Case Report

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Mental Health Manifestations in a Case of Dandy-Walker Variant

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Abstract

Dandy-Walker variant disorder is a defined as cerebellar dysgenesis, enlarged posterior fossa with variation of cerebellar vermis hypoplasia and less commonly agenesis/thinning of the corpus callosum. Commonly reported symptoms in the literature are intellectual disabilities, epilepsy, and developmental delay. The pathogenesis of psychological manifestations in the illness is believed to be attributed to abnormal corticocerebellar tracts, resulting in what is recognized as cerebellar cognitive affective syndrome. Reports about the clinical manifestation and management challenges associated with Dandy-Walker variant coexisting with impulsive/destructive behavior are very few. We report a case of young adult man with Dandy-Walker variant who presented with challenging behaviors outbursts including verbal abusive, physically assaultive, environmental destruction and sexually disinhibited behavior and destructive behavior causing high risk of injury and financial consequences. His presentation suggests and organic psychosis diagnosis in the context of uncontrolled epilepsy. This report highlights the challenges in diagnostic formulation and management of this case; especially the collaboration of different services and the need of monitoring the pharmacotherapy, behavioral support plans and risk management to safeguard the patient and his family. The report also explores the possible factors contributing to the progression of his behavioral and mental health. In comparison with previously published cases.

Keywords: Intellectual disability; Challenging behavior; Epilepsy; Dandy-Walker variant

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Introduction

The terms Dandy-Walker complex or Dandy-Walker Syndrome (DWS) are used to denote the commonest cerebellar congenital anomalies of the posterior fossa; malformation (DWM) (agenesis of cerebellar vermis), variant (DWV) (hypoplastic cerebellar vermis), mega cisterna magna and arachnoid cyst [1-3]. DWS impacts 1/25,000 to 1/35,000 births, with higher incidence in females [4]. The variant form lacks hydrocephalus, with decreased neurological manifestations, and better prognosis [5]. DWS has been reported to be associated with schizophrenia, bipolar disorder, psychosis, obsessive/compulsive behaviors, hyperactivity, and impulsive/destructive behaviors [6-20]. Psychiatric comorbidities such as Attention Deficit Hyperactivity Disorder have been reported with associated Intellectual Disability and challenging behaviors [12].

We report a case of an adult male with Dandy-Walker presenting with organic psychosis and complex challenging behaviors including episodes of temper tantrums; anger outbursts, verbally abusive, physically assaultive, severe destructive and sexually disinhibited behaviors associated with social avoidance. We discuss aspects of risk management and implications of wide system approach.

Case Study

An 18 years old adult male of Arabian descent living with his family and recently left school due to lack of progress and behavioral problems. He was referred to the Mental Health Service, Learning Disability clinic with history of progressive deterioration in his behaviors over a period of one and half years. His behaviors described as episodic but of high intensity starts with him making demands and escalates to becoming aggressive towards his

family and attacking the environment breaking TV screens and any valuable items, he get in his hand. He has been observed by his family as becoming increasingly isolated in his room, refusing to come out during the day and only comes out at night after all goes to bed. He gets in the kitchen but often engage in breaking items and opening all food cans to throw them on the floor. He sometimes goes to his younger brother's room and demands their phone claiming it as his phone. Attempts at managing his behavior often require his father to try containing him by force as he can put himself and others at risk during these episodes. His father continued to sleep during the day and wakes up at night to ensure safety in the house from his behaviors. When out, he attacked the cars with stones and has broken number of family and neighbor's cars. His mood has been volatile, irritable, and easily provoked and often suspicious of all the family members leading to significant problem with relationship.

He has been seen before by child and adolescent psychiatrist, his behavioral problems were noted as social avoidance with nervousness, abnormal movements, and learning disability. He was diagnosed with anxiety and prescribed Escitalopram optimum dose with limited response and repeated episodes of challenging behaviors. His intellectual abilities were tested and his total IQ score reported as 57-60 using Wechsler Intelligence Scale for children-III (WISC-III).

