From Cerebellar Malformation to Suicidal Idea: Case Report

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Abstract

In 1998, Schmahmann described an affective and cognitive cerebellar syndrome. Studies have shown that cerebellum plays a major role in higher functions since involved in cognitive and emotional processing. Papers reported studies of the direct implication of cerebellum in the psychopathology of described psychiatric disorders such as schizophrenia. Furthermore, a series of cases reported that posterior fossa malformations appear to be related to psychiatric disorders suggestive of schizophrenia. In this paper, we report a case of Dandy Walker Variant revealed by melancholic depression with suicidal ideation in a patient followed for schizophrenia. We then review the evidences supporting a possible link between the two conditions.

Keywords: Schizophrenia • Dandy walker • Cerebellum

Introduction

Dandy-Walker Complex (DWC) is the most common spectrum of the posterior fossa malformation including Dandy Walker Malformation (DWM). This last pathological entity is characterized by an enlarged posterior fossa, elevated tentorium cerebelli and hydrocephalus, complete or partial agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle; Dandy Walker Variant (DVW) comprises hypoplasia of the cerebellar vermis, communication between the cystic dilatation of the fourth ventricle and the arachnoid space without hydrocephalus. The mega-cisterna magna is described by an enlarged cisterna magna with cerebellum and intact fourth ventricle [1].

Indeed, these malformations are mainly expressed through neurological symptoms, and a number of clinical observations found cognitive or psychiatric affective manifestations such as acute or chronic psychosis or cerebellar cognitive affective syndrome also called syndrome of schmahmann [2,3]. Inhere reported clinical observations supports that cerebellum plays an important role in the superior cognitive functions. In this sense, we report a case illustrating an association between posterior fossa malformations and psychiatric symptoms before instituting arguments in favor of a possible cerebellar participation in the pathophysiology of revealed psychiatric disorders.

Case Report

Our patient was a 20 years old male, who left school 3 years before presenting in our consultation for learning difficulties. The patient lived with his father in the past year after spending earlier 10 years with his mother since his parents were divorced.

The childhood was without particularity except a difficult schooling. By his 17 year-old, the patient presented a gradual social withdrawal state, strange behavior, hallucinatory attitude, and sleep disorder. The diagnosis of schizophrenia was established and the patient was treated by 2 mg/day of Risperidone. A restricted response was obtained and the patient has still retained a generally disturbed functioning as well as verbal acoustic hallucinations. The follow-up was marked by the irregularity of consultations and a compromised therapeutic compliance. A year ago at the time when the patient moved to live with his father, this last one was not convinced of the diagnosis, and decided to stop the follow-up of his child before requesting a consultation since got worst with total seclusion, hostility toward loved ones with the presence of suicidal ideation. At admission, we found an isolated patient, with a weird contact, a marked psychomotor retardation, attitudes toward hallucinations, and a bizarre unsystematized delusion. The patient showed also a depressed mood with feelings of worthlessness, damnation, and ruin "I think I'm finished, the world no longer exists. . . " thoughts of death and suicidal ideation, and a clear context of anorexia and insomnia. The cognitive assessment was a marked psychic repercussion with memory complaints, difficulty to concentrate and huge distractibility. Finally, the patient was disoriented in time.

In front of this clinical profile expressing a psychotic, depressive and cognitive dimension; a treatment based on Quetiapine at 400 mg/day and Fluoxetine at 20 mg/day was indicated. A blood assessment of a potential inflammatory, infectious, endocrine or deficiency condition and was normal. Psychometric tests were performed including Positive and Negative Syndrome Scale (PANS) and Calgary depression scale for patients with schizophrenia with cognitive tests were not performed due to deep cognitive impairment.

The evolution in the 21st day was marked by a strong improvement of depression; vanished suicidal ideas, hopelessness and depressed mood. Calgary's score increased from 21 to 12. The psychotic symptoms showed a partial improvement with vanishing delusion and regression of disorganized speech and thought. However, negative symptoms and hallucinations persisted. The score of the positive, negative, and general psychopathology scales fell from 23, 47 and 63 to 19, 42 and 51, respectively. Magnetic Resonance Imaging (MRI) was required to explain the partial response to treatment without any cognitive improvement. MRI revealed a cystic dilatation of the mega cisterna magna with hypoplasia of the lower cerebellar vermis evoking a Dandy Walker Variant (Figure 1).

Subsequently, the Quetiapine dose was increase to 600 mg/day. After 2 months, the evolution was marked by an improvement in functioning, a decreased acoustic hallucinations and a continuous stabilization of the negative signs, with persisting marked cognitive deficit.

Discussion

We have reported a clinical case of a severe depression with suicidal ideation occurred in a patient followed for persisting schizophrenia,
significant cognitive impairment and persistent symptoms after the reinstatement of treatment that directed to an organic cause consisted of a cerebellar malformation evoking a Dandy Walker Variant.

The literature review found twenty DWV cases suggesting a potential link between psychosis and DWM. Inhere reported case, psychotic symptoms were characterized by early onset, the presence of a family history of psychosis, cognitive impairment, and resistance to treatment [2,4,8]. Indeed, our case filled this description except the family history of psychosis. Besides, the case presented a melancholic depression and suicidal ideation that are clinical features not found in the literature.

This malformation remains rare without epidemiological studies. Santoro et al. estimated the overall DWV prevalence of 0.79 per 100,000 births. DWV is the most frequent form and concerned 2.08 per 100,000 births [7]. They are isolated in half of cases or associated with other cerebral or extra-cerebral malformations.

This syndrome is expressed by severe neurological symptoms, psychomotor retardation, hydrocephalus, intracranial hypertension, hypotonia and extra cranial congenital anomalies [8]. Less common manifestations might include focal neurological signs such as strabismus, nystagmus, cranial nerve palsies, truncal ataxia, or language disorders. DWV could also manifest by epileptic seizures especially in case of associated supratentorial malformations [9]. These manifestations remain less severe in the DWV, which might not express neurological symptoms [8].

The cognitive alterations are established in DWV patients and are reported in most cases as an important clinical dimension such as our patient. These symptoms vary according to the severity of malformations and mainly affect higher brain functions, namely social cognition and the language [10,11]. On the other hand, normal lobulation of vermis and the absence of other brain abnormalities seem to be associated with a better cognitive prognosis [12,13].

So far, a little data concerning the association between this malformation and psychiatric disorders is available. Stambollu et al. studied 187 cases and found an associated psychiatric disorder in 18.6% of patients. Among these cases, 51% were DWV [9]. The same study found that patients with psychiatric disorders presented [15,16], 1% with psychotic disorder. Recent study listed cases of an association between posterior fossa malformations and psychotic disorders found in 24 cases, including 5 cases with schizophrenia diagnosis, while schizophrenia-like diagnosis was found in 4 cases [2]. However, no clinical criterion distinguishes schizophrenia cases from so-called schizophrenia-like cases. Acute psychosis cases, especially first psychotic episode have been identified while the routine use of imaging diagnostic tool revealed malformations [8,14,15].

The association of DWM with psychiatric disorders poses problems of the role played by the malformation as a causal factor or risk factor, the possibility of a fortuitous co-occurrence or the presence of common factors to both conditions given the importance of neurodevelopmental factors in the pathophysiology of schizophrenia.

The major hypothesis of a link between abnormalities of the cerebellum and psychosis added to motor control and coordination role, there is growing possibility of a fortuitous co-occurrence or the presence of common factors.


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