Porencephaly and psychosis: a case report and review of the literature

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Abstract

Introduction: Malformations of the cerebral cortex are often associated with developmental delay and psychoses. Porencephaly is a rare congenital disorder of central nervous system involving a cyst or a cavity filled with cerebrospinal fluid, in brain’s parenchyma.

Case presentation: We present a 25 years old woman with her first psychotic episode. She also suffers from porencephaly in the frontotemporal lobes region. It is emphasized that the two consistently abnormal brain regions in schizophrenia research had significant damage in this patient since her birth.

Review of the literature: There is a total of only five cases of schizencephaly or porencephaly and psychosis. Their clinical characteristics as well as the imaging results are described.

Conclusion: It is unclear if porencephaly and psychosis concur by chance or are causally related. The area where the porencephalic cysts appear seems to be of relevance. This case highlights the need for further research.

Key words: schizophrenia, porencephaly, temporal region

Introduction

Malformations of the cerebral cortex are often associated with developmental delay and psychoses. Porencephaly is an extremely rare congenital disorder of central nervous system involving a cyst or a cavity filled with cerebrospinal fluid, in brain’s parenchyma. It is caused by either a local damage in brains hemisphere from ischemia or hemorrhage after birth (most common). It can also occurs as the consequence of abnormal development before birth (less common cases) [1-2].
Specifically, congenital brain lesions include two types of porencephaly: genetic porencephaly, resulting from maldevelopment during early neuronal migration and encephalophaloplastic porencephaly, which is late prenatal or perinatal vascular lesion due to arterial ischemic stroke or venous thrombosis. Porencephalic cysts could be located in any lobe or lobes of two brain hemispheres [3].

Prefrontal cortex and limbic system hypotheses are the predominant neuroanatomical hypotheses of psychosis. The demonstration of decreased prefrontal gray or white matter volumes, or both, disturbed prefrontal metabolism and blood flow, decreased hippocampal and entothinal cortex volume, in psychotic patients, are some of the most strong indications that prefrontal cortical and medial temporal regions are associated with psychosis [4-6].

**Case Presentation**

A 25-year-old woman admitted via the emergency department of Psychiatry Clinic of Attikon General Hospital with psychotic symptoms. She was escorted with her mother and brother. She didn’t have any psychiatric history but the last three days, she was too frightened, she couldn’t sleep properly and said that people talked to her through the television set. On the day of her admission, she stayed home, closed the windows and called her husband at work, many times, asking for his help, because something very bad would happen. Her husband tried to calm her, but that was impossible. Finally, he went home and he found her lying in bed, she didn’t offer any explanations for her behavior.

On mental state examination, the patient was oriented in person, space and time. She displayed persecutory delusions that were related to violent incidences that occurred the previous days in Athens. There were no auditory hallucinations. Patient’s affect was blunted and she did not display any insight into her condition. She had psychomotor retardation and her speech was slow. At that time, she was calm, she told us she suffers from insomnia the last days and she was very co-operative, almost subdued. The neurological examination revealed right arm weakness (proximal 3/5, distal 2+/5 according to the MRC scale of evaluation), pyramidal type rigidity with increase of deep tendon reflexes, muscle atrophy and stereoagnosia of the same arm. The Mini-Mental State Examination was normal (30/30). PANSS (Positive: +17, Negative: -22, General Psychopathology: 35). A neuropsychological examination showed few alterations, with impairment of verbal memory, attention and ability to plan (WAIS: 64, Verbal: 69, Performative: 63).

According to her medical history, she suffers from porencephaly in the front-temporal lobes region since her birth. The patient had no speech development problems or severe mental
retardation which is often associated with porencephaly. However, she had a slight spastic paresis in her right arm.

A cranial MRI was performed (fig 1 and fig 2) in order to get a more detailed image of her brain. It confirmed the nature of the lesion and revealed the existence of a large porencephalic cyst on the left frontal and temporal lobes. The examination revealed also atrophy in the left side of brainstem due to wallerian degeneration.

Fig 1

Fig 2

The patient was not very social as a child; she had only few friends, however she managed to complete secondary education. She grew up and lived till her 23 years old in a small village. She had no personal relationships until she met his husband and left with him for Athens. She kept their house but her husband did all external jobs and never left home alone. Few days before her illness, they had a fight and the husband threatened to leave her alone.