He was also a known case of Epilepsy diagnosed by neurologist at age 12 years and was kept on antiepileptic medication Topamax 50 mg twice daily with no clear monitoring of his seizures. The diagnosis of DWV was made by the neurologist as an outcome of the investigation and report of a Brain Magnetic Resonance Imaging (MRI) done at the age of 12 years showing: Absent left cerebellar hemisphere and severely dysplastic right cerebellar hemisphere, and vermis with large retro-cerebellar Cerebro-Spinal Fluid (CSF) cyst, supratentorial Ventriculomegaly (VM) with thinning of carpus callosum along with dysmorphic right hippocampus with suggestion of right temporal lobe atrophy **(Figure 1).**

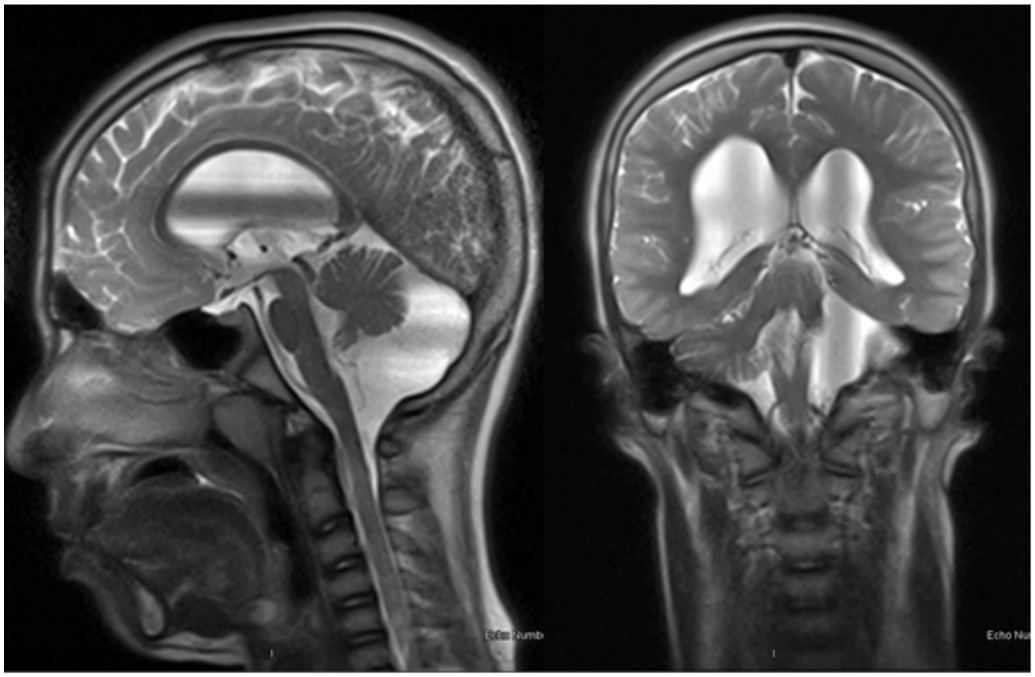

Figure 1 MRI images of the patient showing appearance of DWV.

The initial psychiatric assessment was done taking relevant history and collateral information from the patient, his parents and reviewing his electronic medical records. His parents described uncontrolled episodes of severe destructive behaviors, breaking Television screens, and his parents tend to buy two new televisions every month. He was reported to breaking cars, door handles and plates frequently. He forcefully takes his siblings' belongings and breaking them while keeping his own items safe and not allowing other to reach them. Moreover, he was developing frequent and sudden episodes of aggression/assault towards strangers, neighbors and family members for no apparent reason. The parents reported that his siblings were injured during these episodes of aggression. With increasing social isolation, he refused to come out of his room and neglecting self-care along with few shouting episodes in anger if asked to self-care. He also showed sexually disinhibited behaviors towards female members of the family causing distress and avoidance His sleep has been disturbed as he wakes all night and only sleep for shot period during the day. There is no history of alcohol or drugs use and no family history of psychiatric illness.

