Focus on Medical Sciences Journal **Case Report**

May 2018, Volume 4, Issue 2

Dandy-Walker Syndrome Associated with Recurrent Mania Episode: A Case Report

Fateme Hosseini Biouki [1], Fateme Hosseini [1], Reza Bidaki [1, 2], Maryam Akrami [1, *], Mohammad Ebrahim Ghanei [3], Elham Hosseinizade [1], Ehsan Zarepur [4], Mohammad Talebpoor Ardakani [1]

[1] Research Center of Addiction and Behavioral Sciences, Shahid Sadoughi University of Medical Sciences, Yazd, Iran
2 Diabetes Research Center, Shahid Sadoughi University of Medical Sciences, Yazd, Iran
3 Radiology Department, Shahid Sadoughi University of Medical Sciences, Yazd, Iran
4 Student Research Committee, Shahid Sadoughi University of Medical Sciences, Yazd, Iran

** Corresponding author: Maryam Akrami, Research Center of Addiction and Behavioral Sciences, Shahid Sadoughi University of Medical Sciences, Yazd, Iran. E-mail: marzieh_akrami@ymail.com*

Submitted: 01.05.2018

Accepted: 02.29.2018

Keywords:
Dandy-Walker Syndrome
Ataxia
Bipolar Disorder

Abstract

Introduction: Dandy-Walker Syndrome (DWS) is a congenital brain malformation. The psychiatric aspect of DWS has not been addressed sufficiently. To our knowledge, there are few reports about psychiatric presentation of this syndrome, especially bipolar disorder.

Case Presentation: We report a 48-year-old male with bipolar disorder type I diagnosis with recurrent mania episode apparently sensitive to benzodiazepines. The brain imaging was done to investigate the causes of ataxia and impairment of consciousness after prescription of benzodiazepine. According to the neurological consultation, CT scan, and MRI findings, DWS was diagnosed. Following shunt insertion, the psychiatric treatments were more effective for control of psychiatric signs.

Conclusion: This case report implicated the importance of studying and accurate diagnosis in the preoperative administration of medication, especially in patients with recurrent mania episode and impairment of brain function following the sedating drugs administration.

INTRODUCTION

Different diseases can occur in different parts of our body. Human's brain have so many functions and any type of disease or injury to nerves can lead to so many problems. The role of the cerebellum in movement coordination has been widely recognized. It plays an important role in cognitive, emotional, and behavioral processes [1-5]. Multiple studies have reported the effect of the cerebellum in the pathophysiology of various psychiatric disorders like mood disorders and schizophrenia. The model of behavioral dysfunctions as well as cognitive impairments that is related to cerebellum abnormal functions can be provided with the illustration of the cerebellar cognitive-affective syndrome with congenital or acquired cerebellar lesions [2-5].

The Dandy-Walker Syndrome (DWS) involves the multiple developmental abnormalities of the cerebellum, including Dandy-Walker variant, Dandy-Walker malformation, mega cisterna magna, and posterior fossa arachnoid cyst [5]. DWS has been characterized as a triad of malformations: dilatation of the fourth ventricle, complete or partial agenesis of the cerebellar vermis, and large posterior fossa with a displacement of the tentorium. The DWS has been variably explaining in association with atypical psychosis [6, 7]. Remarkably, it has been reported that confounding factors in defining the treatment of complicated bipolar disorder organic psychosis or somatic diseases may be misdiagnosed [8].

Here we describe a patient with recurrent mania episode apparently sensitive to benzodiazepines. He was a patient with DWS that was misdiagnosed until 3rd decades of his age. The comorbidity of bipolar disorder with DWS is very rare. We don't know the relationship between these disorders. It is interesting when a very rare congenital disease in cerebellar functions is detected following the course of psychiatric disorder.

CASE PRESENTATION

A 48-year-old male, rural and low socio-economic with a history of recurrent mania episode from 9 years ago admitted to

the psychiatric ward with previously diagnosed with bipolar disorder type I, according to DSM-IV-TR. Re-hospitalization was due to episodes of agitation, aggressiveness, decreased the need for sleep, an increase in goal-directed activity and hyper sexuality. There was no family history of neurological or psychiatric illnesses. Familial history of medical illness and psychiatric disorder were negative. The psychosocial intervention wasn't done and psycho pharmacotherapy wasn't effective. The patient wasn't symptom-free anytime.
The patient was under treatment of antipsychotic and mood stabilizers, the first documented hospitalization occurred six years ago, due to delusional thoughts, suspiciousness, insomnia and aggressive behavior. The patient was diagnosed with bipolar disorder type I and substance dependency (opium) and treated with Olanzapine 5mg twice a day and Sodium Valproate 200mg twice a day, Clonazepam 2mg daily (when necessary) After two days with the consent of a person is discharged and he discontinued the drugs after few months. In the following six years, the patient had seven compulsory admissions to local psychiatric hospitals.

