Kluver-Bucy syndrome - a dreadful neuropsychiatric disorder following herpes simplex encephalitis - a case report

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Abstract: Kluver-Bucy syndrome was described in the 50s of the 20th century as a group of neuropsychological symptoms such as changes in behaviour, emotions and memory. Various reasons for its causation have been mentioned in the literature. Here we are presenting a young male who presented to us with behaviour disturbances, emotional lability, forgetfulness following fever with altered sensorium, which was diagnosed and treated as herpes simplex encephalitis. On examination he had apathetic look, reduced word output, difficulty in executive function, memory and emotional lability. He was started on antipsychotics (risperidone), on follow up his behavioural symptoms improved with no improvement in memory and naming. There is need for early diagnosis and treatment of brain fever to prevent dreadful sequelae, along with early initiation of rehabilitation and modification of pathological behaviours.

Keyword: Kluver-Bucy syndrome, herpes simplex encephalitis

INTRODUCTION
In the 1930s, Klüber and Bucy gave their names to an intriguing cluster of behavioural changes that were noted after bilateral removal of the anterior temporal lobes in primates (1). The Klüber-Bucy presentation includes:
1. "Psychic blindness," or the inability to recognise the emotional significance of objects;
2. Hypersexuality, often directed indiscriminately;
3. Altered emotional behaviour, particularly placidity;
4. Increased oral exploratory behaviour and the ingestion of inappropriate objects (pica);
5. "Hyper metamorphosis," or the tendency to react to every visual stimulus; and

According to Lilley (1983) - Three of the afore-mentioned symptoms are required for the diagnosis of Klüber-Bucy Syndrome (2). The causes of Klüber-Bucy syndrome in human from literature are bilateral amygdala ablation (4), diseases of central nervous system (herpes simplex encephalitis), hypoxic anoxic encephalopathy, Pick’s disease, cranial trauma, Reye’s syndrome, CO poisoning, subdural haemorrhage etc(2,5). First case of Klüber-Bucy Syndrome in humans was reported in a 22 yrs old male following Herpes simplex meningoencephalitis (HSE)(4). Here we are presenting a case of neuropsychiatry disorder, sequelae of HSV Encephalitis.

CASE REPORT:
21 yrs old male presented with complaints of inappropriate behaviour, disturbance in emotions and memory for past two years. The complaints started following an episode of prolong fever with altered sensorium requiring hospitalisation for one month duration. Based on clinical profile, neuroimaging and cerebral fluid analysis, he was diagnosed as a case of Herpes Simplex Encephalitis. In months time he was found to have disturbed sleep, change in eating behaviour (voracious eating, pica). He started making obscene gestures (fondling genitals) and made inappropriate sexual advances towards the females including mother and sisters. He was found to be walking nude in the house and outside requiring frequent instruction to dress adequately. Family members noticed he had difficulty in remembering the instructions, names of his close friends and objects of common use like pen, bucket, mug etc. When taken to construction site, he was unable to handle the instruments, which he used to be expert previously. His interaction became limited to short, simple sentences. He was noticed to be crying and smiling while watching serials and conversing with others, which was unusual. His behaviour ranged from uncontrolled angry outburst to being complete passive at times. His physical examination was normal. Central nervous system examination: ill sustained attention and concentration. Lobar function revealed features of frontal and temporal lobe involvement- defective executive function, utilization behaviour, motor perseveration, defective motor sequencing, blunted emotional reaction, impaired visual memory, reduced word output and impaired visuo-spatial memory. Cranial nerve examination showed, slow saccades. Fundus showed temporal pallor. Motor and sensory system – deep tendon reflexes brisk He also had positive frontal release signs (grasp and rooting reflexes).

On the Mini- Mental State Examination (MMSE), he scored 13 out of 30. His Mental status examination revealed - decreased psychomotor activity with inappropriately smile. Talk-quantum, tone, rate decreased with increased reaction time and lack of prosody. Poverty of thought content observed though there were no obsessions or delusions. There was no perceptual pathology. His judgement and reality testing abilities were intact. Mood subjectively happy, objectively- inappropriate with emotional lability. His haematological investigations, Serum copper and ceruloplasmin were within normal range, Negative VDRL and HIV Non Reactive. CSF analysis: protein-145mg/dl, glucose- 73mg/dl,
Psychological assessment revealed gestalt disturbance of organic aetiology impairing cognitive functioning. With this history, clinical features and laboratory findings, according to ICD-10, he was diagnosed as a case of - F-07 Personality and Behavioral disorder due to brain disease, disorder and dysfunction. F-07.1 Post encephalitic syndrome. He was started on haloperidol 5mg later shifted to risperidone 4mg due to extra pyramidal symptoms with benzhexol 2mg and diazepam 10mg. After initiating treatment, he showed significant improvement. For past 8 months, on regular follow up he continues to show improvement in his behaviour with minimal to no improvement in amnesia and difficulty remembering and naming persons and common objects.

DISCUSSION –
This patient presented with clinical features suggestive of a diagnosis of Klüver-Busy syndrome (2). The symptom complex was preceded by fever with altered sensorium of 1 month duration, diagnosed as Herpes simplex encephalitis (HSE) by serological testing of CSF for HSV antibody (IgG) and latter confirmed by HSV DNA PCR, which has diagnostic sensitivity of 95% and specificity of 99%(4). Herpes simplex encephalitis (HSE) is the most common cause of fatal viral encephalitis in our part of world with majority of illness caused by type 1 variant (90%). Lot of case reports and series have been documented in literature about dreaded complications of HSE presenting as Klüver-Bucy syndrome at times even Korskoff’s Psychosis in the survivors (6, 7, 8, 11, and 14). When the clinical features of the patient was analysed we could see the involvement of frontal and temporal lobe with limbic involvement. Symptoms like dis-inhibition, poor performance on planning, poor social insight, inappropriate behaviour are marker of frontal lobe especially orbito-frontal cortex (disinhibition); dorso-lateral prefrontal cortex (dysexecution) (15). Unlike that of change in memory, affect and eating behaviour whichseems to be related to temporal lobe involvement.

The limbic involvement is substratianted by difficulty in processing, integration, inhibition and recall of stimuli (16). These findings from history, lobar function test and neuroimaging conforms the predilection of HSV for the frontal and temporal lobes of the cerebrum (12, 9, 10). HSV is a potentially treatable viral infection but rate of mortality and morbidity increases with delay in treatment onset and untreated cases (70%) as evident in our patient. Hence early diagnosis and management with acyclovir is mandatory (6, 7, and 8). Poor prognostic markers are age more than 30 years, long duration of illness, deep coma before initiation of therapy, and delay or non-use of acyclovir. It should be strongly suspected in any patient with a clinical presentation suggesting encephalitis. For the behaviour disturbances no specific treatment documented, still few drugs that are used are oral Carbamazepine, haloperidol, other atypical anti-psychotic and anticholinergics (8, 13) - as in our patient there was good response to risperidone, for symptom management. Along with pharmacotherapy for behavioural disturbances, there is need of therapeutic assistance for rehabilitation of selected cognitive process and behavioural modification of pathological behaviours because of permanent nature of damage caused by the illness.

CONCLUSION
Klüver-Bucy syndrome is a neuropsychiatric disease, where the intensity of personality and behaviour disturbance does not correspond to neurological deficit. With multiple aetiology, early diagnosis and treatment of treatable causes like HSV can prevent dreadful complications, like in the present case. Cognitive rehabilitation can be of help in long run with pharmacological management for pathological behaviours.

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