His developmental history shows that he was born at 24 weeks gestation with a birth weight of 1.4 kg and G6DP deficiency. His postpartum periods were complicated by Neonatal Intensive Care Unit (NICU) admission and need for intubation along with incidence of intraventricular hemorrhage. He had febrile

convulsions, chronic lung disease and Patent Ductus Arteriosus that was operate on at age 3 years. In addition, he had global developmental delay in cognitive and motor skills. He started speaking at 2 years of age and walked after the second year. He became dry by day at 4 years of age and needed significant support to achieve toilet training. Although attended mainstream school initially, however he has been supported all through with limited academic achievements. His education was interrupted by his behaviors and physical health issues needing to attend hospital appointments. He graduated from high school through special needs support educational program with poor academic performance.

In his mental state examination, when visited at the Emergency Department, he appeared slightly distressed, wearing night clothing, looking suspicious and was not cooperative to interview. He was nervous and shy asking to leave the room avoiding answering questions. He had fleeting eye contact and attempting to leave the room. His speech was clear, slow but with low tone and volume with delayed response. He appeared moody, irritable and his affect was changing, while being reactive only when talking about his interests. He holds paranoid ideas claiming that he is been watched and all people are against him wants to harm him but unable to expand but denied any hallucinations and suicidal or homicidal ideation. He appeared to have some insight into his behavior but not admitting to taking responsibility.

Neurological and general physical examination was unremarkable. Blood Investigations including; complete blood count testing, blood chemistry, thyroid function tests were within normal range and Electrocardiogram (ECG) was normal. He was referred for Neurologist review and a repeat electroencephalography which showed abnormal epileptiform discharges on the left posterior parietal region.

His behavior was managed using medical and nursing interventions and support from his family. His presentation was discussed with the neurologist and diagnostic formulation of organic psychosis in the contest of uncontrolled epilepsy was agreed. A plan to optimize his antiepileptic medication and use of small dose antipsychotic medication has been agreed. A behavioral approach to support the risk management plan and offer of short hospital admission was made. His family agreed with management plan except for the admission as he started to settle in the ED following the introduction of antipsychotic and expressed need to try treatment at home. Nursing and Occupational Therapy involvement to complete assessments of his need for support and monitoring of his medication at home agreed. His medication was adjusted with a to gradually withdraw Topiramate and introduce Sodium Valproate and use small dose Risperidone, psychoeducation given. He was visited at home for follow up by the Intellectual Disability team and he showed evidence of recovery whiting three weeks of discharge with significant reduction in his behavior and improved mental state.

Results and Discussion

The cerebellum is traditionally known to be primarily involved in regulation of coordination and motor functions. However, research indicates its potential role in sensory, procedural, linguistic actions along with its effect on irritability, affect, anger, aggressive behavior, and abnormal crying and/or laughter [21,22]. Previous clinical studies reported that stimulation of the surface of the vermis enhanced control of emotions and decreased aggressive bouts and feelings of anger [23,24]. Hence, cerebellar malformations; especially vermin hypoplasia are suggested to be significantly associated with anger, increased attacks of aggression, abnormal mood.

In this paper, we summaries radiological features, psychiatric manifestations and other mental health diagnoses in a group of 18 cases with DWS, with a special focus on the variant form of the disease **(Table 1)**. Interestingly, only 6/18 cases reported aggressive/hostile behaviors with vermin hypoplasia or cerebellar hemisphere hypoplasia [15] being a common radiological feature. However, none of them seemed to have similar severity in the abusive, assaultive and destructive behaviors in our case; suggesting that absence of one cerebellar hemisphere and/or temporal lobe atrophy and/or thinning of the corpus callosum might further substantiate the severity of the behavioral presentation.

Table 1: DWS: Case Reports with Radiological and Behavioural Features [5, 6, 8,9,10, 12, 14,16,17,13,18,20].

