Case Report

Dandy–Walker Variant with Schizophrenia: Comorbidity or Cerebellar Cognitive Affective Syndrome?

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ABSTRACT

Dandy–Walker complex (DWC) is a series of neurodevelopmental anomalies involving the posterior cranial fossa. The cerebellum has long been considered to be involved in motor coordination and balance. However, it has now been noted to play an important role in higher order cognitive, emotional, and behavioral functions. The concept of cerebellar cognitive affective syndrome, describing a coherent spectrum of cognitive and behavioral disturbances in adults following cerebellar damage has long been proposed. There have been reported cases of co-occurring psychiatric symptoms and DWC in literature, but the conclusive evidence for an association between the same remains lacking. Herein, we report a case of schizophrenia presenting along with Dandy–Walker Variant.

Key words: Affective syndrome, cerebellar cognitive, Dandy–Walker variant, schizophrenia

INTRODUCTION

Dandy–Walker complex (DWC) is a series of neurodevelopmental anomalies involving the posterior cranial fossa. It comprises of DWC malformation (cystic dilatation of the fourth ventricle with enlarged posterior fossa, complete or partial agenesis of the cerebellar vermis, elevated tentorium cerebelli, and hydrocephalus), DWC variant (variable hypoplasia of the cerebellar vermis with or without enlargement of the cisterna magna, communication between the fourth ventricle and arachnoid space and no hydrocephalus), mega-cisterna magna (enlarged cisterna magna with intact cerebellum and fourth ventricle), and posterior fossa arachnoid cyst.[1] The cerebellum has long been considered to be involved in motor coordination and balance. However, it has now been noted to play an important role in higher order cognitive, emotional, and behavioral functions.[2] Schmahmann and Sherman gave the concept of “cerebellar cognitive, affective syndrome” (CCAS), describing a coherent spectrum of cognitive and behavioral disturbances in adults following cerebellar damage.[3] There have been reported cases of co-occurring psychiatric symptoms and DWC in literature, but the conclusive evidence for association...
between the same remains lacking. Herein, we report a case of schizophrenia in a patient with Dandy–Walker Variant.

CASE REPORT

A 25-year-old male presented with a 3 years history of psychotic symptoms. Detailed evaluation of the case revealed delayed motor milestones along with history of truancy, bullying, oppositional behavior, and poor academic performance throughout childhood and adolescence. The patient also indulged in occasional cannabis and alcohol use from 18 years of age. At the age of 22 years, he developed suspiciousness, hallucinations, and aggressive behavior along with significant sociooccupational decline. There was no significant past or family history. Detailed physical examination revealed dysdiadiokinesia and dysmetria in the left upper limb. Patient had delusion of grandiosity, second and third person auditory hallucinations on mental status examination. Higher mental functions revealed impaired concentration, deficits in immediate and recent memory, poor general fund of information, concrete thinking, impaired judgment, and absent insight. Patient’s routine blood investigations were within normal limits. His urine drug screen was also negative. A plain magnetic resonance imaging brain showed inferior vermis hypoplasia with communication of the fourth ventricle with cisterna magna without hydrocephalus, suggestive of Dandy–Walker variant [Figure 1]. Intelligence quotient was 81 (borderline intelligence) as tested by Wechsler Adult Performance Intelligence Scale. Neuropsychological assessment revealed moderate impairment in working memory and ideational fluency. In temporal lobe testing, his speech had poor productivity, paucity of content of speech, impairment in visual integration, moderate impairment in verbal learning and memory functions, and severe impairment in visual learning and memory functions. No impairment was noted in other lobe functions. The patient was diagnosed as a case of schizophrenia and started on oral antipsychotic olanzapine which was optimized to a dose of 15 mg/day. He showed partial improvement in the psychotic symptoms through patient’s speech and cognitive deficits remained static. One month postdischarge, patient continued to have stable cognitive deficits with partial resolution of psychotic symptoms.

DISCUSSION

The involvement of cerebellum in a variety of psychiatric diseases, such as schizophrenia, autism, depression, and obsessive–compulsive disorder has been substantiated by evidence from morphological, metabolic, and functional neuroimaging studies. Psychiatric presentation in DWC has been varied with the spectrum ranging from psychotic to cognitive symptoms. In a case series of DWC with co-morbid psychiatric disorders, common clinical features found were: Juvenile onset of symptoms, family history of psychosis, atypical psychiatric symptoms, and high prevalence of cognitive deficits and treatment refractoriness. Schmahmann and Sherman further elaborated the role of cerebellar lesion in neuropsychiatric symptoms with the introduction of CCAS. The CCAS is a coherent spectrum of cognitive and behavioral disturbances consisting of (i) executive dysfunctions such as disturbances in planning, set shifting, abstract reasoning and working memory, (ii) visuospatial deficits, (iii) mild language symptoms such as anomia and agrammatism and (iv) behavioral and affective disturbances. The symptoms observed in the CCAS are consistent with predictions derived from anatomical and neuroimaging studies, which show extensive neural circuits connecting prefrontal, superior temporal, posterior parietal, and limbic cortices with the cerebellum and the disruption in these circuits could lead to the CCAS.

Impairments in cognition such as attention, working memory, learning, verbal fluency, motor speed, and executive functions have been thought to be a core feature of schizophrenia. In addition, reduced cerebellar vermis volume has been consistently seen in drug naïve schizophrenia patients. The cognitive impairments in schizophrenia have been proposed to arise out of dysfunctional cortico-cerebellar-thalamic-cortical circuits conceptualized as “cognitive dysmetria” leading to difficulty in prioritizing, processing, coordinating,
and responding to information.[9] Measures of functional connectivity suggest that long-range interactions between frontal and posterior regions are abnormally reduced in patients with schizophrenia[10] which may contribute in the development of delusions and hallucinations.

The index case had the clinical features observed in psychiatric presentation in DWC except for a family history of psychosis, and the neuropsychological assessment showed deficits in the aforementioned domains along with behavioral and affective symptoms as observed in CCAS. Partial improvement in psychotic symptoms on olanzapine and stable cognitive deficits hints toward possible treatment refractoriness. Thus, this case study helps in illustrating the role of cerebellum in schizophrenia and also in management including prognosis of CCAS. While many advances are still needed and anticipated in the field of etiopathogenesis of schizophrenia, however, the index case could be a representation of the CCAS and not just a simplistic co-morbid case of a functional psychiatric disorder.

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**Conflicts of interest**
There are no conflicts of interest.

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