Personal History

The patient was born at term by normal vaginal delivery without complications of pregnancy. Premorbid physical growth has been reported as normal. Pacific childhood spent without trouble but lots of detailed history of childhood growth was not available. Due to financial difficulties and lack of facilities has not gone to school and was engaged in farming and past oralism.
He smoked cigarette one pack/day from 28 years ago and from nine years ago, he used half a gram of opium a day without other drug addiction. He had 4 brothers and 2 sisters and none of them had a history of mental illness. Routine lab tests were normal (Table 1).
He was discharged after each admission with a diagnosis of bipolar disorder type I and substance dependency and was treated with Risperidone, Sodium Valproate plus Chlorpromazine. Since the last admission to a psychiatric ward, six months ago, patient was diagnosed with bipolar disorder type I and prescribed the following therapy: Sodium Valproate 500 mg twice a day, Risperidone 4 mg/daily, Clonazepam 2mg PRN, Subsequently, after receiving the first dose of Clonazepam, mental status examination revealed poor rapport, mild cognitive deficits. The patient could not walk alone and then fall without reducing the level of consciousness.

Assessment

CT scan showed severely comminuted hydrocephaly with enhancement of fissure and sulci were seen. For more evaluation, MRI without contrast was performed and dilatation of all cerebral ventricle (communicating hydrocephaly) and white matter (atrophy as well as periventricular high signal change on T2 FLAIR in both cerebral hemisphere) were seen. The MRI scan showed no evidence of space occupying lesion in supra and infratentorial structures that DWS was offering these symptoms. There was no family history of neurological or psychiatric illnesses. Therefore, based on the imaging studies, history, and clinical judgment, he was diagnosed as DWS.
Repeated previous period of ataxia and gait disorder after taking benzodiazepine and spontaneous recovery was notable. Informed written consent was obtained to report this case. Following surgery, some problems include ataxia and mood instability were declined.

CT scan findings

Non-enhanced CT scan demonstrated partial agenesis of the vermis, resulting in communication between the fourth ventricle and the cisterna magna in the normal sized posterior fossa (Fig 1A). Also the lateral ventricles were dilated with

Table 1: Neurological Findings (Physical Examinations)	
1.	Cranial nerves were intact
2.	Motor function: 2-A: muscle force: 5/5 , 2-B: lower extremity reflexes increased and asymmetric but deep tendon reflex upper limbs seemed to be symmetric
3.	Abdominal reflex was absent
4.	Hoffman sign was negative
5.	Coordination test (finger to nose) indicated a slight clumsiness and dissymmetric
6.	Heel to shin obviously was abnormal
7.	Rapid attention movement seemed to be dysdiadochokinesia
8.	Tandem gait was obviously abnormal
9.	Romberg test with the open eye was abnormal.
	Psychological examinations: A: Mini-mental state (MMSI) test score was 7 B: interpretation of bender visual motor gestalt test score was 18 C: Wechsler memory scale was 53 that under normal class D: In testing of scoring Rey Andre card was reported severe visual memory test In general, all tests confirmed the brain damage

Pharmacological Interventions

abnormal orientation (Fig 1B). There was no evidence of intra or extra-axial hematoma or abnormal calcification.

Imaging findings

Study of the MRI revealed the partial agenesis of the vermis, resulting in communication between the fourth ventricle and the cisterna magnain normal sized posterior fossa (Fig 1C). There was an associated with moderate hydrocephalus (Fig 1D) and partial agenesis of the corpus callosum (Fig 1D). Also, MRI demonstrated the significant dilatation of occipital horns of lateral ventricle and thin white matter in the posterior cerebrum (colpocephaly) (Fig 1E).

