Dandy-Walker Syndrome with psychotic symptoms: a case report

Caso clinico di sindrome di Dandy-Walker con sintomi psicotici

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SUMMARY. Here we report the case of a patient with psychotic symptoms apparently resistant to antipsychotic treatments. Since the last admission in a psychiatric division the patient was diagnosed with Bipolar Disorder type I and then referred to our Outpatients Unit of Treatment Resistant Psychosis, where she was subsequently re-diagnosed with Dandy-Walker Syndrome. The Dandy Walker Complex is a congenital brain malformation involving the fourth ventricle and the cerebellum. We investigated the cognitive impairment of the patient and found deficits prominently in executive functions. This report may add further evidence on the importance of a correct diagnosis prior to defining a patient as treatment resistant and highlights cerebellar dysfunctions that may contribute to neuropsychiatric symptoms and cognitive impairment.

KEY WORDS: Dandy-Walker Syndrome, treatment resistant schizophrenia, cognitive impairment, cerebellum, psychosis.

INTRODUCTION

The contribution of the cerebellum to functions other than movement coordination has been widely recognized. Cerebellar structures are considered to be involved in cognitive, emotional and behavioral processes1. Multiple evidence has pointed out the involvement of the cerebellum in the pathophysiology of several psychiatric illnesses such as schizophrenia and mood disorders2. The description of the cerebellar cognitive-affective syndrome (CCAS) in subjects with either congenital or acquired cerebellar lesions has provided a model of behavioral dysfunctions and cognitive impairments related to abnormal functions of the cerebellum3.

The Dandy-Walker Complex (DWC) comprises multiple developmental abnormalities of the cerebellum, including: Dandy-Walker Variant, Dandy-Walker Malformation, Mega Cisterna Magna, and posterior fossa arachnoid cyst4. The DWC has been characterized as a triad of malformations: dilatation of the fourth ventricle, complete or partial agenesia of the cerebellar vermis, enlarged posterior fossa with displacement of the tentorium. The DWC has been variably described in association with atypical psychosis5,6. Here we describe a case of atypical psychotic symptoms with cognitive impairment in a patient referred to our Unit on Treatment Resistant Psychosis, then diagnosed with Dandy-Walker Syndrome. Notably, it has been reported that confounding factors in defining treatment resistant schizophrenia may be misdiagnosis, organic psychosis or somatic diseases7,8.

CASE REPORT

A 29-year-old female with a 9-year history of psychiatric symptoms was admitted to our Unit on Treatment Resistant Psychosis after being previously diagnosed with either Schizophrenia, Delusional Disorder, Schizoaffective Disorder or Bipolar Disorder type I, according to the Diagnostic and Statistic Manual of Psychiatric Disorders IV version, text revised (DSM-IV-TR). Before our observation, the patient reports that she was first hospitalized with the diagnosis of Schizophrenia at the age of 20.
DISCUSSION AND CONCLUSIONS

This report shows a case of neuropsychiatric symptoms potentially related to impairments in cerebellar functions.

The spectrum of psychiatric symptoms in DWC ranges from psychotic to cognitive ones, impacting symptom domains similar to those impaired in schizophrenia. In clinical practice, it has been estimated that 20 to 50% of schizophrenic patients have no adequate response to antipsychotics and are categorized as treatment resistant. However, misdiagnosis has been regarded as a prominent cause of non-response and should be thoroughly investigated before considering a patient as treatment resistant.

This case report may support previous evidence on the role of the cerebellum in the pathophysiology of psychiatric symptoms. It has been observed that schizophrenic patients have altered cortico-cerebellar connectivity. The cortico-thalamic-cerebellar-cortical circuit (CTCCC) has been proposed to have a role in the coordination and monitoring of the fluent execution of mental activity. Disruption of this circuitry has been postulated to underlie cognitive impairment and clinical symptoms of schizophrenia. Structural MR analyses have yielded consistent reports of cerebellar atrophy in schizophrenia. Furthermore, the conceptualization of the CCAS as a result of acquired or congenital cerebellar lesions envisions a precise role of the cerebellum in cognition and behavior. Prominent features of the CCAS include deficient planning of actions, impaired abstract reasoning and working memory, decreased spatial cognition, decreased verbal fluency, impulsiveness, disinhibition, psychotic symptoms, and aggressive behavior. Interestingly, a significant number of these features were found in the report described. Therefore, the clinical presentation of this case is consistent with the view that brain developmental abnormalities involving the cerebellum may contribute to psychotic and cognitive symptoms resembling schizophrenia-like disorders.

BACS and WAIS-R results revealed prominent deficits in executive functions but not in verbal memory, which is frequently impaired in schizophrenic patients. Neuroimaging studies have shown a relatively consistent pattern of task-related cerebellar abnormalities in schizophrenia, particularly in the vermis. Instead, neocerebellar regions may be activated during tasks as memory encoding and retrieval in healthy volunteers. Therefore, vermis abnormalities such as those observed in our patient are not expected to impact
memory functions. These findings may support the role of distinct regions of the cerebellum in the pathophysiology of different neuropsychiatric symptoms and may putatively explain the lack of impairment obtained from our patient at the Verbal Memory Task. Therefore, this cognitive pattern may contribute to discriminate schizophrenic patients from cases, such as that described herein, where psychotic symptoms might be related to morphological brain lesions.

REFERENCES