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An Impulsive Child- An Unusual Syndrome, A Common Cause, Uncommon Sequealae

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A 14-year female, 9thstandard student,came with complaints ofseizures for two and a half years, behavioural disturbances for four months and memory disturbances for two months. Her first seizure occurred whileenroute to home from school and was witnessed by friends and auto driver. There wastonic posturing of all four limbs, staring look, tongue bite and frothing.It was followed by postictal confusion, and the entire episode lasted for 15 minutes.

Her second episode of seizure occurred the following day in the railway station while waiting for the train. It began as tingling of right upper limb and lowerlimb, followed by posturing of all four limbs. There was postictal confusion, but no tongue bite or frothing. The entire episode lasted 15mins and was associated with weakness of right UL and LL, which later improved over a few hours.

She was prescribed antiepileptic drugs after the two episodes and was seizure-free for more than a year when she again had her 3rd episode of seizure.This seizure was a generalized tonic-clonic seizure with postictal confusion, but no tongue bite or frothing. Following this episode, her parents have observed that she has had frequent staring episodes lasting for few minutes occurring at 1-2 times a day. During this episode, she cannot recognize the person or surroundings and stares blankly. There is no history of myoclonic jerks, nocturnal seizures, or history of febrile seizures in childhood.There is no family history of seizures.

She had behavioural disturbances for the last four months, in the form of being very aggressive and abusing her friends, brother and parents verbally and physically. There was a history of asking the same question repeatedly or saying the same sentence repeatedly. She became very stubborn and was demanding in nature with stereotyped behaviour patterns. There was a tendency to keep non-edible objects into the mouth and on condemningthis act,she used to become even more aggressive.Shehad hyperphagia and developed certain food faddisms with a preference for sweet foods leading to weight gain.

Her parents noticed that her academic performance in the school declined. On enquiring further, the patient complained that she was not able to memorize what she has read and also faced problem in paying attention in the classes. The decline in memory and intellect lead to a poor academic performance, which caused depressed mood and crying spells occasionally.

Following the above changes in her mood and behaviour, her parents consulted a psychiatrist, and she was prescribed antipsychotic drugs. However, her behavioural disturbances continued to worsen further. Of late, she had difficulty in maintaining personal hygiene, as it was difficult for her to perform brushing, bathing etc.

She was subsequently admitted to our department. During hospitalization too, she was aggressive and abusive towards her parents, other patients, nurses, and doctors.She even tried to hit the nurses while giving injections and hit the doctors while trying to examine.

There was no history of any cranial nerve symptoms, motor weakness, sensory complaints, incoordination, and bowel or bladder incontinence. Moreover, there was no history of any headache, fever, or vaccination.

Her past history was insignificant, and she received antiepileptics and antipsychotics in the past. She was born of non-consanguinous parentage, full-term normal vaginal delivery, attained all milestones on time. She takes a mixed diet with regular bowel and bladder habits.

On examination, she was obese,BMI 30.76 kg/m^2.She was well dressed, with aggressive behaviour, using abusive language. Her MMSE was 20/30, and she lost points in attention, calculation(5),recall(3), orientation to time(1) and place(1).On lobar function test, frontal, temporal and occipital lobes were affected. On language functions, fluency and repetition were affected.

Frontal lobe tests revealed decreased attention, impaired judgement, abstract thinking,with preserved insight. Random-A test showed omission errors,and she could not perform the motor Luria test.

Temporal lobe examination showed impaired immediate,recent memory and new learning ability with preserved remote memory. Her parietal lobe functions were within normal limits. Occipital lobe examination showed prosopagnosia, with normal visual fields and colour vision.

Cranial nerve examination was within normal limits. Motor system examinationrevealed normal tone, bulk and power of both upper and lower limbs. Deep tendon reflexes were present (1+ in both upper and lower limbs),and plantars were flexor.Other superficial reflexes were preserved. Examination of the sensory system, cerebellar function, gait, spine, and cranium was within normal limits.