Authors	Radiological features	Psychiatric symptoms	Diagnoses	Authors	Radiological features	Psychiatric symptoms	Diagnoses
Marques et al. (2019)	Vermian hypoplasia	Irritability	DWV	Marques et al. (2019)	Vermian hypoplasia	Irritability	DWV
	Mega cisterna magna	Impulsive behavior	Schizophrenia		Mega cisterna magna	Impulsive behavior	Schizophrenia
		Persecutory delusions	mental retardation			Persecutory delusions	mental retardation
Dawra et al.	Vermian hypoplasia	Abusive behavior	DWV	Dawra et al.	Vermian hypoplasia	Abusive behavior	DWV
(2017)	Fourth ventricle communication with cisterna magna	Decreased self-care	Mental retardation	(2017)	Fourth ventricle communication with cisterna magna	Decreased self-care	Mental retardation
		Inappropriate laughter/crying				Inappropriate laughter/crying	

Sinha et al. (2017)	Vermian hypoplasia	Grandiose delusions	DWV	Sinha et al. (2017)	Vermian hypoplasia	Grandiose delusions	DWV
	Fourth ventricle communication with cisterna magna	Auditory hallucinations	Schizophrenia		Fourth ventricle communication with cisterna magna	Auditory hallucinations	Schizophrenia
		Moderate impairment in memory	Borderline intelligence			Moderate impairment in memory	Borderline intelligence
Rohanachandra et al. (2016)	Cerebellar hemispheres hypoplasia	Aggression	DWV	Rohanachandra et al. (2016)	Cerebellar hemispheres hypoplasia	Aggression	DWV
	Fourth VM	Sextual impulses	Schizophrenia w/ obsessive compulsive symptoms		Fourth VM	Sextual impulses	Schizophrenia w/ obsessive compulsive symptoms
	CSF filled cleft connected to right sigmoid sinus	Delusions of grandiosity/ control			CSF filled cleft connected to right sigmoid sinus	Delusions of grandiosity/ control	
		Somatic hallucinations				Somatic hallucinations	
BATMAZ et al. (2016)	Third & fourth VM	High speed of thoughts	DWM	BATMAZ et al. (2016)	Third & fourth VM	High speed of thoughts	DWM
	Posterior fossa cyst	Grandiose attitude	Bipolar disorder		Posterior fossa cyst	Grandiose attitude	Bipolar disorder
	Cerebellar hypoplasia		Borderline intelligence		Cerebellar hypoplasia		Borderline intelligence
Zincir et al. (2014)	Vermian hypoplasia	Persecutory delusions	DWV	Zincir et al. (2014)	Vermian hypoplasia	Persecutory delusions	DWV
	Fourth VM	Auditory hallucinations	Schizophrenia		Fourth VM	Auditory hallucinations	Schizophrenia
			Borderline intelligence				Borderline intelligence
Buonaguro et al. (2014)	Vermian hypoplasia	Persecutory delusions	DWV	Buonaguro et al. (2014)	Vermian hypoplasia	Persecutory delusions	DWV
	Enlarged cisterna magna	Cognitive deficits	Delusional Disorder		Enlarged cisterna magna	Cognitive deficits	Delusional Disorder
	Fourth VM		Bipolar Disorder		Fourth VM		Bipolar Disorder
		Abusive/ impulsive behavior	Schizoaffective Disorder			Abusive/ impulsive behavior	Schizoaffective Disorder
Kim et al. (2013)	Vermian hypoplasia		DWV	Kim et al. (2013)	Vermian hypoplasia		DWV
	Enlarged cisterna magna		Major depressive disorder		Enlarged cisterna magna		Major depressive disorder
	General VM				General VM		
Ryan et al. (2012)	Mild VM	Paranoid ideation	DWV	Ryan et al. (2012)	Mild VM	Paranoid ideation	DWV
	Pominence of basilar cisterns	Auditory hallucinations	Schizophrenia		Pominence of basilar cisterns	Auditory hallucinations	Schizophrenia
	Prominent CSF space dorsal and inferior to the cerebellum				Prominent CSF space dorsal and inferior to the cerebellum		

