

CASE REPORT**A Multi-Disciplinary Team Approach to Klüver-Bucy Syndrome:
A Case Report**

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Abstract

Klüver-Bucy syndrome was first diagnosed in humans in 1955, after a group of people who had experienced temporal lobectomy. It is a rare neuropsychiatry disease and of which little is understood about its pathophysiological processes. Here we present a 60-year-old man seen in the outpatient psychiatric department in a tertiary hospital in Kuala Lumpur who presented to us with hyper sexuality, impulsivity, docility, amnesia and hyperphagia for the past 10 months. He was diagnosed with Herpes Simplex Virus with encephalitis 18 months ago and was shown to have bilateral meningoencephalitis of his temporal lobes. Thus, a diagnosis of Klüver-Bucy was made. We have taken a multi-disciplinary team approach to treat his illness and specific goals has been laid out in each discipline. A written consent has been taken by the patient and his family for the publication of this report.

Keywords: Klüver-Bucy Syndrome, Herpes- Simplex, Malaysia, Encephalitis, Hyperphagia

Introduction

Klüver-Bucy syndrome is a rare behavioral impairment which is linked with damage to both anterior temporal lobes of the brain. It causes patients suffering from it to have hyperphagia, hyperorality and hypersexuality. Other symptoms may include visual agnosia, docility, amnesia, and seizures. Brain injury and herpes encephalitis may further complicate this disorder, which may then lead to permanent brain damage [4].

Case Report

Mr A, a 60-year-old married gentleman presented with high fever in March 2016. He was diagnosed to have Herpes Simplex Virus at that time based on the clinical symptoms as well as the lumbar puncture findings which showed the detection of Herpes Simplex Virus 1, with a viral load of 127,248 copies/ml. However, he developed a complete change in behaviour since May 2016, 2 months after the onset of Herpes Simplex Virus. He had an increase in libido and was constantly asking for sexual favours

from his wife, much to his wife's dismay, and thus putting a strain in their marriage. He also had an abnormally great desire for food which resulted in overeating and especially an increased affinity to sweet food. This resulted in him being caught stealing food from supermarkets and other food stalls for which his family had to repeatedly show proof that he was not well. His family also complained of him having temper tantrums, childlike behaviour and was disinhibited. His family reported him to have poor judgement and impulsive; constantly being dared to do silly things by his friends, of which he abides. He also had poor sleep and problems with his memory. His functionality was also deteriorating, and has stopped working for the past 1 year, since the behavioural changes were noted after the infection. He must be reminded to perform daily activities such as bathing and practising his general self-hygiene.

Mr A has undergone several blood and radiological investigations. He first had a Computed – Tomography (CT) Brain done in June 2016 which showed prominent meningeal enhancement with no dominant focal mass and no evidence of intracranial haemorrhage. He then had a Magnetic Resonance Imaging (MRI) done in July 2016 which showed progressive cerebritis and meningoencephalitis in the temporal lobes bilaterally, right more than left. There was also a small rim-enhancing lesion in the right hippocampus. He also had another CT-brain done in May 2017 which showed hypodense right temporal lobe with reduced brain volume might be due to previous infarction or encephalomalacia from previous infection or trauma. Apart from that, his electroencephalography (EEG) report in May 2017 showed left fronto-parieto- central epileptiform discharges were seen, which may suggest the focus of the epileptic discharge or underlying structural

lesion. Moreover, he had blood investigations done in May 2017 which revealed he had hyperlipidaemia and borderline diabetes mellitus. The other blood parameters were unremarkable. He was referred to the medical department for his metabolic parameters.

Occupational therapy assessment of Activities of Daily Living revealed Mr A was able to perform most of his duties but lacked in hygiene and safety issues. Based on The Lawton Instrumental Activities of Daily Living Scale he only scored 2 out of 8 points, which shows a rather low functioning capability. He was unable to perform if there was no reward such as food present. He was able to manage his finances minimally based on colour and exchange. Another exciting find noted is that Mr A had problems with his sensory integration. He was only able to identify strong flavours of food based on his gustatory sensation. Main tastes identified were sweet and salty, he was unable to identify other taste sensations such as bitter, sour and umami. This issue with sensory integration could explain his affinity towards sweet biscuits. Besides that, he also had problems with his olfactory sensation, and couldn't identify things based on smell, which could suggest a sensory modality disorder and further testing needs to be done. The primary goals for this therapy would be to maintain his independence in activities of daily living and to modify his negative behaviour to positive behaviour as well as to improve interpersonal skills.

