A CASE OF POSTICTAL PSYCHOSIS WITH INTERICTAL BEHAVIOURAL CHANGE SUGGESTIVE OF GESCHWIND-GASTAULT SYNDROME IN TEMPORAL LOBE EPILEPSY - CASE REPORT
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Abstract: Postictal psychosis is one common entity that often complicates chronic epilepsy, in patients with severe seizure clusters that include tonic clonic seizures, bilateral cerebral dysfunction and complex partial seizures especially temporal lobe epilepsy. Similarly as reported by Norman Geschwind in 1974, interictal behavioural syndrome in temporal lobe epilepsy (TLE) has features of hyper religiosity, hyposexuality, hypergraphia, humourlessness. In subsequent studies these features have been found not to be entirely specific to TLE and the presence of this specific entity is questioned later. Here we present a case of a 36 year old male with a unique presentation of postictal psychosis with interictal hyper religiosity, circumstantial speech that was suggestive of Geschwind syndrome in TLE.

Keyword: postictal psychosis, temporal lobe epilepsy, Geschwind syndrome

INTRODUCTION:
Psychiatric presentation in epilepsy could be classified into disorders strictly related in time to seizure occurrence, interictal psychiatric disorders and disorders clearly attributable to brain pathology causing epilepsy(1). Among the disorders having temporal association to seizure occurrence it could be related to preictal, ictal and post ictal states which includes various presentation like psychosis, depression and anxiety as mentioned in ILAE proposal(1,2). Psychoses in epilepsy in particular could be closely linked to seizures (ictal or postictal psychosis), those linked to seizure remission (alternative psychosis), psychoses with a more stable and chronic course (such as interictal psychosis), and iatrogenic psychotic processes related to antiepileptic drugs (3,4). Vuilleumier and Jallon found that 2-9% of patients with epilepsy have psychotic disorders(1,4,5). Perez and Trimble reported that about half of epileptic patients with psychosis could be diagnosed with schizophrenia(6). As per ILAE proposition among the post ictal disturbances postictal delirium and postictal psychosis are the most common aspects seen in the patients. Even though described as early as 1836 by Esquirol as ‘postictal fury’, it was only after a century of neglect that Logsdail and Toone in later part of 1980 introduced the criteria of postictal psychosis and emphasized its common occurrence and the need for treatment(1,7,8). Postictal psychosis constitutes about 25 to 30 percent of psychoses of epilepsy(7). Prevalence rate of around 6% among the epileptic cases has been reported in two elementary studies(1,9,10). The most consistent risk factors include bilateral widespread CNS injury, encephalitis, EEG slowing, seizure cluster that includes tonic-clonic seizure and presence of GTCS with TLE focus (1,9,11). The psychosis is characterised by plenomorphic picture with fluctuating combination of thought disorder, auditory and visual hallucination, delusions (grandiose, religious, persecutory) with a lucid interval of 2 hours to a week (usually over 6 hours) after seizure onset. Psychosis usually lasts for at least 15 hours and less than 2 months(1,7,8). Religious and violent behaviour can be prominent in post ictal psychosis and various reports of suicides have been reported(10,11). The mean age of onset is 32-35 years with the first psychotic presentation occurring at an average of 15-20 years after seizure onset(1,4,11). Logsdail and Toone’s criteria as mentioned below gives a clear set of guidelines for diagnosing postictal psychosis:

1] Episode of psychosis (often with confusion and delirium), developing within 1 week of a seizure or cluster of seizures
2] Psychosis lasting at least 15 hours and less than 2 months;
3] Mental state characterized by delirium or delusions (e.g., paranoid, non paranoid, delusional misidentifications) or hallucinations (e.g., auditory, visual, olfactory) in clear consciousness;
4] No evidence of:
   a) History of treatment with antipsychotic medications or psychosis within the past 3 months,
   b) Antiepileptic drug toxicity
   c) EEG demonstrating nonconvulsive status,
   d) A recent history of head trauma or alcohol/drug intoxication or withdrawal (other than benzodiazepines used for epilepsy).

