CASE REPORT

Schizencephaly: Psychosis, a Rare Manifestation

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Abstract

Schizencephaly is an uncommon congenital malformation of the central nervous system which affects the development of the cerebral cortex. It is defined as the gray matter lined filled cleft with cerebrospinal fluid that extends from the pial surface to the ventricle. There exists a paucity in scientific literature regarding the association between schizencephaly and psychosis. We report a case of a 36-year-old male, presented with worsening crisis of aggression, disorganized behavior and auditory hallucinations over 20 years. CT brain scan revealed unilateral left open lip schizencephaly, a finding during his recent admission to the psychiatry ward.

Keywords: Schizencephaly, Malformation, Psychosis

Introduction

Schizencephaly is an anomaly of the neuronal migration due to insults to the migrating neuroblasts during the third to fifth gestational months [1]. There are 2 types of schizencephaly; Type 1 (closed lip) which does not communicate with the ventricular system, and Type 2 (open lip) which does communicate, as described by Yakolev and Wadsworth [2]. Such occurrences are incredibly rare with only 1.54 cases per 100,000 population [3]. Multiple causes have been implicated in its aetiology, including ischaemia, germline mutations, intrauterine infections, and exposure to drugs. Patients with this condition clinically present with developmental delay, seizures, and abnormal motor skills in those severely affected. The correlation between the malformation, seizures and developmental delay are well described, however only few cases of psychosis were reported [4] (Table 1).

Alexander RC, et al., reported 2 cases of schizencephaly that exhibited psychotic symptoms. The first case describes a woman with childhood seizures that displayed low mood, feelings of hopelessness, sleep and appetite disturbances, auditory hallucinations, together with religious and persecutory delusions. Her MRI findings disclosed bilateral closed lip schizencephaly. The second case involves a man with left unilateral schizencephaly having auditory hallucinations, persecutory delusions, thought insertion, plus withdrawal and suicidal ideations [5].

Tibrewal, et al., also reported 2 patients exhibiting similar symptoms; the first
patient, a 25-year-old male presented with persecutory delusions and auditory hallucinations, while the second patient was a 23-year-old female who experienced bizarre delusions, with poor sleep, irritability and social withdrawal. CT brain scans revealed left sided unilateral closed lip schizencephaly and right-sided parieto occipital closed lip schizencephaly respectively [1].

A further case of schizencephaly was also reported in Brazil with the patient displaying perceptual disturbances, disorganized behavior and persecutory delusions [4].

There have also been reports of schizencephaly related to bipolar disorder [6,7].

Our patient, who had a recent admission to the psychiatric ward shares some interesting similarities with the previous case reports. These similarities may shed some light on the possibility of a connection between the schizencephaly and psychosis.

**Case Report**

Mr K – a 36-year-old male, was brought into the outpatient department for setting fire to his own house and displaying aggressive behaviour towards family members. Furthermore, he also experienced auditory hallucinations, persecutory delusions and sleep disturbances. Prior to this, he has had multiple admissions to the psychiatric hospital mainly for irritability and aggressive behavior.

Mr K was diagnosed with epilepsy at the age of 8 when he started to have recurrent episodes of generalized tonic-clonic seizure. He was started on an anti-epileptic but defaulted subsequently.

Mr K’s first contact with the psychiatric unit was at the age of 16. He presented with aggressive and destructive behaviours together with auditory hallucinations. Since then, he has had multiple admissions for similar presentations. Frequent scheduled appointments to the psychiatric outpatient department showed poor seizure control despite strict adherence to medications and adequate medication dosage which were monitored by serum levels of the anti-epileptic. A typical anti-psychotic was also added to his treatment regime.

Mr K was born full term via vaginal delivery via a non-consanguineous marriage. His antenatal history was uneventful. He had delayed developmental milestones. It was noted that he only started walking at the age of 4, with learning difficulties becoming evident at the age of 9 when he stopped schooling.

There were no family history of mental health illness and history of illicit substance use.

His mental state examination revealed persecutory delusions and auditory hallucinations with a poor insight to his illness.

Neurological examination showed right sided facial asymmetry, right upper limb contracture and length discrepancy of the lower right limb.

Blood examinations, including full blood count, biochemistry and thyroid function tests were unremarkable. Mr K’s electroencephalography shows left focal epileptic discharge with probable underlying focal structural abnormality. CT brain (Figure 1) revealed a cleft at the left cerebral hemisphere extending from cortex to left
lateral ventricle. MRI brain was not done in view of restricted resources in the locality.

He was treated with Haloperidol 5mg per day during this admission. Carbamazepine 200mg twice daily was also given for seizure control. There were no episodes of seizure observed during his 2-month stay in the ward. Upon discharge, he only experienced residual auditory hallucinations which resolved during subsequent visits.

Figure 1. Head CT scan dated 29-Jul-2016

Table 1. Schizencephaly cases in literature

<table>
<thead>
<tr>
<th>Author(s)</th>
<th>No. of cases</th>
<th>Anatomical Anomaly</th>
<th>Diagnosis</th>
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<td>2</td>
<td>Unilateral &amp; bilateral schizencephaly</td>
<td>Psychosis</td>
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<td>Bipolar Disorder</td>
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<td>Komori K, Komori M, Hayashi J, 2006</td>
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<tr>
<td>Authors</td>
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<tr>
<td>Arora M et al, 2009</td>
<td>1</td>
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<td>Bipolar Disorder</td>
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<td>Melo MC et al, 2013</td>
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<td>Tibrewal P et al,</td>
<td>2</td>
<td>Unilateral closed lip schizencephaly</td>
<td>Psychosis</td>
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</table>

**Discussion**

Schizencephaly, meaning split brain is a rare congenital anomaly which affects the formation of the cerebral hemisphere in the first 3-5 months of gestation [8]. It was initially described by Yakolev and Wadsworth in 1946 [2]. The exact pathogenesis of this anomaly is unknown.

The highlight of this case is the significance of the link between schizencephaly and the psychotic manifestation. Schizencephaly often goes undetected due to overshadowing of other medical conditions such as epilepsy. It has been associated with psychosis with limited literature exploring the relationship in further detail.

The relationship between abnormal brain structure and manifestation of psychotic symptoms have been a topic of discussion over the years.

Epilepsy is present in about half the cases of schizencephaly but are not strictly related to the severity of the malformation as opposed to motor deficits and mental retardation [9]. Based on the available case reports, there are no definite set of symptoms that are pathognomonic of schizencephaly. In our case, Mr. K experienced persistent auditory hallucinations and delusions despite seizures being well controlled. This suggests a possible association between schizencephaly and psychotic symptoms.

The neuro-developmental anomaly may have a pathoplastic effect on the manifestation of psychosis and this may influence treatment response [6]. Low dose of typical anti-psychotic-in this case haloperidol, shows a good response in symptom control without the side effects.

More research is warranted to find a true link between schizencephaly and the emergence of psychosis. A better understanding of this rare disorder and how brain processes are affected may provide a broader perspective of the psychosis spectrum.

**References**


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