She was started on antipsychotic therapy (olanzapine 10 mg). The psychotic symptoms improved progressively during the stay, with amelioration of the psychomotor retardation. It was clear that there was a significant improvement in relation to the delusional persecutory ideas and a complete remission of these symptoms was achieved. PANSS (Positive: +10, Negative:-18, General Psychopathology: 19). The patient was discharged after a 2-week inpatient stay and received follow-up outpatient care with psychiatric appointments.

Discussion –Review of the literature

The present case has clinical manifestation of porencephaly, i.e. mild mental retardation and spastic paresis in her left arm accompanied by motor impairment. However, this patient did not have speech development problems or seizures. The location of porencephalic cyst, in frontal and temporal lobes, is associated with psychotic features in this patient, or damaged brain regions possibly triggered the patient’s potential vulnerability to psychotic symptoms?
We reviewed the scientific literature for other cases with porencephaly and psychosis to investigate the above hypothesis. Dr Pae and Dr Kim this year report the case of a patient who developed a psychotic episode, possibly, associated with multiple leukoencephalomalacia and Porencephaly changes in the brain cortex. In this case, a 30-year-old woman had persecutory ideas and visual hallucination. Multiple focal leukoencephalomalacia changes in the left frontal lobe, bilateral occipital lobes and left basal ganglia, and a large porencephalic change in the right temporal lobe were found in magnetic resonance image (MRI) [7].

Schizencephaly is also a rare congenital neurodevelopment disorder of brain cortex which is characterized by abnormal slits or clefts in the cerebral hemispheres. That malformation is a form of porencephaly. It appears with paresis, mental retardation and seizures, too [8].

Alexander et al reported two cases with schizencephaly associated with psychosis. First patient had seizures, mental retardation and psychotic features like delusions and auditory hallucinations. Second patient had monoplegia, mental retardation and delusions [9]. Relan et al presented a case with unilateral schizencephaly cleft associated with biopolar disorder. This patient was overactive and oversocial. She also had mild motor impairment, mental retardation and appeared persecutory ideation and auditory hallucinations [10].

Cysts on the left temporal lobe have been associated with psychosis. Alves da Silva et al described the case of a patient associated with the presence of an arachnoid cyst in the left temporal lobe. He appeared psychotic symptoms as delusions and hallucinations [11]. Vakis et al reported a case of young woman with a left temporal lobe arachnoid cyst who appeared psychosis-like syndrome [12].

Overall, there was only one case of porencephaly and psychosis and three cases of schizencephaly (which is considered to be a variation of porencephaly) and psychosis.
## Table 1

<table>
<thead>
<tr>
<th>Number of cases</th>
<th>Anatomical Damages</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Porencephaly case</strong></td>
<td>1</td>
<td>Multiple focal leukoencephalomalacia changes in the left frontal lobe, bilateral occipital lobes and left basal ganglia, and a large porencephalic change in the right temporal lobe</td>
</tr>
<tr>
<td><strong>Schizencephaly cases</strong></td>
<td>2</td>
<td>unilateral <em>schizencephaly</em> cleft and bilateral <em>schizencephaly</em> cleft</td>
</tr>
<tr>
<td><strong>Relan P et al 2002</strong></td>
<td>1</td>
<td>unilateral <em>schizencephaly</em> cleft</td>
</tr>
<tr>
<td><strong>Current case</strong></td>
<td>1</td>
<td>a large <em>porencephalic</em> cyst on the left frontal and temporal lobes.</td>
</tr>
</tbody>
</table>

## CONCLUSION

It is unclear whether porencephaly and psychosis concur by chance in this case or that there is actually an increased risk for psychosis in patients with porencephaly. Our review showed that there are only two cases with porencephalic cysts and psychosis as well as 3 cases of schizencephaly. These damaged brain regions are associated with psychotic symptoms. The presence of neurodevelopment anomalies may have pathoplastic effect on the manifestation of psychosis. We assume that this patient’s brain lesions have a crucial
influence on her vulnerability to psychotic symptoms. She had no past and familiar psychiatric history and this was her first psychotic episode. Another vulnerability factor is that her IQ was 64, indicating mild mental retardation. This is associated with a higher incidence of psychotic disorder, than in normal population\(^{[13]}\). Her porencephalic cyst extended from left frontal to the left temporal lobe. Lesions in these areas have been associated with psychosis. This case highlights the need that in patients with similar clinical features as described above, radiological examination should be carried out so as to discover damaged brain lesions.

**References**