EEG obtained at this period shows diffuse background slowing or increase in inter ictal epileptiform abnormalities(4,8,10–12). Post ictal confusion is another common entity associated with seizures. While epileptic seizures begin abruptly, recovery of normal function is usually gradual. It starts from the completion of seizure to full recovery of normal function. It may take few minutes to few hours in certain cases, usually characterised by confusion, drowsiness and head ache. In complex partial seizure
altered sensorium is itself part of seizure. It may itself be mistaken for mental confusion and disorientation. Symptoms like automatism, fugue like state seen in complex partial seizure could be further mistaken for behavioural problems. In complex partial seizure the transition from state of unresponsiveness to alertness take place over minutes whereas in GTCS recovery would be protracted to about 15-30 minutes. It is difficult to differentiate postictal confusion and postictal psychosis in a case of complex partial seizure. As per the Logsdail and Toone's criteria 'lucid interval' between termination of seizure activity and onset of psychosis clearly differentiates both condition(1,4). Similarly, while considering interictal personality change, there exists a debatable question as to whether any typical personality traits specific to epileptic patients have been observed. In 1975, Waxman and Geschwind proposed a syndrome of interictal behavioural changes which they believed to be common in TLE(13).

Geschwind –Gastaut syndrome, as it came to be known, comprised alterations in sexual behaviour (principally hypo sexuality), religiosity and a tendency towards extensive, and in some cases overt emotional traits (deepened emotionality, sadness, hypermetabolism), whereas those with left temporal foci showed ruminative intellectual tendencies (religiosity, philosophical interests, humourlessness, sense of personal destiny)(11). Further studies using BFI also reported the syndrome to be not very specific when compared with patients with other form of epilepsy, and mere existence of such separate entity came for questioning (1,4,6). Nevertheless, till date certain graphia), found to be more commonly occurring in patients with TLE even though they are not specific to them. With this brief introduction, we proceed to our current case scenario, where both aspects have been seen in the individual leading to a unique clinical presentation.

CASE REPORT:
Mr. A, 36 years old unmarried male, studied till 10 th standard, working as a painter, from urban background was brought to psychiatric OPD with the complaint of episodes of aggressive and violent behaviour associated with hearing voices of God, claiming to have power of God, followed by wandering behaviour on and off for past 4 years. His detailed history revealed that he was born out of a normal delivery. In early and middle childhood, he had no significant disturbance in early and middle childhood. He had no significant complaint of hearing voices subsided within a week of onset. After a week he remained confused, restless, pacing in the room, giving irrelevant replies and not comprehending his mother’s requests. After this 2 hours period he himself laid down, took the diet given to him, answered relevantly when questioned and complained of severe bilateral throbbing headache with no associated physical complaints. He was able to recognize his mother and other family members. He remained confused and disoriented. He was laid in the bed and appeared well oriented as inferred from his answers and behaviour during the next 6 hours. Later after that, in night time his mother noticed him to behave in an odd manner as he was kneeling down in front of Lord Jesus portrait, holding the Bible in his hands for about an hour. On questioning he mentioned he could hear the voice of lord conversing to him and claimed to have power of the lord. He was found to be extremely irritable, agitated and mentioned he has to ‘purify the world, as commanded by the lord’. Following this he left the house and his whereabouts were not known for next 3 days. After 3 days he returned back with no clear details of his whereabouts and mentioned he followed the voice of Lord Jesus. His complaint of hearing voices subsided within a week of onset. After a week he started going to work and maintained well with minor symptoms being noticed, not aware of the events happened during the episode clearly. After 1 month, again while he returned from work in evening hours he was found to be dull, slow in his activities and remained seated for about an hour with frequent rubbing of nose and lip smacking movements. This was associated with unresponsiveness and followed by sudden fall with loss of consciousness for few minutes and urinary incontinence at that time. After about 30 minutes of confused and disoriented behaviour he remained quiet, took diet, answered relevantly with no symptoms for next few hours. Later again in midnight he behaved in abnormal manner such as kneeling in front of God’s portrait for a long time at midnight, mentioning about hearing voices of Lord Jesus conversing to him and claiming to have power of the Lord. He remained responsive to stimuli but was extremely irritable and aggressive mentioning he has to ‘purify the world’. After this he wandered away for 2-3 days with no clear details of his whereabouts, saying that he followed the voice of Lord Jesus. After returning home his complaint of hearing voices and aggressiveness subsided completely within 7 days.
Following this he continued to have such episodes once in a month with similar presentation and symptoms subsiding within 7-10 days either with symptomatic treatment or without treatment. In the workplace also, he used to behave in an odd manner like mixing paints inappropriately, splashing paints on painted walls. Following these acts, he would leave from there without answering anyone and being totally unaware of such happenings later. Exact duration and frequency of such behavior in work place was not known clearly. His GTCS reduced in frequency but he continued to have tonic clonic movements following few such episodes about once in every three months. Initially he had such behaviour once a month for about 2 years, later it progressed to more than 2-3 episodes every month in last 6 months with similar presentation. In between the period of such episodes for about 2-3 weeks period patient use to go to work regularly, maintained well with his day to day activities with no history suggestive of aggressive or psychotic symptoms. His sleep, appetite, self care was good and no such odd behaviour was noticed. As the episodes of such behaviour progressed following rules and orderliness in all his activities in home and work, in between the period for about 2-3 weeks, for about 6 months ago, he was forcefully admitted in a private hospital where he was found to be highly aggressive and agitated, expressing delusion of grandiosity and auditory hallucination. On the next day, without responding to others, he suddenly jumped from the 3rd floor of the hospital and couldn’t give any reason for the act. There were no signs of depressive features prior to the act. EEG taken at that time showed bilateral epileptiform discharges and within a week he was discharged as his symptoms had improved with antiepileptics and antipsychotics which he discontinued after discharge. In between the episodes, even though he maintained well in his work and daily routine as mentioned earlier, his family members noticed that he had become involved in reading the Bible repeatedly, preaching the values to others, making notes of verses repeatedly for which he used about 4-5 notebooks every month, frequently advising others to be true as mentioned in the Bible and avoiding his friends for the same reason. He also use to help school children with drawings voluntarily, and would give excessive details with biblical verses in his speech for most of the topics. He also advised the family members to stop looking for marriage proposals as he said was due to the purity in life which one was expected to achieve through the Bible. He also advised others to be true as mentioned in the Bible and avoiding his friends for the same reason. He also use to help school children with drawings voluntarily, and would give excessive details with biblical verses in his speech for most of the topics. He also advised the family members to stop looking for marriage proposals as he had no desire to lead a marital life. He was also found to be following rules and orderliness in all his activities in home and work, more strictly which was unusual of his usual behaviour. Even though patient maintained well with medications he discontinued the treatment for about a month. With continuing episodic pattern as described above and the change of certain traits in day to day activities in intervening periods, following one such episode when he tried to walk away, he was again forcefully brought to our OPD. He was found to be highly aggressive, irritable but oriented, with paranoid delusional ideation of grandiosity, circumscribed by sound and person auditory hallucination which was suddenly followed by an episode of GTCS. He was treated for GTCS and then got admitted in psychiatric ward after 3 days with persisting psychotic symptoms.

