sexual hyperactivity	Dietary habits	Seizures	Cranial imaging
1 7/M	CA	-	HO	Present	Placid	Pelvic thrusting	-	GTCS	Diffuse cerebral atrophy
2 14/M	HSE	Present	HO	-	Violent,Irritable	Rubbing of genitals	-	GTCS	B/ T P involvement
3* 12/F	NCC	-	HO	Present	Violent	Hypersexuality	Bulemia	CPS	Multiple NCC
4 11/M	Head injury	-	HO	Present	Offensive	Hypersexuality	Bulemia	CPS	Temporal atrophy
5* 3/M	TBM	-	HO	Present	Hostile	Hypersexuality	-	-	Mild hydrocephalus
6 9/ M	CA	-	HO	-	Apathetic	Pelvic thrusting	-	GTCS	Cerebral atrophy (FT)

TBM – Tuberculous meningitis, CA – Cerebral anoxia, HSE – Herpes simplex encephalitis, NCC – Neurocysticercosis, T P-Temporoparietal, GTCS- Generalized tonic clonic seizures, CPS- complex partial seizures, FT –frontotemporal, HO- Hyperorality

• - detailed in text

Case 1 – Neurocysticercosis

A 12-year-old girl presented with recurrent partial seizures of 6 months duration. Some of the seizures were associated with post-ictal Todd’s palsy. Seizures were well controlled with carbamazepine. She presented with headache, recurrent vomiting, and abnormal behavior. The abnormal behavior included explosive temper, hyperorality and increased sexual instincts in the form of stroking sex organs. She would become placid when someone scolded her for her abnormal behavior. Neurological examination revealed bilateral papilledema and no other deficits. Cranial CT revealed multiple contrast enhancing granulomas with perilesional edema scattered bilaterally in both the cerebral hemispheres. The CT findings were suggestive of neurocysticercosis (Figure 2). She improved symptomatically with steroids and mannitol. There was an improvement in the frequency of outbursts, hyperorality, and sexual orientation. At the last follow-up in the outpatient clinic she was seizure-free and had no behavioral abnormalities.

Case 2 - Neurotuberculosis

A 3-year-old male child was referred to our neurology outpatient clinic for progressive blindness and abnormal behavior. When he was 2 years old, he had fever, vomiting, and altered consciousness and was diagnosed as a case of tuberculous meningitis. Cerebrospinal fluid (CSF) examination done at that time showed proteins of 210 mg/dl, sugar 30 mg/dl, and a cell count of 412/cum with 85% lymphocytes. Enzyme linked immunoabsorbant assay (ELISA) and polymerize chain reaction (PCR) for mycobacterium were positive in CSF. EEG showed diffused slow waves. Cranial MRI revealed massive hydrocephalus and basal enhancing exudates (Figure 3). He was started on anti-tuberculous drugs and steroids. He regained consciousness with anti-tuberculous drugs over the next 3 weeks. However, he was unable to speak, comprehend, or perform activities of daily living including self-care. Parents observed gradual deterioration in his vision, which progressed to total blindness. After 3 weeks he developed gross behavioral abnormalities. The patient nibbled virtually everything on which he could lay hands and even ingested a few metallic objects. He made constant attempts to squeeze the genitals. He appeared shy but became hostile on slight provocation.

Self-inflicted, deep bite marks were present on both the hands. He could sit and stand but had a spastic gait. Neuropsychological examination could not be done due to lack of cooperation. Parents denied further investigations and neurosurgical management.

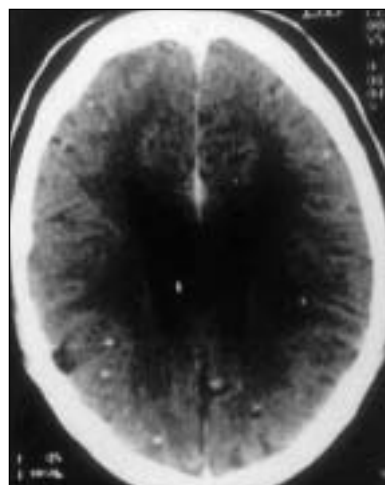


Figure 2: Cranial MRI demonstrating multiple neurocysticerci in different stages.