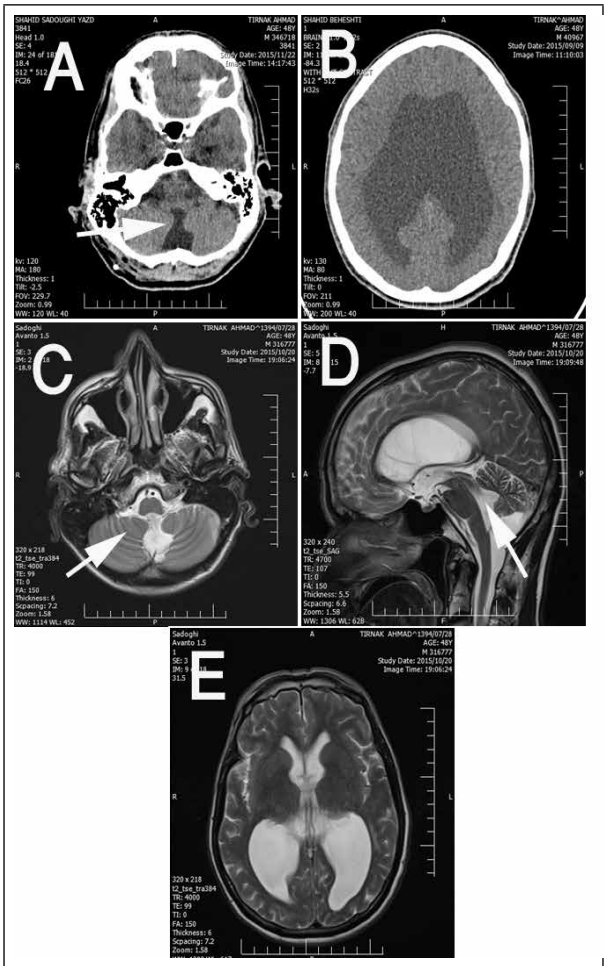

Figure 1: Non-enhanced CT scan demonstrates partial agenesis of the vermis, and communication between the fourth ventricle and the cisterna magna (blue thick arrow) in normal sized posterior fossa (A). Non-enhanced CT scan showing dilatation of lateral ventricles with abnormal orientation (B). Axial T2-weighed image depicting agenesis of the inferior vermis, (yellow arrow) resulting in communication between the fourth ventricle and the cisterna magna (C). Sagittal T2-weighed image showing partial agenesis of posterior body of corpus callosum, also linear signal void in aqueduct of sylvius (red arrow) due to stenosis and non-communicating hydrocephalus (D). Axial T2-weighed image depicting colpocephaly (E).

DISCUSSION

DWS is considered as a relatively very rare brain disorder most of the time, it is diagnosed in early life period [8], but sometimes it remains undiagnosed or misdiagnosed until the later childhood and only solitary cases of adulthood onset of symptoms which reported a broad spectrum of presenting symptoms. The more common type is about women transmitted as an autosomal dominant, it may be purely coincidental discovery [9, 10]. The psychiatric aspect of DWS has not been addressed sufficiently. To our knowledge, there are few studies of psychiatric presentation of this congenital malformation [11, 12].

To date, the reports about this syndrome in association with bipolar disorder type I are rare. This report presents a 49 year-old male in rural areas, illiterate and shepherd, unlike previously reported cases, sings of the disease until the age of 49 the conditions of employment and living patients who don't subtle movements not recognized.

This report shows the importance of neurological examination and procedural surgical intervention for the decline of psychiatric symptoms. Respect to treatment, the same treatment as the primary mood disorder was indicated effective. Due to patient sensitivity to psychotropic and sedative drugs-similar to other brain disorder- the lower dose of drug should be administrated.

The psychiatric symptoms related mood disorder was declined after psychopharmacologic intervention in some studies [13]. But about our patient, the treatment wasn't successful until after shunt insertion. Following surgery, the psychopharmacological interventions were more effective. Finally, we have concluded that justification whether DWS is one of the risk factors for bipolar disorder or other precipitate affective disorder is too soon.

Another hypothesis which calls for more research is the common genetics factor between DWS and bipolar disorder. The patients with severe and recurrent psychiatric disorders may have a general medical condition, it may be missed and even the patient experience serious side effects with low dose of drugs. A psychiatrist should know these situations and doubt to an organic brain disease in these situations.

If this syndrome is with other anomalies, therefore its prognosis will be the worse [14]. About our patient, if we presume that bipolar disorder is an abnormality of the brain, therefore the concordance of these may effect on prognosis. We emphasize on exact and complete physical examination of psychiatric patients.

CONCLUSION

We expect the comorbidity of bipolar disorder type I with DWS lead to poor prognosis, more next recurrence, and resistance to treatment. This case report implicated the importance of studying and accurate diagnosis in the preoperative administration of medication, especially in patients with recurrent mania episode.

ACKNOWLEDGMENTS

We thank the patient and his family who let us the opportunity to share the knowledge with others for the goal of scientific advances.

CONFLICT OF INTEREST

Non to declare.