NEUROLOGIST OPINION:
Initially following GTCS patient got admitted in neurology ward. He was diagnosed as a case of complex partial seizure [temporal lobe epilepsy] with postictal behavioural disturbance and secondary generalisation of seizure at present. Past occurrence of primary GTCS was also recorded. His cranial nerve function, motor system, sensory system, cerebellar functions, autonomic system and cognitive functions were reported to be normal at the time of discharge. Bilateral epileptiform discharges were noted in EEG taken on second day and B/L mesial temporal sclerosis was seen on MRI. He was advised to continue the treatment with T.carbamazepine 200 mg 1-0-1; T.sodium valproate 200 mg 1-2-2; T.folic acid 1-0-0 with periodic review every month. Patient was discharged and referred to psychiatric OPD for persisting psychotic symptoms mainly. On admission in psychiatric ward his general examination was found to be normal and his systemic examination including cardiovascular and respiratory systems were normal. His detailed psychiatric examination was normal with MMSE score of 27/30. Fundi in both eyes were normal. His mental state examination revealed circumstantial speech, fleeting auditory hallucination, with restricted affect and no signs of grandiosity and irritability. He was not clearly aware of the events in preceding 3 days. His blood count, electrolytes, glycemic status, renal function including BUN, serum calcium level were normal and he was non reactive for VDRL, HIV antibodies. His cardiac status including ECHO, ECG findings were normal and his endocrine status was also found to be normal as per opinion obtained. Within a week his hallucination subsided, but his circumstantial speech, involving moral and ethical values from the Bible for all queries persisted. He had a sense of orderliness in his activities not amounting to obsession and expressed disinterest in sexual and marital life which he said was due to the purity in life which one was expected to follow as told in the Bible. EEG done again on the 10th day showed background slowing with no epileptiform discharges. Psychological assessment done later showed that he had adequate cognitive function, with an ambivert personality with traces of affective colouring, manic features and intact reality testing. Patient improved with antiepileptics along with low dose of antipsychotics and benzodiazepines. Figure 1: MRI brain of the patient with B/L mesial temporal sclerosis.

