Kluver – Bucy Syndrome Following Herpes Simplex Encephalitis

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Introduction

Kluver-Bucy syndrome (KBS) is a constellation of neurological signs which include hypermetamorphosis (compulsive exploration), hyperorality, reduced aggression, hypersexuality and visual agnosia. The syndrome is seen in pathological states which destroy anterior and medial temporal lobes bilaterally. Head injury, Central Nervous System (CNS) infection and degenerative diseases are the common causes. We describe a patient who developed Kluver-Bucy syndrome following Herpes simplex encephalitis.

Case Report

A 21 year male with history of continuous fever of four days, headache and vomiting of two days and altered sensorium of one day was found to be confused and disoriented with temperature of 102 deg F, pulse of 110/minute regular, respiration of 24/min and blood pressure of 120/70 mm. There was no skin rash, lymphadenopathy, pallor or cyanosis.

CNS examination revealed a disoriented patient, not responding to verbal commands, withdrawing all limbs to noxious stimuli, eyes opening spontaneously but not maintaining eye contact. The pupils were normal sized and reacting and he had global aphasia, bilateral symmetrical brisk deep tendon reflexes, absent abdominal reflexes and bilateral upgoing plantar response. There were no involuntary movements and no signs of meningeal irritation. The optic fundi were normal.

Blood hemoglobin was 14.0gm%, TLC 9000/cu mm, DLC P 64%, L 28%, M 02%, E 04%, no malarial parasites were seen, blood culture was sterile after 48 hrs, urine routine and microscopic examination was normal, blood sugar random was 90mg%, BUN was normal, serum ALT 50 IU/L, serum AST was 48 IU/L, ECG was sinus tachycardia, X-ray chest showed clear lung fields, CSF was normal pressure and clear appearance, proteins were 30mg%, sugar 48mg%, cells 50/cu

mm, predominant lymphocytes, microscopic examination for bacteria, mycobacteria and fungus was negative. MRI revealed hyperintensities on T2 weighted images in both frontal and temporal lobes suggestive of Herpes Simplex Encephalitis (Fig 1).

He was managed with intravenous acyclovir 30mg/kg and supportive therapy. Patient became afebrile forty- eight hours after starting therapy, was alert and responsive to verbal commands but remained aphasic. Over the next seven days he began exhibiting strange behavioral abnormalities as follows: (1) could move all limbs but he made no attempt to get up from bed. (2) started pelvic thrusting movements, which increased whenever anybody approached him. (3) became hyperphagic and began biting objects including a thermometer and chewed his own thumb. (4) was unable to sustain attention. An EEG done showed periodic bursts of slow waves in the left fronto temporal region. Carbamazepine 200 mg 8 hourly was added which reduced hyperphagia and pelvic thrusting movement. Patient remained bed-ridden with aphasia and did not make further improvement over next one month and was subsequently lost to follow up.

Discussion

In 1939, H Kluver and PC Bucy discovered peculiar behaviour in monkeys who under went bilateral temporal lobectomy [1]. The monkeys became docile, hyperphagic and hypersexual (especially male monkeys) and displayed visual agnosia (“psychic blindness”) distraction. Human KBS has been described and about 200 cases have been reported in literature. The complete syndrome is rarely seen in humans. The well known causes of human KBS are head trauma, encephalopathy, encephalitis, subarachnoid hemorrhage, Alzheimer’s disease, Pick disease, bilateral temporal infarction, porphyria, hypoglycemia, adrenoleukodystrophy and neuroleptic medications [2].

Damage to the amygdala results in flattened

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Fig. 1 : MRI T2 weighted image, showing bilateral temporal lobe hyperintensities.

emotions. Patients with bilateral amygdala damage cannot differentiate between relevant and irrelevant information [3]. KBS animals are unable to identify the biological significance of stimuli with the result that they cannot discriminate between food from non-food items which is known as psychic blindness [4]. Herpes simplex encephalitis (HSE) is the most common infection causing KBS due to predilection of the virus to selectively affect the temporal lobes. Ninety percent of HSE is caused by herpes simplex virus type 1. Diagnosis is suggested by fever, focal neurological signs (aphasia, complex partial seizures), T2 hyperintensity on MRI, which is due to inflammatory edema [5] and abnormal CSF. Diagnosis is confirmed by isolating viral DNA from CSF by PCR amplification. While PLEDs are suggestive of HSE, they

are not diagnostic [6]. Acyclovir is specific therapy for HSE, but is most useful if given before the patient becomes comatose [7]. KBS can be partially controlled with drugs like Carbamazepine [8] and medroxyprogesterone, which decreases sexual drive. No large studies have been carried out to determine the exact incidence of KBS in HSV encephalitis. A limited study of 19 cases revealed 50% mortality with one third of the survivors developing severe neurological deficits like Kluver Bury syndrome and Korsakoff's psychosis [9].

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Answers to MCQs

1. d	2. c	3. d	4. e	5. e
6. b	7. a	8. b	9. c	10. b
11. c	12. b	13. b	14. a	15. d
16. b	17. b	18. a	19. b	20. b
21. d	22. d	23. d	24. a	25. d
26. a				