Aside from that, he is also being treated simultaneously by the Psychology team as well. Based on his Mini-Mental Status Examination (MMSE) he only scored 12 out of 30 which indicates severe dementia. He mainly lacked in the orientation, registration and recalled sections. Based on the Factor

analysis of the frontal systems behaviour scale (FrSBe) which is a short behaviour evaluation scale with proven validity for the evaluation of behaviour instabilities linked with injury to the frontal and subcortical brain regions, [14] he showed a mildly elevated before scale of 66 marks which indicates mild apathy. However, he showed a very much increased after the scale of 90 marks which is clinically significant and shows apathy, disinhibition and executive dysfunction. On the Dementia Rating Scale which used to measure cognitive impairment, he scored 100 out of a possible 144 points, which indicates a severely impaired level of cognitive functioning. The primary goal of psychological treatment was behaviour modification. Behaviours such as overeating and disinhibition were the primary targets. Cognitive stimulation therapy was applied to help him to develop healthier habits.

Mr.A was also treated by the neuropsychiatry team. A Neuropsychiatry Unit Cognitive Assessment Tool (NUCOG) [7] is a cognitive screening tool which covers 5 main domains that are attention, memory, language, executive and visuospatial function [16]. It was performed on him on the 14th of June 2017, in which he scored 44.5/100. This indicates severe cognitive impairment. He was prescribed oral Carbamazepine 200 mg twice a day and oral Zolpidem 10 mg on night. However, the oral Carbamazepine was increased to 400 mg twice a day on the next visit as his symptoms were only mildly improved. Subsequently, this dosage was maintained as his sexual behaviours, impulsivity and general irritability had reduced. This could be seen based on the Overt Behaviour Scale which is a tool used to document an array of challenging behaviours such as verbal and physical aggression, inappropriate sexual and social behaviour, repetitiveness,

absconding tendencies and lack of initiation displayed by people with brain injuries living in communities [8]. He had scored 37 points on the initial one done on 1st June 2017 and only 10 points on the latest one done on 27th July 2017, whereby majority of the reduction was in physical aggression and inappropriate sexual and social behaviour. However, a reduction of fear and overeating symptoms were still there and has largely contributed to hyperlipidaemia for which he was started on oral Atorvastatin 20 mg on night.

Discussion

Klüver-Bucy syndrome was discovered twice, first by Brown and Schäfer around 1887, and then by Klüver and Bucy in 1936 [3]. Klüver and Bucy, and predominantly Klüver, documented the condition as a chance to explain the role of the temporal lobe in behaviour [9]. This syndrome usually never occurs in isolation, rather it happens as a portion of an intricate behavioral syndrome that usually comprises of aphasia and amnesia, but of course may include some or all of its other components [11]. The most common presentation seen in humans besides amnesia and aphasia is hyperorality and emotional changes that include irritability, disinhibition and anger [13]. The top three causes of Klüver-Bucy syndrome are Bilateral temporal lobectomy or amygdectomy, traumatic brain injury and meningoencephalitis especially Herpes-Simplex Encephalitis [10] as seen in the case of Mr. A.

In view of this syndrome being so rare, and so complexed the treatment is often extremely difficult, and when even initiated it can be unsatisfactory. It possesses a great deal of difficulty, to both the patient itself as well as the family and community. Often, we see caregivers facing a burn out and may

become depressed themselves. A lot of the times, a full recovery is almost impossible due to the organic nature of this illness, coupled with the lack of insight from the patient himself. Hence, treatment must focus on an all rounded approach, one that deals with controlling patients symptoms, improving his daily activities, as well as a psychological approach. Another important factor which most physicians miss out is to give adequate support to caregivers.

Pharmacologically, multiple modalities may be used to treat the patient, from antidepressants to antipsychotics and mood-stabilizers depending on the main presentations. In this case, Mr. A mainly presented with symptoms that were affecting his mood in the sense of irritability, temper tantrums, increased libido and disinhibition. Hence, we opted to use a mood stabiliser for him. Pharmacologically, Carbamazepine is an antiepileptic, but it is also used to treat multiple psychiatric and organic disorders [1]. It has especially been proven to treat some symptoms especially hyper sexuality and impulsivity effectively in Klüver-Bucy syndrome [6]. Carbamazepine may influence the pathophysiology of organic disinhibition through reduced stimulation, enhancement by other mechanisms, or it may offer a better symptomatic relief than other medications [2].

Besides that, a combination of occupational therapy and psychological therapy is deemed to be useful to address the issues faced by patients suffering from this syndrome, such as inability to maintain his activity of daily living and maintaining his self- hygiene as well as improving his cognitive functions. Occupational therapy is also useful in this case, as Mr.A had problems with his gustatory sensation, and was having a high affinity for sweet foods. Apart from that, psychologically Cognitive

Stimulation Therapy was used on him which is an engagement in a range of activities and discussions intended at the general improvement of cognitive and social functioning [5].

In conclusion, Klüver-Bucy syndrome is an assemblage of neurological symptoms that results from a deficit in the temporal lobe [12]. Symptoms can prove to be very challenging to both the caregivers and treating doctors as it involves many different areas to treat. Its treatment is always best approached multifactorial with a multi-disciplinary team approach method as each approach has its own unique benefits to fully rehabilitate the patient well and to reduce caregiver burden.

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