

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 (DWV) 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 (PANSS) 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 felled 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,

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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. In here reported case, psychotic symptoms were characterized by early onset, the presence of a family history of psychosis, cognitive impairment, and resistance to treatment [2,4,6]. 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 DWM prevalence of 6.79 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 or 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. DMV 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 DWM 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. Stambolliu et al. studied 187 cases and found an associated psychiatric disorder in 16.6% 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 [6,14,15].

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 evidence against this view. In fact, traumatic, vascular, or tumor cerebellar damage are separately manifested from motor symptoms, namely cognitive and emotional symptoms [16]. This syndrome described by Schmahmann is called the Cerebellar cognitive affective syndrome [17,18]. It refers to a constellation of deficits including working memory impairment, problems with planning, reasoning, behavioral disorders, reduced affect display and appears to be all associated with lesions of the posterior lobes and the cerebellar vermis [17,19,20]. Imaging studies support this finding by showing in normal subjects a co-activation of the cerebellum within areas responsible of cognitive and emotional processes [21]. This cognitive and affective dimension also characterizes schizophrenia, hence imaging studies of schizophrenic patients that found structural and functional

abnormalities in the cerebellum [22]. Indeed, morphological MRI produced a studies series reporting cerebellar atrophy. A large study involving 1349 patients found a moderate but very significant reduction in cerebellar volume in schizophrenic patients that is more marked in adults as well as in young people. This initially suggests a neurodevelopmental rather than neurodegenerative process [23]. Even more, the reduced volume of the gray matter is greater in the posterior cerebellum (lobe VI, Crus I) [24]. On the other hand, a series of studies of functional imaging in schizophrenic patients found a defect in cerebello-thalamo-cortical pathway [25,26]. The same abnormalities were found in subjects with high risk of developing psychosis [27]. Finally, given the cerebellum role in social cognition including facial recognition, it appears to be involved in the impairment of social cognition in schizophrenic patients [28,29].

Moreover, the cognitive and affective syndrome has an affective dimension reported by studies and described as a change in personality, a reduced affect display, dis-inhibition and behavioral disorders [17]. The DWM literature does not report any major depressive disorder except in a 33-year-old woman marked by impulsivity [30]. We therefore put forward the hypothesis that depressive disorders could be underdiagnosed in this population or masked by other neuropsychiatric manifestations of cerebellar involvement. The presence of depression in 35.5% of patients with degenerative cerebellar disease is an additional supporting argument [31]. Conversely, a study found vermis volume reduction in patients with recurrent depression [32]. However, these data present a viewpoint since degenerative diseases are affecting other brain areas which are linked to depression expression.

Conclusion

Despite being rare, the posterior fossa malformations seem to constitute a vulnerability factor to psychotic disorders. Recently, it was shown that cerebellum is involved in higher brain functions and emotions. Besides, the imaging data confirmed the link between morphological and functional abnormalities of the cerebellum and psychotic disorder. However, further powerful studies are still required to confirm or refute this hypothesis.

References

1. WBosemani, Thangamadhan, Gunes Orman, Eugen Boltshauser and Aylin Tekes, et al. "Congenital Abnormalities of the Posterior Fossa." *Radiographics* 35(2015): 200-220.
2. Tréhout, Maxime, Norbert Zhang, Marie Blouet and Alin Borha, et al. "Dandy-Walker Malformation-Like Condition Revealed By Refractory Schizophrenia: A Case Report and Literature Review." *Neuropsychobiology* 77(2019): 59-66.
3. Graf, Heiko, Beate Franke and Birgit Ablter. "Cerebellar Cognitive Affective Syndrome in Dandy-Walker Variant Disorder." *J Neuropsychiatry Clin Neurosci* 25(2013): E45-E46.
4. Isidro-García, Guillermo J, José A Espina-Barrio and Marcia Guitián-Domínguez. "Dandy-Walker Variant and Refractory Psychosis." *Actas Esp Psiquiatr* 45(2017): 179-84.
5. Zincir, Selma Bozkurt, Yiğit Kivılcım, Filiz Izci and Umit Basar Semiz. "Schizophrenia-Like Psychosis and Dandy-Walker Variant Comorbidity: Case Report." *Psychiatry Investig* 11 (2014): 102.
6. Williams, Aislinn J, Zhenni Wang and Stephan F Taylor. "Atypical Psychotic Symptoms and Dandy-Walker Variant." *Neurocase* 22 (2016): 472-475.
7. Santoro, Michele, Alessio Coi, Ingeborg Barišić and Ester Garne, et al. "Epidemiology of Dandy-Walker malformation in Europe: A EUROCAT Population-Based Registry Study." *Neuroepidemiology* 53 (2019): 169-179.
8. Sasaki-Adams, Deanna, Samer K. Elbabaa, Valerie Jewells and Lori Carter, et al. "The Dandy-Walker Variant: A Case Series of 24 Pediatric Patients and Evaluation of Associated Anomalies, Incidence of Hydrocephalus and Developmental Outcomes." *J Neurosurg Pediatr* 2 (2008): 194-199.