Gan et al. (2012)	Cerebellar vermis agenesis	Auditory hallucinations	DWM	Gan et al. (2012)	Cerebellar vermis agenesis	Auditory hallucinations	DWM
	Encephalatrophy	Persecutory delusions	Schizophrenia		Encephalatrophy	Persecutory delusions	Schizophrenia
		Disinhibition				Disinhibition	
	Hypoplasia of cerebellar hemispheres and vermis	Restlessness	DWV		Hypoplasia of cerebellar hemispheres and vermis	Restlessness	DWV
		Unreasonable phobia				Unreasonable phobia	
		Uncontrollable worries without reasons				Uncontrollable worries without reasons	
	Mega cisterna magna	Auditory hallucinations	Schizophrenia		Mega cisterna magna	Auditory hallucinations	Schizophrenia
	Ischemic foci in frontal, temporal, and parietal lobes	Persecutory delusions	Mega cisterna magna		Ischemic foci in frontal, temporal, and parietal lobes	Persecutory delusions	Mega cisterna magna
		Jealousy				Jealousy	
	Posterior fossa arachnoid cyst	Agitation and distraction	Posterior fossa arachnoid cyst		Posterior fossa arachnoid cyst	Agitation and distraction	Posterior fossa arachnoid cyst
		Loss of interest	Mild mental retardation			Loss of interest	Mild mental retardation
		Repetitive behavior				Repetitive behavior	
		Delusions				Delusions	
Aimua et al. (2012)	Vermian hypoplasia	Aggressive behavior	DWV	Aimua et al. (2012)	Vermian hypoplasia	Aggressive behavior	DWV
	Fourth VM		Bipolar disorder		Fourth VM		Bipolar disorder
	Communication between cisterna magna and fourth ventricle				Communication between cisterna magna and fourth ventricle		
Lingeswaran et al. (2009)	Vermian hypoplasia	Excessive cheerfulness	DWV	Lingeswaran et al. (2009)	Vermian hypoplasia	Excessive cheerfulness	DWV
	Posterior fossa cyst	Disinhibited in speech/behavior,	Bipolar affective disorder		Posterior fossa cyst	Disinhibited in speech/behavior,	Bipolar affective disorder
	Mega cisterna magna	Restlessness			Mega cisterna magna	Restlessness	
		Disobedience				Disobedience	
Prakash et al. (2009)	Vermian hypoplasia	Overactivity	DWV	Prakash et al. (2009)	Vermian hypoplasia	Overactivity	DWV
	Fourth VM	Abusive/ destructive behavior	ADHD		Fourth VM	Abusive/ destructive behavior	ADHD
		Lying and stealing	Hyperkinetic conduct disorder			Lying and stealing	Hyperkinetic conduct disorder
			Mild mental retardation				Mild mental retardation
Papazisis et al. (2007)	Vermian hypoplasia	Severe delusional ideas	DWV	Papazisis et al. (2007)	Vermian hypoplasia	Severe delusional ideas	DWV
	Fourth VM	Auditory hallucinations	OCD		Fourth VM	Auditory hallucinations	OCD
	Mild general VM		Schizophrenia		Mild general VM		Schizophrenia
			Mild mental retardation				Mild mental retardation
Turner et al. (2001)	Vermian hypoplasia	Hostile behavior	DWV	Turner et al. (2001)	Vermian hypoplasia	Hostile behaviour	DWV
	Cystic Fourth VM	Paranoid ideation	Schizophrenia		Cystic Fourth VM	Paranoid ideation	Schizophrenia

Schizophrenia was a common psychiatric comorbidity with DWS [7-11,15-20]. Marques et al. [17] reported that some patients can present with schizophrenia-like symptoms and might live up to 20 years with missed diagnosis of DWS that could have been reached *via* a simple brain imaging. Mental retardation was also fairly common with DWS, along with bipolar disorder [7,10,11-14,16-19,8]. In our case we did not find features suggestive of psychosis, however the severity of his challenging behaviors might lead to use of antipsychotic medication as part of the risk management plan.

Conclusion

We highlight the need for multidisciplinary assessments and interventions from specialist services recognizing the support needed for families caring for such high intensity and severity challenging behaviors in adult cases. Such cases may need high level of support to engage in activities outside home environment and careers need to be trained on behavioral management approach to manage his risk and improve his quality of life.

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Conflict of Interest

No conflicting relationship exists for any author.

Informed Consent

Written informed consent was obtained from the patient and his guardians for their anonymized information to be published in this article.

HMC Approval for Publication Obtained from the MRC

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