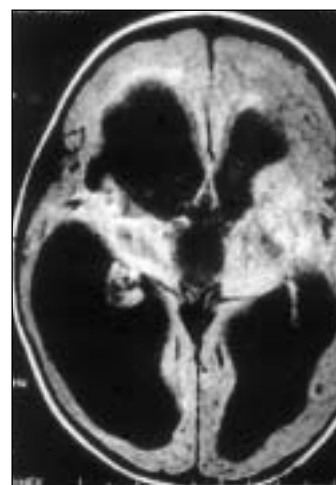


Figure 3: Cranial MRI demonstrating the hydrocephalus and exudates in patient of TBM.

Discussion

Though the anatomical basis of KBS with other diseases is controversial, its intimate association with temporal lobe damage has been widely reported.^{8,9} Demonstration of diffuse cerebral atrophy in patients with heat stroke, and isolated symmetrical damage to the amygdalae and their cortical connections following cancer treatment has been widely confirmed by imaging and positron emission tomography. It has been conjectured that this syndrome also results from the disruption of pathways connecting the dorsomedial thalami with prefrontal cortices and other limbic areas which are essential for memory and regulation of impulses and emotions.^{10,11,12}

Localizing studies (clinical and anatomical) indicate the involvement of the phylogenetically old medial temporal lobe regions and bilateral lesions of Ammon's horn as a sine qua non for the production of the syndrome. The mechanism of injury in these cases, apart from direct lesions of the temporal lobe also involves compression of both hippocampi.¹³ Involvement of temporal lobes by hydrocephalus in tubercular meningitis and the inflammatory reaction of the brain to the antigen liberated by the degenerating cysticercal cyst may probably explain the clinical features observed in our patients with tuberculous meningitis and NCC. The patient with KBS in association with NCC probably suggests that in certain clinical settings this syndrome can be reversible. Early recognition of such pathologies and appropriate treatment is essential to prevent long-term consequences.

Certain features of KBS (especially self stimulation of genitals and hyperorality) are common to children with severe learning difficulties arising from a variety of causes. The underlying mechanism, which is rather complex, is a mixed effect of generalized cognitive and sensory deficits in a child, often developing in an environment not sensitive to its special needs. Disentangling the non-specific and specific etiological factors is rather difficult.¹⁴ Of the 6 patients, 5 had recurrent seizures and in none of them have we observed any temporal worsening in these symptoms in the post-ictal phase. A number of changes in sexual behavior are associated with temporal lobe epilepsy and improvement with temporal lobectomy. As many as 71% of the patients with temporal lobe epilepsy exhibit altered sexual behavior associated with ictal events. Inter-ictal hyposexuality occurs in 80% of the patients; 20% exhibit a variety of paraphilias. When patients with hyposexuality have unilateral temporal lobectomy, libido frequently increases and may become pathological. Conversely, aberrant sexual behavior as part of seizure semiology of temporal lobe epilepsy has also been well documented.¹⁵ Perhaps

episodes of seizures reflect transient bilateral dysfunction of the temporal lobe. Such temporary functional bilateral lobectomy could cause the same syndrome as an anatomical lobectomy and would be similar to the pathophysiological mechanism, which may account for our patient's behavior. Carbamazepine and leuprolides have been found to decrease the sexual behavioral abnormality in some individuals with KBS. Other medications such as haloperidol and anticholinergics may also be useful in treating behavioral abnormalities associated with KBS.¹⁶

There were major limitations in our observations. There was reluctance and embarrassment on the part of the parents to provide elaborate details pertaining to hypersexuality. Detail neuropsychological evaluation was not possible as most of the patients were not cooperative to do these tests. There was a long delay between the onset of the symptoms and the diagnosis. Detailed diagnostic workup could not be done because of non-affordability of the workup. The follow-up was difficult due to lack of cooperation and unwillingness to consult a psychiatrist.

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