FUNDING/ SUPPORT: NO

There is no funding for the present study.

AUTHORS' CONTRIBUTION

Fateme Hosseini Biouki, Fateme Hosseini, Mohammad Talebpoor and Maryam Akrami visited the patient, participated in data gathering, and wrote the primary draft and Reza Bidaki and Ehsan Zarepur visited the patient, wrote the primary draft, revised and submitted it

REFERENCES

1. Mormina E, Petracca M, Bommarito G, Piaggio N, Cocozza S, Inglese M. Cerebellum and neurodegenerative diseases: Beyond conventional magnetic resonance imaging. World J Radiol. 2017;9:371-88. http://dx.doi.org/10.4329/wjr.v9.i10.371 www.ncbi.nlm.nih.gov/pubmed/29104740
2. Phillips JR, Hewedi DH, Eissa AM, Moustafa AA. The cerebellum and psychiatric disorders. Frontiers in public health. 2015;3:66.
3. Sasannejad P, Rezaei F, Bidaki R, Zarepur E. Rare Presentation of Moyamoya Disease with Sub acute Presentation in Iran. Iran J Child Neurol. 2018;12:89-93. www.ncbi.nlm.nih.gov/pubmed/29379567
4. Imanian M, Ghasemzadeh MJ, Zarepur E, Zarepur A, Sarbandi Farahani R, Sarbandi Farahani R. The Relationship between Pneumonia with Parental Smoking in Children under 10 Years Old: A Case-Control Study. International Journal of Pediatrics. 2018;6:7791-6.
5. Parisi MA, Dobyns WB. Human malformations of the midbrain and hindbrain: review and proposed classification scheme. Mol Genet Metab. 2003;80:36-53. www.ncbi.nlm.nih.gov/pubmed/14567956
6. Bozkurt Zincir S, Kivilcim Y, Izci F, Semiz UB. Schizophrenia-like psychosis and dandy-walker variant comorbidity: case report. Psychiatry Investig. 2014;11:102-4. http://dx.doi.org/10.4306/pi.2014.11.1.102 www.ncbi.nlm.nih.gov/pubmed/24605131
7. Gan Z, Diao F, Han Z, Li K, Zheng L, Guan N, et al. Psychosis and Dandy-Walker complex: report of four cases. Gen Hosp Psychiatry. 2012;34:102 e7- e11. http://dx.doi.org/10.1016/j.genhosppsych.2011.08.012 www.ncbi.nlm.nih.gov/pubmed/21983277
8. Agrawal S, Thakur P. Dandy-Walker malformation. BMJ Case Rep. 2009;2009. http://dx.doi.org/10.1136/bcr.07.2008.0559 www.ncbi.nlm.nih.gov/pubmed/21686746
9. Tadakamadla J, Kumar S, Mamatha GP. Dandy-Walker malformation: An incidental finding. Indian J Hum Genet. 2010;16:33-5. http://dx.doi.org/10.4103/0971-6866.64936 www.ncbi.nlm.nih.gov/pubmed/20838490
10. Mendelson SG. Maternal grand mal seizure leads to a surprising diagnosis of Dandy-Walker variant. J Obstet Gynecol Neonatal Nurs. 2011;40:458-62. http://dx.doi.org/10.1111/j.1552-6909.2011.01267.x www.ncbi.nlm.nih.gov/pubmed/21771071
11. Bakhla AK, Sarkhel S, Kumar R. Bipolar I disorder in a patient with Dandy-Walker malformation. Psychiatry Clin Neurosci. 2010;64:212-3. http://dx.doi.org/10.1111/j.1440-1819.2009.02057.x www.ncbi.nlm.nih.gov/pubmed/20447018
12. Papazisis G, Mastrogianni A, Karastergiou A. Early-onset schizophrenia and obsessive-compulsive disorder in a young man with Dandy-Walker variant. Schizophr Res. 2007;93:403-5. http://dx.doi.org/10.1016/j.schres.2007.03.019 www.ncbi.nlm.nih.gov/pubmed/17478081
13. Eslami Shahre Babaki M, Estilaee F. Bipolar Disorder in a Young Girl with Dandy-Walker Syndrome. Iran J Psychiatry. 2015;10:212-3. www.ncbi.nlm.nih.gov/pubmed/26877755
14. Kolble N, Wisser J, Kurmanavicius J, Bolthauser E, Stallmach T, Huch A, et al. Dandy-walker malformation: prenatal diagnosis and outcome. Prenat Diagn. 2000;20:318-27. www.ncbi.nlm.nih.gov/pubmed/10740205