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CASE REPORT
http://dx.doi.org/10.4306/pi.2014.11.1.102
Print ISSN 1738-3684 / On-line ISSN 1976-3026
OPEN ACCESS

Schizophrenia-Like Psychosis and Dandy-Walker Variant Comorbidity: Case Report

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Dandy-Walker variant is a developmental malformation consisting of cerebellar hypoplasia and cystic dilatation of the fourth ventricle. Previous research has proposed a possible role for the cerebellum in cognition and in schizophrenia. In this paper we report a schizophrenia-like psychotic disorder in a 30 year-old woman with Dandy-Walker variant. The patient was treated with risperidone 6 mg/day, biperiden 4 mg/day and risperidone depot 50 mg injections fortnightly, and most of the symptoms were ameliorated within 2 months. The similar cognitive profile to populations with cerebellar pathology and rarity of the condition strongly suggests that there may be direct relationship between cerebellar pathology and appearence of psychotic symptoms. **Psychiatry Investig 2014;11:102-104**

Key Words Schizophrenia, Dandy-Walker, Cerebellum, Comorbidity.

INTRODUCTION

Dandy Walker 'syndrome' was first defined by Dandy and Blackfan[1] in 1914, but the its precise characteristic properties described by Hart et al.[2] in 1972 . Dandy-Walker complex (DWC) is a series of neurodevelopmental anomalies in the posterior fossa, including Dandy-Walker malformation, Dandy-Walker variant (cerebellar hypoplasia and cystic dilatation of the fourth ventricle), mega-cisterna magna and posterior fossa arachnoid cyst.[3] Although these four subtypes of DWC are believed to occur between the 7th and 10th week of gestation and are regarded as a continuum in embryology,[4] they anatomically have their own unique characteristics.[3] There are some rare case reports on coincidence of schizophrenia and DWC in medical literature,[5-9] but the relationship between psychiatric symptoms and the DWC is still unclear because of the lack of data. Cerebellum plays an important role in cognition and a variety of psychiatric disorders[10,11] including schizophrenia.[12] Neuroimaging studies showing abnormalities in cerebellar structure[13] and function, especially the vermis, suggest the possible role of cerebellum in the pathophysiology of schizophrenia. These abnormalities can be detected both in chronic patients and at the time of onset of the disorder.[13,14]

Here we report a case of schizophrenia in a 30-year-old woman with Dandy Walker variant. The documentation of this case may contribute to the understanding of the pathophysiology of schizophrenia.

CASE

30-year-old female patient with a 2-year history of psychosis presented with schizophrenia-like symptoms including, auditory and visual hallucinations, sense of insecurity, delusions of reference, of persecution, of being controlled; inapropriate and labile affect; irritability, aggression, workplace absenteeism, sleep disruption, and having suicidal and homicidal thoughts was brought to the emergency department with her brother and sister. The patient was being followed in different outpatient clinics with irregular use of various psychotropic medications such as risperidone, olanzapine, sertraline etc. This was her first hospitalization. About two weeks before admission, she manifested strange behaviors (covering toilet windows with toothpaste, having a knife under the bed to sleep, occasionally trying to attack to relatives with a knife). Premorbid educational performance and social adjustment were reported as normal. Family history of psychosis in one of his brother was reported. No remarkable anomaly was found by physical

Received: January 23, 2013 **Revised:** April 30, 2013
Accepted: May 9, 2013 **Available online:** January 21, 2014

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examination and laboratory testing. The mental status examination revealed slowness of thought, stereotypic and vicious thought content, blunted affect, absence of spontaneous speech, psychomotor retardation, social withdrawal, poor rapport and lack of insight. Borderline intelligence activity was found on Weschler Adult Intelligence Scale-revised in Turkish (WAIS-RT) and full intelligence quotient was 75. Her overall cognitive functions were obviously impaired; (poor attention, impaired recent and remote memory, visuospatial distortion, simple arithmetic) but the abstract reasoning was roughly good. Electroencephalogram revealed moderate bioelectric disorganization in each frontal and right temporooccipital regions and the presence of sharp wave discharges in the both centroparietal regions was reported. MR scanning disclosed cerebellar vermis hypoplasia and large cerebellar medullary system in the 4th ventricular posterior cisterna magna (key-hole deformity) that is compatible with Dandy-Walker variant (Figure 1). Initial treatment with Risperidone 50 mg depot and 6 mg/day p.o, biperiden 4 mg/day p.o treatment and lorezepam 2.5 mg/day p.o achieved nearly 35% reduction in Positive and Negative Symptoms Scale (PANSS) scores (from 144 to 90) within 10 days. Treatment with injectable risperidone 50 mg fortnightly eliminated positive psychotic symptoms and most of the negative symptoms within two months except social isolation.