9. Stambolliu, Emelina, Myrsini Ioakeim-Ioannidou, Kimonas Kontokostas and Maria Dakoutrou, et al. "The Most Common Comorbidities in Dandy-Walker Syndrome Patients: A Systematic Review of Case Reports." *J Child Neurol* 32 (2017): 886-902.
10. Bedeschi, Maria Francesca, Maria Clara Bonaglia, Rita Grasso and Alda Pellegri, et al. "Agenesis of the Corpus Callosum: Clinical and Genetic Study in 63 Young Patients." *Pediatr Neurol* 34 (2006): 186-193.
11. Paul, Lynn K, Warren S. Brown, Ralph Adolphs and J. Michael Tyszka, et al. "Agenesis of the Corpus Callosum: Genetic, Developmental and Functional Aspects of Connectivity." *Nat Rev Neuro Sci* 8 (2007): 287-299.
12. Boddaert, N, O. Klein, N. Ferguson and P. Sonigo, et al. "Intellectual Prognosis of the Dandy-Walker Malformation in Children: The Importance of Vermian Lobulation." *Neuroradiology* 45 (2003): 320-324.
13. Bolduc, Marie-Eve and Catherine Limperopoulos. "Neurodevelopmental Outcomes in Children with Cerebellar Malformations: A Systematic Review." *Dev Med Child Neurol* 51 (2009): 256-267.
14. Gan, Zhaoyu, Feici Diao, Zili Han and Kanglai Li, et al. "Psychosis and Dandy-Walker Complex: Report of Four Cases." *Gen Hosp Psychiatry* 34 (2012): 102 e7-102 e11.
15. Ryan, Molly, Ernesto Grenier, Anthony Castro and Charles B. Nemeroff. "New-Onset Psychosis Associated with Dandy-Walker Variant in an Adolescent Female Patient." *J Neuropsychiatry Clin Neurosci* 24 (2012): 241-246.
16. Baillieux, Hanne, Hyo Jung De Smet, André Dobbeleir and Philippe F Paquier, et al. "Cognitive and Affective Disturbances Following Focal Cerebellar Damage in Adults: A Neuropsychological and SPECT Study." *Cortex* 46 (2010): 869-879.
17. Schmahmann, Jeremy D and Janet C. Sherman. "The Cerebellar Cognitive Affective Syndrome." *Brain* 121 (1998): 561-579.
18. Bielawski, Michael and Helen Bondurant. "Psychosis Following a Stroke to the Cerebellum and Midbrain: A Case Report." *Cerebellum Ataxias* 2 (2015): 1-4.
19. Manto, Mario and Peter Mariën. "Schmahmann's Syndrome-Identification of the Third Cornerstone of Clinical Ataxiology." *Cerebellum Ataxias* 2 (2015): 1-5.
20. Tavano, Alessandro, Rita Grasso, Chiara Gagliardi and Fabio Triulzi, et al. "Disorders of Cognitive and Affective Development in Cerebellar Malformations." *Brain* 130 (2007): 2646-2660.
21. Riedel, Michael C, Kimberly L. Ray, Anthony S. Dick and Matthew T. Sutherland, et al. "Meta-Analytic Connectivity and Behavioral Parcellation of the Human Cerebellum." *Neuroimage* 117 (2015): 327-342.
22. Andreasen, Nancy C and Ronald Pierson. "The Role of the Cerebellum in Schizophrenia." *Biol Psychiatry* 64 (2008): 81-88.
23. Moberget, T, NT Doan, D. Alnæs and T. Kaufmann, et al. "Cerebellar Volume and Cerebellocerebral Structural Covariance in Schizophrenia: A Multisite Mega-Analysis of 983 Patients and 1349 Healthy Controls." *Mol Psychiatry* 23 (2018): 1512-1520.
24. Moberget, Torgeir, Dag Alnæs, Tobias Kaufmann and Nhat Trung Doan, et al. "Cerebellar Gray Matter Volume is Associated with Cognitive Function and Psychopathology in Adolescence." *Biol Psychiatry* 86 (2019): 65-75.
25. Ferri, Jamie, J. M. Ford, B. J. Roach and J. A. Turner, et al. "Resting-State Thalamic Dysconnectivity in Schizophrenia and Relationships with Symptoms." *Psychol Med* 48 (2018): 2492-2499.
26. Cao, Hengyi, Oliver Y. Chén, Yoonho Chung and Jennifer K. Forsyth, et al. "Cerebello-Thalamo-Cortical Hyperconnectivity as a State-Independent Functional Neural Signature for Psychosis Prediction and Characterization." *Nat Commun* 9 (2018): 3836.
27. Bernard, Jessica A, Derek J. Dean, Jerillyn S. Kent and Joseph M, et al. "Cerebellar Networks in Individuals at Ultra High Risk of Psychosis: Impact on Postural Sway and Symptom Severity." *Hum Brain Map* 35 (2014): 4064-4078.
28. Hoche, Franziska, Xavier Guell, Mark G. Vangel and Janet C. Sherman, et al. "The Cerebellar Cognitive Affective/Schmahmann Syndrome Scale." *Brain* 141 (2018): 248-270.
29. Mothersill, Omar, Charlotte Knee-Zaska and Gary Donohoe. "Emotion and Theory of Mind in Schizophrenia—Investigating the Role of the Cerebellum." *Cerebellum* 15 (2016): 357-368.
30. Kim, Ji Hyun, Tae Ho Kim, Young Chil Choi and Soon-Cheol Chung, et al. "Impulsive Behavior and Recurrent Major Depression Associated with Dandy-Walker Variant." *Psychiatry Investig* 10 (2013): 303.
31. Leroi, Iracema, Elizabeth O'Hearn, Laura Marsh and Constantine G, et al. "Psychopathology in Patients with Degenerative Cerebellar Diseases: A Comparison to Huntington's Disease." *Am J Psychiatry* 159 (2002): 1306-1314.
32. DelBello, Melissa P, Stephen M. Strakowski, Molly E. Zimmerman and John M. Hawkins, et al. "MRI Analysis of the Cerebellum in Bipolar Disorder: A Pilot Study." *Neuropsychopharmacology* 21 (1999): 63-68.

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