Case Report

Kluver-Bucy syndrome developed after convulsion: A case report

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Abstract. Kluver-Bucy syndrome is characterized by increased appetite, hypersexuality, hypermetamorphosis, memory disorders, visual agnosia, stagnancy, aphasia, bulimia, polyuria, and polydipsia. A 14 year old girl had generalized tonic-clonic convulsions at admission, and an incomplete Kluver-Bucy syndrome with hypersexuality, recent memory disturbance, hypermetamorphosis, speech disturbance, hyperactivity, agitation, aggressiveness, and hallucinations, developed the following day. Here in, we report a case of KBS in a child with epilepsy.

Key words: Child, hypersexuality, hallucinations, agitation, epilepsy

1. Introduction

Kluver-Bucy syndrome (KBS) is characterized by increased appetite, hypersexuality, hypermetamorphosis, memory disorders, visual agnosia, stagnancy, aphasia, bulimia, polyuria, and polydipsia (1). Kluver and Bucy was first described KBS, in 1939, as a neurobehavioral syndrome in rhesus monkeys in whom rhinencephalon and bilateral lobes were rejected (2). Terzian reported a case of KBS after bilateral temporal lobectomy as the first human case (3). The potential causes of KBS include Herpes simplex encephalitis, head trauma, subarachnoid hemorrhage, epilepsy, bilaterally thalamic infarct, bilaterally temporal lobe resection, glioblastoma, shigellosis, Pick disease, adrenoleukodystrophy, hypoglycemia, Reye syndrome, Alzheimer disease, porphyrinuria, anoxic-ischemic encephalopathy, neurocystocercosis, tuberculosis meningitis, arachnoid cyst, Huntington chorea, Parkinson disease, Systemic lupus erythematosis, carbon monoxide intoxication, leukoencephalopathy associated with methotrexate, radiation, and stroke (4).

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In this report, we describe a case of KBS in a 14 year old girl with epilepsy who presented with incomplete Kluver-Bucy syndrome with hypersexuality, recent memory disturbance, hypermetamorphosis, increased appetite, speech disturbance, hyperactivity, agitation, aggressiveness, and hallucinations.

2. Case reports

A 14 year old girl with epilepsy was admitted to our emergency department. She had periodic convulsions once every 15 days in the last year. On the day before admission, she had had at least three generalized tonic clonic convulsions and each had continued for 15 minutes. She had no fever, headache, vomiting, personality change, head trauma or intoxication in her history. She had stopped taking her antiepileptic drugs (valproic acid and carbamazepine) without any reason two weeks before admission. There were no pathological features in her family history.

On physical examination, the patient's general condition was moderate and vital signs were normal. On neurological examination, she was lethargic, the pupils were myotic, pupil reflexes to penlight were positive bilaterally, deep tendon reflexes were normal bilaterally, and plantar response was flexor bilaterally. There were no signs of meningeal irritation.

Laboratory tests revealed the following: hemoglobin 12,8 g/dL, hemotocrit 36%, white blood cell 18230/mm^3 and platelet count was 348000/mm^3. Cerebrospinal fluid protein was 14

g/dL, glucose 78 mg/dL, chloride 127 mEq/L, and there were no cells in cerebrospinal fluid by direct microscopic investigation.

Electroencephalography showed an epileptic anomaly in the left hemisphere. Cranial computed tomography scan and brain magnetic resonance imaging were both normal excluding organic lesions such as brain tumors, subarachnoid hemorrhage. Anti epileptic therapy with valproic acid and carbamazepine was restrated. Next day, the patient experienced hypersexuality, memory disturbance, enlarged hands and feet described as hypermetamorphosis, increased appetite, speech disturbance, hyperactivity, agitation, hallucinations, and aggressiveness. With these signs and symptoms, she was diagnosed with KBS and haloperidol was started. The patient was discharged from the hospital on the fourth day, with anti-epileptic drugs. She is still being followed without epileptic seizures or signs and symptoms of KBS by the pediatric neurology department.

3. Discussion

KBS is a rare condition that may occur after temporal lobe trauma. Clinically, visual agnosia, hypermetamorphosis, increased appetite, memory disturbances, and bulimia can be seen (1).

Ictal genital automatism with postictal KBS symptoms are mostly seen in patients who have bilateral temporal convulsive activity (5). Postictal nasal whipping is seen in about 46-51% of patients with temporal lobe epilepsy, while it is seen in 10-12% of patients with extratemporal epilepsy (6). Although the exact mechanism of KBS is not clear, injury of the amygdala is often seen with these symptoms (7). Varon et al. reported KBS after partial complex seizure in a patient who had no structural temporal lobe injury (8). Our patient had symptoms including hypersexuality, hyperorality, increased appetite, recent memory disturbance, agitation, aggressiveness, and hallucinations during follow up. With these sings and symptoms, she was diagnosed as KBS which developed after convulsion.

KBS can be seen in all conditions related to temporal lobe, like herpes simplex encephalitis of the temporal lobe, hypoxia, temporal lobe epilepsy, cerebrovascular conditions, and cranial trauma. Generally, recovery is seen 1-3 months after acute disease, but this period may be shorter or longer (1-9,10).

Wong at al. reported a case of KBS that occurred after encephalitis with poliuria, polidipsia, narcolepsy, personality changes, amnesia, and hypersexuality (11). Yilmaz et al. has reported a child with incomplete KBS developed during acute encephalitis (12).

Neuroleptics, carbamazepine, dopamine blockers, benzodiazepines, and fluvoxamine can be used in treatment of KBS (2,3-13). Our patient was started on haloperidol therapy for behavioral changes, and valproic acid and carbamazepine were continued. A month after discharge, the patient recovered completely and convulsions did not recur. Haloperidol therapy was stopped and the anticonvulsive therapy was continued without any change.

In conclusion, we would like to emphasize that patients with epilepsy should be closely followed for KBS especially in patients with hypersexuality, hallucinations, hypermetamorphosis, increased appetite, memory disturbance, speech disturbance, hyperactivity, agitation, and aggressiveness. Overall the prognosis of the syndrome depends largely on the underlying pathology.

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