With the above symptoms, a possibility of limbic encephalitis, temporal lobe epilepsy and Kluver-Bucy syndrome was considered. She was investigated accordingly. MRI brain showed subnormal sized left hippocampus with a paucity of digitations and mild dilatation of adjacent temporal horn.PET – CT revealed hypoperfusion in the left cerebral hemisphere, right cerebellar hemisphere and also in the left parieto-occipital gyrus.It also showed left hippocampus subnormal in size compared to the right.

Biochemical CSF analysis was normal. CSF viral panel was positive for HSV-1 and HSV-2. Electroencephalograph showed slowing in the parieto-occipital region on the right side and spike and sharp wave discharges in the left posterior temporal region.

She was treated withantiepileptics(carbamazepine), antipsychotics(quetiapine), and antivirals(acyclovir).Though initially she seemed to improve, laterher condition did not improve, and behavioural disturbances worsened further. An autoimmune panel of CSF was sent at this time, and it came positive for LGI (Leucineglioma inactivated protein) antibody. Following this, she was started on intravenous immunoglobulin, and her condition improved drastically. She was back to near normalcy and was able to attend school, carry on her activities of daily living.

I. Discussion

We describe here a case of limbic encephalitis with Kluver-Bucy like presentation secondary to herpes simplex encephalitis, triggering an autoimmune response to LGIantibody.

Klüver and Bucy[1,2] described the following peculiar symptoms in rhesus monkeys after the removal of both temporal lobes including the uncus and hippocampus: 1) psychic blindness, 2) oral tendencies, 3)hypermetamorphosis, 4) emotional changes, 5) changes in sexual behaviour. The syndrome in humans occur following bilateral temporal lobectomy, Alzheimer's disease, Picks disease,viral encephalitis, trauma,paraneoplastic encephalitis, andtoxoplasmic encephalitis[3,4].

Partial KBS is diagnosed based on the presence of three or more of the aforementioned symptoms.

Our patient had oral tendencies, visual agnosia, emotional,behavioural disturbances, memory disturbances and language disturbances, thus fulfilling as partial Kluver-Bucy syndrome. Kluver-Bucy syndrome is usually accompanied by dementia, aphasia or disturbances in language function. Partial Kluver-Bucy is more common in humans and has the same localizing value as complete form with affliction of the medial and anterior temporal regions.

As the disease progressed, it evolved into a partial Kluver-Bucy with limbic encephalitis, with waxing and waning features which cautioned us to work up for an autoimmune aetiology. Thus the viral infection could have triggered autoimmune limbic encephalitis.

This case highlights the clinical phenomenology of Kluver-Bucy syndrome in autoimmune limbic encephalitis triggered by a viral infection and the reversibility on managing it appropriately.

II. Summary

1. Kluver-Bucy syndrome should be kept in mind in the differential diagnosis of patients who present with visual agnosia,hypersexuality,behavioural disturbances, hypermetamorphosis.
2. Kluver-Bucy syndrome usually manifests along with other behavioural, memory, emotional disturbances, stereotyped motor acts, language disturbances. The presence of these symptoms can obscure a diagnosis of Kluver-Bucy syndrome.Hence, a high index of clinical suspicion is needed to make the diagnosis. This could be one reason for under-reporting of these cases.
3. Kluver-Bucy syndrome can develop after a latent period of 2 to 4 weeks or longer[6].In our case, the symptoms of Kluver-Bucy (hyperorality, emotional changes, fearlessness) developed after, more than a year after the first seizure episode.A similar observation was made by Shoji et al[5]. The reason for the delay is uncertain but could suggest a spread of the virus from temporal lobes to limbic areas.
4. The chronicity of the symptoms, behavioural, and psychiatric changes, coupled with changes in MRI could be tempting to label these patients as a case of psychiatric illness or temporal lobe epilepsy. Thus it is essential to exercise caution in such cases, avoiding delay to arrive at the correct diagnosis for timely intervention.
5. An autoimmune limbic encephalitis could evolve in the cases over days to months mimicking medial temporal lobe epilepsy.This could be due to triggering of autoantibodies, especially to LGI,which interact with epilepsy-related channels.The presence of such antibodies warrants immunotherapy as it is highly treatable.

6. Management involves mood stabilizers, antipsychotics and antiepileptics primarily. In cases where an autoimmune reaction is triggered, IVIG therapy is useful.

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