DISCUSSION

In this case report, Dandy Walker variant and schizophrenia may be found coincidentally together or any cerebellar dysfunction due to Dandy Walker variant may cause or contribute to the appearance of psychotic symptoms. Considering previous case reports, psychiatric symptoms have been related with DWC but, the spectrum of mental symptoms varies quite widely between cases.[15] The varieties of structural abnormality of the brain in these cases might partly contribute to the diversity of the psychiatric symptoms since the position and the severity of the anomalies in the previously mentioned cases were not exactly the same. Despite the wide range of symptoms some common clinical features had been observed in a series of our cases: juvenile or young adult age onset, high frequency of family history of psychosis, atypical psychiatric symptoms, high prevalence of cognitive deficit, borderline intellectual functioning and refractoriness to treatment.[15]

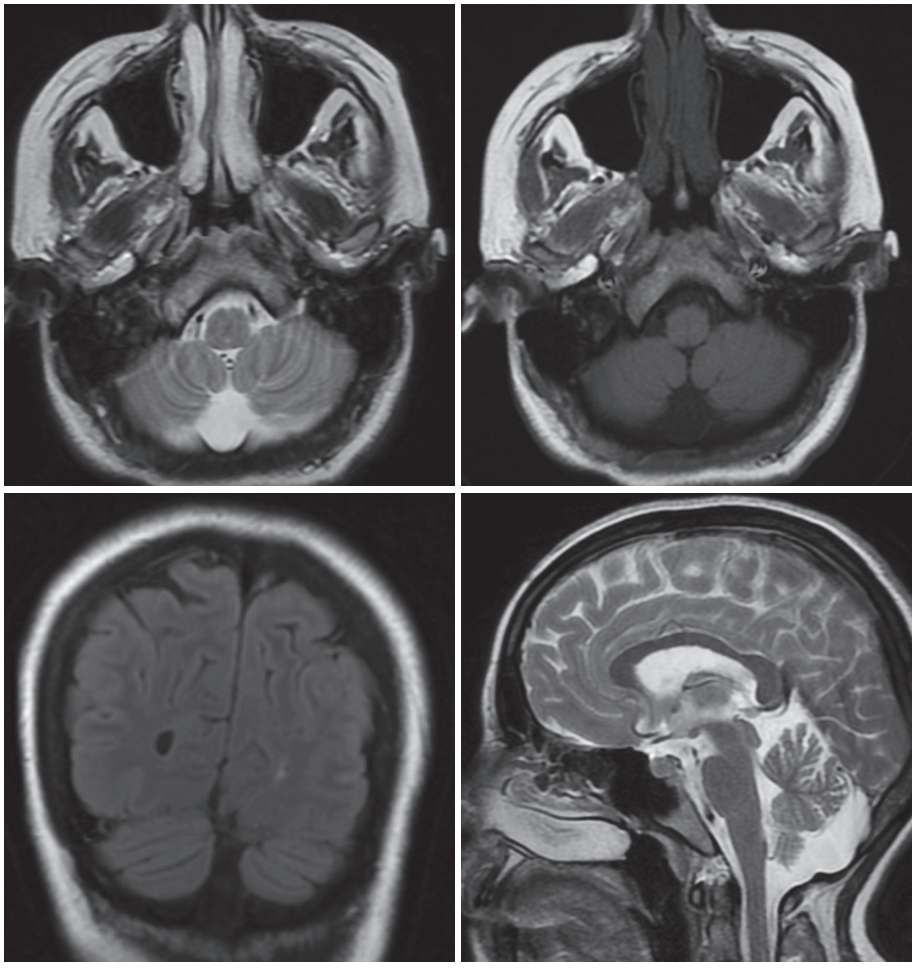

Figure 1. Magnetic resonance images of the case demonstrates Dandy walker variant.

Most of these features were also found in the cases previously reported.[5-9]

Andreasen suggested a model to explain "misconnections" between cortical regions and the cerebellum mediated through the thalamus in schizophrenia (the cortico-cerebellar-thalamic-cortical circuit).[16] Psychotic symptoms such as hallucinations may occur in case of an abnormality in this circuit.[17] It is also considered that cerebellar feedback pathways through the thalamus to the cerebral cortex may play a role in peduncular hallucinations.[18,19] Some postmortem and neuroimaging studies have demonstrated cerebellar gray matter volume reduction and a smaller anterior vermis in schizophrenic patients relative to normal or psychiatric control subjects.[20] There was a significant negative correlation between cerebellar gray matter volume reduction and halucinations.[18] Moreover, in a study it was found that there was a significant correlation between delusions and activation of cerebellum.[19]

In the present case, Dandy Walker variant might contribute to the activation of cerebellum. In the light of these findings, it might be suggested that cerebellar dysfunction may interfere in the emergence of psychotic symptoms such as hallucinations and delusions.

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