DIFFERENTIAL DIAGNOSIS AND TREATMENT:
While working up this case, various possibilities were considered before arriving at a diagnosis. Patient had initial presentation of primary GTCS and currently complex partial seizure with secondary generalisation of seizure. This was evident from objective finding of epileptiform discharges in EEG taken twice during such behaviour change and bilateral mesial temporal sclerosis in MRI. The behavioural change following the episode has to be considered now with following possibilities
1] Postictal confusion
2] Postictal psychosis
3] Complex partial seizure with interictal psychosis
4] Ictal transient Psychosis
5] Alternative psychosis (forced normalisation)

The semiology of events in each episode consists of automatisms, followed by loss of consciousness and an interval of adequate orientation which was followed by psychotic symptoms that resolved completely within 10 days. This suggests post ictal psychosis, because even during the expression of psychotic features, patient had adequate orientation as seen during admission and also by history. This rules out the possibility of post ictal delirium. Complete resolution of symptoms within 10 days and presence of psychotic symptoms only in subsequent episode, with a lucid interval of about 6 hours rules out the possibility of inter ictal separate psychotic disorder and favours postictal psychosis as per the Logsdail and Toones criteria(8). Long standing recurrent seizure and GTCS associated with TLE focus seen in this patient is one common associated risk factor for postictal psychosis. With reduction in GTCS following the current presentation, possibility of forced normalisation phenomenon was considered but the abnormal EEG changes persistently recorded in the episodes goes against it (18,19).
Regarding personality traits Geschwind and Waxman proposed certain traits to be specific to patients with TLE as mentioned earlier. This patient on BFI scored positive for traits of circumstantiality, hypergraphia, moralism, stickiness, hyposexuality, philosophical interest which is sufficient to diagnose Geschwind-Gastaut syndrome. Whether it was specifically related to his TLE or not remains a matter of debate(15).

TREATMENT AND FOLLOW UP:
With lack of specific diagnostic criteria for these entities, treatment guidelines have also not been well established. As mentioned in literature, patient was put on antiepileptics in liaison with the neurologist. Adequate dose of antipsychotic was added initially to reduce aggression and psychosis, which was then tapered slowly. On discharge his psychotic symptoms completely remitted and his aggression and psychosis, which was then tapered slowly.

Patient was further maintained on low dose antipsychotics along with antiepileptics in follow up. Other types of advice like need for regular medical check-ups, risk of working at heights (as patient was a painter) and risk of legal problems during periods of wandering was explained to patients and family members. Need for regular follow up was also explained as these cases have high chance of progressing to interictal psychotic disorder(20).

DISCUSSION:
Even though the patient has been diagnosed to have post ictal psychosis as per Logsdail and Toone’s criteria and Geschwind - Gestaut syndrome as per Bear - Fedo inventory, the current psychiatric classification of ICD-10 or DSM-IV does not include any such criteria or nomenclature. This has significantly hampered the diagnosis of this rather common condition and lead to misdiagnosis as had happened earlier in this patient. International League Against Epilepsy(ILAE)’s Commission on Psychobiology of Epilepsy has classified the neuropsychiatric disorders in epilepsy (2) and has given a detailed classification proposal to forthcoming ICD-11 and DSM-V for inclusion of these disorders, as lack of awareness has led to the condition being misdiagnosed or left undiagnosed, posing a serious threat, as studies have reported high suicide rate, severe aggression and violence proneness(9,11,21) during these periods which could be treated effectively, if diagnosed early. The presence of such entity has been questioned in current literature. We conclude this with a quote from Dostoevsky, a famous Russian writer, who said the following about his own epileptic seizures: "For several instants I experience a happiness that was impossible in an ordinary state, and of which other people have no conception. I feel full harmony in myself and in the whole world, and the feeling was so strong and sweet that for a few seconds of such bliss one could give up ten years of life, perhaps all of life. I felt that heaven descended to earth and swallowed me. I really attained God and was imbued with him. All of you healthy people don't even suspect what happiness is, the happiness that we epilepts experience for a second before an attack." Thus epilepsy has its own different presentation in each individual which has to be considered carefully while evaluating each patient.

REFERENCES: