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Dandy–Walker malformation and intermittent explosive disorder: A case report

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Abstract

Dandy–Walker malformation is a group of brain malformations that sometimes present with psychotic features, violent and impulsive behavior, or mood symptoms. Here, we present a case report of a patient with Dandy–Walker malformation who presented with intermittent explosive disorder. A young man, aged 18 years, was brought to the author's hospital [Hamad Medical Corporation] with anger outbursts, irritable mood, and violent behavior. His magnetic resonance imaging scans showed typical alterations of Dandy–Walker malformation. He also had mild intellectual disabilities and epilepsy. After a few weeks of treatment with sodium valproate 1000 mg/day and risperidone 2 mg/day, his condition improved, and his violent behavior was significantly reduced in 3 months, 6 months, and 1 year of follow-up. There is broad consensus that Dandy–Walker malformation is associated with psychosis and other behavioral abnormalities because of a possible disruption in the prefrontal, thalamic, and cerebellar circuits. The link between Dandy–Walker malformation and intermittent explosive disorder may help us understand this type of brain malformation as a potential psychiatric comorbidity.

Keywords

Cerebellum, epilepsy, impulsivity, malformation, violent behavior

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Introduction

Dandy–Walker malformation (DWM) is the most frequent cerebellar malformation. It is defined by hypoplasia and upward rotation of the vermis cerebelli, cystic enlargement of the fourth ventricle, and an enlarged posterior fossa with cranial shifting of the lateral sinus, tentorium, and torcula herophili.[1] In 1914, Dandy and Blackfan initially documented this deformity, which Taggart and Walker published in 1942. In 1954, Bender coined the term "Dandy–Walker malformation" to describe the condition. In addition to these essential characteristics, DWM may present with other central nervous system (CNS) abnormalities and deformities, such as agenesis of the corpus callosum, visual impairments, and epilepsy.[1]

Dandy–Walker variant (DWV) is defined as cerebellar dysgenesis, enlarged posterior fossa with variation in cerebellar vermis hypoplasia, and agenesis or thinning of the corpus callosum, which is not common.[2] DWV is a less severe type of Dandy–Walker syndrome with fewer severe radiological abnormalities, such as a lack of hydrocephalus, less severe neurological symptoms, and better prognosis than DWM.[3]

Several case reports have linked DWM to psychotic symptoms, obsessive-compulsive symptoms, mood disorders, and hyperactivity.[4–9] We performed an extensive literature review using the PubMed, PsycInfo, and Google Scholar databases. To the best of our knowledge, only few case

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reports described DWM with impulsive and destructive behavior.[10,11]

According to the *Diagnostic and Statistical Manual of Mental Disorders* (5th ed.; *DSM-5*), intermittent explosive disorder is characterized by impulsive and aggressive outbursts, which might be verbal or physical. Furthermore, these outbursts are impulsive, unplanned, and difficult to predict. The outbreaks occur without trigger or are not proportionate to the initial trigger or stressor. To qualify for a diagnosis, outbursts must occur approximately twice a week for at least 3 months (American Psychiatric Association, 2013).

Road age, interpersonal violence, child abuse, and property damage are common behavioral aspects of intermittent explosive disorder. Violent and aggressive behavior instills distrust in family members, and friendships and relationships are likely to suffer as a result.[12]

This case report explores the difficulties in diagnosing and managing this condition, particularly the need for collaboration among various services, for monitoring the medication and behavioral support plans, and for providing risk management to protect the patient and his or her family.

Case presentation

An 18-year-old adult male dropped out of school due to lack of progress and behavioral difficulties. He was referred to the Mental Health Service after a history of progressive deterioration in his impulsive actions for over a year and a half. His behavior has been described as episodic and intense, beginning with demands and progressing to hostility toward his family. At other times, he became verbally abusive. His impulsive aggressiveness caused relationship problems because his violent outbursts featured insulting claims directed at his sisters; this happened multiple times, even when no apparent issues were present. At several instances, the patient grew so irritated that he threw objects around the house, including kitchen plates, books, food cans, and refrigerator contents, damaging television screens and expensive goods he could get his hands on. His family reported that he had become increasingly isolated in his room, refusing to leave during the day, and only leaving at night after everyone had gone to bed. He routinely smashed items and opened all food cans and dropped them on the floor when he entered the kitchen. Attempts to manage his behavior occasionally required his father to use force to restrain him, as he could injure himself and others during these instances. His father continued to sleep during the day and woke up at night to ensure safety from his actions. When he had these rage outbursts, he would leave his house and damage vehicles, including those of the family and neighbors, using stones. His mood has been volatile, impatient, and easily aroused, and he is frequently skeptical of all family members, resulting in a serious problem in the relationship. With increasing social isolation, he refused to leave his room and neglected self-care, yelling angrily if prompted. He also displayed sexually unrestrained behavior toward female family members, generating distress and avoidance. He stayed awake all night and slept during the day. He removed his brothers' belongings and broke them while keeping his own safe and preventing others from accessing them. These attacks, as described earlier, were episodic in nature and unprovoked; they occurred around two to three times on a weekly basis, and in between the episodes, he was generally manageable with no concerns from his family. However, his appetite, concentration, energy level, and interest were not generally affected between episodes; hence, he did not meet the criteria for major depression.

He was previously seen by a child and adolescent psychiatrist, who diagnosed his behavioral problems as social avoidance with anxiety and mild intellectual disability. According to his family, he had neither manic nor depressive symptoms nor any hallucinations or obsessive thoughts. According to his parents, their child's mental abilities were also evaluated (they provided us with his previous report evaluation), and he achieved a total intelligence quotient (IQ) score of 57 to 60 on the Wechsler Intelligence Scale for Children–III (WISC-III).[13]

He had epilepsy, which was diagnosed by a neurologist when he was 12 years old. He had been on antiepileptic medicine for several years and had no seizure activity. Based on the clinical investigation and brain magnetic resonance imaging (MRI) performed at the age of 12 years, the neurologist made the diagnosis of DWM. His MRI scan showed an absent left cerebellar hemisphere and a severely dysplastic right cerebellar hemisphere and vermis with large retro-cerebellar cerebrospinal fluid (CSF) cyst and supratentorial ventriculomegaly (VM) with thinning of the corpus callosum along with a dysmorphic right hippocampus with a suggestion of right temporal lobe atrophy (Figure 1).

There was no family history of psychiatric illness nor of alcohol or drug use. His early developmental history indicated that he was born at 24 weeks of gestation with a birth weight of 1.4 kg. The postpartum period was complicated by Neonatal Intensive Care Unit (NICU) admission, the need for intubation, and the occurrence of intraventricular hemorrhage. He had febrile convulsions and chronic pulmonary illness, and underwent surgery at the age of 3 years for patent ductus arteriosus. Despite the difficulties mentioned above, he graduated from high school through a special needs support educational program.

Mental status examination

The patient was a young gentleman. He was wearing a casual dress, looked suspicious, and was uncooperative during the interview. He was nervous and asked to leave the room and avoided answering questions. The patient had minimal eye contact. His speech was clear, slow, but with a low tone and volume, with delayed but relevant responses. He appeared

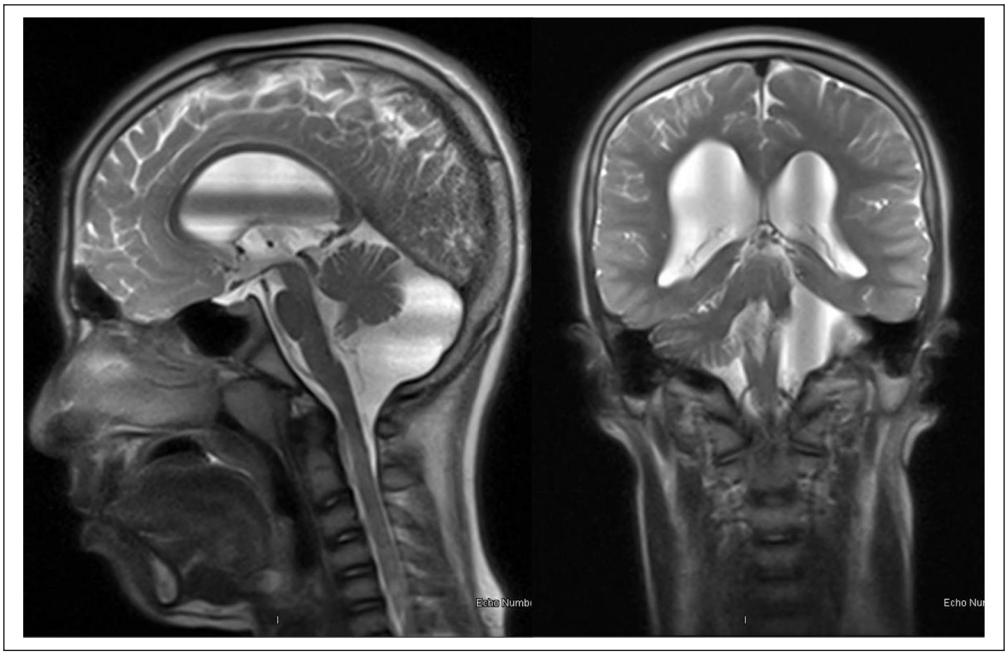

Figure 1. Magnetic resonance imaging scan of the patient showing the appearance of DWM.

irritable, and his affect was labile while being reactive only when talking about his interests. He became defensive when asked about his anger outbursts. He seemed to have insight into his behavior but did not admit to taking responsibility. He denied death wishes, suicidal ideation, homicidal ideation, and abnormal perceptions and obsessions. No overt delusions were observed during assessment.

Physical examination and investigations

His neurological and general physical examinations were unremarkable. Blood investigations, including complete blood count testing, blood chemistry, and thyroid function, showed results within the normal range, and his electrocardiogram (ECG) was normal. He was referred for a neurologist review and repeat electroencephalography, which showed abnormal epileptiform discharges in the left posterior parietal region.

Management

His presentation was evaluated by a neurologist, and an agreement was reached regarding the diagnostic formulation of intermittent explosive disorder. His behavior was managed through a multidisciplinary approach that included medical, nursing, and psychological therapy, and family support. A plan to switch his antiepileptic medicine to sodium valproate and low-dose antipsychotic medication was agreed upon. The involvement of nursing and occupational therapists to assess his need for support and monitoring of his medication at home was agreed upon. His medications were adjusted to gradually withdraw topiramate and introduce sodium valproate and a small dose of risperidone; psychoeducation was provided. His follow-up was carried out at home, and he showed recovery within a few weeks of discharge, with a significant reduction in his aggressive behavior and improved mental state; he was followed and assessed again at 6 months and after a year, and he continued to show stable mental state and behavior.

Discussion

The cerebellum has long been acknowledged for its involvement in motor coordination, but it is also becoming well known for its function in complex cognitive behavior. Historically, the cerebellum has received far less attention than the neocortex in both humans and model organisms. However, this tide is changing as advances in neuroimaging, neuropathology, and neurogenetics have resulted in the clinical classification and gene identification of various developmental disorders affecting cerebellar structure and function, associated with significant overall neurodevelopmental dysfunction.[14]

The tendency to exhibit impulsive and ill-planned behavior that is inappropriate for the situation is referred to as impulsivity.[15] It manifests itself behaviorally as impulsive acts (the difficulty of ending an ongoing response) but also as impulsive choices (failing to postpone gratification).[16]

In a previous study, main component analysis of the Barratt Impulsiveness Scale resulted in a three-factor model of impulsivity that includes increased motor activation, decreased attention, and reduced planning.[17] Given that these

three variables have been linked to impulse control disorders, different psychiatric ailments may be linked to impulsivity via distinct patterns of these underlying mechanisms.

Schizophrenia is a common psychiatric comorbidity associated with DWM. Gama Marques et al.[18] reported that some patients present with schizophrenia-like symptoms and might live up to 20 years with a missed diagnosis of DWM that could have been reached by simple brain imaging.

Mental retardation is also reasonably associated with DWM,[19] along with bipolar disorder.[5] In our patient, we did not find features suggestive of schizophrenia, depression, or bipolar disorder. However, the severity of his challenging behaviors might have led to him being placed on antipsychotic medication as part of the risk management plan and treatment for challenging behavior.[20]

We hypothesize that the cerebellar lesion played a significant role in the presence of impulsive behavior in our patient's case, as previous clinical studies of patients with posterior cerebellar lesions reported difficulties controlling their behavior and emotions, language deficits, and lack of concentration.[20–22] Furthermore, cerebellar cognitive affective syndrome is characterized by executive function deficiencies accompanied by disinhibited and inappropriate behavior, socially aberrant behavior, personality changes, and language impairments.[23]

Furthermore, animal lesion studies have demonstrated the importance of the cerebellum in perseverative behavior and behavioral disinhibition.[24] Lesions in the posterior vermis caused a delay in behavioral inhibition during the extinction trials.[25] Animals that received vermis lesions when they were young displayed delayed perseverative behavior as adults, lack of attention to novel stimuli, and behavioral disinhibition.[26] These findings suggest that the cerebellum is an important component of circuits that control the inhibitory processes for initiating actions.

We highlight the need for multidisciplinary assessments and interventions from specialist services that recognize the support needed to families caring for such high-intensity and severely challenging behaviors in adult cases. Such patients may require a high level of support to engage in activities outside the home environment, and caregivers need to be trained on behavioral management to manage the risks and improve the quality of life.

Conclusion

Considering previous case reports, psychiatric symptoms have been related to DWM, but the spectrum of mental symptoms varies widely between cases. The variety of structural abnormalities in the brain in these cases might partly contribute to the diversity of the psychiatric symptoms since the position and severity of the anomalies are never precisely the same in different patients. Comprehensive studies are required to better understand the relationship between impulsive behavior and the cerebellum. It is crucial to be aware of psychiatric symptoms, including impulsivity and destructive behavior, in cases of congenital cerebellar pathologies. It would be helpful to follow these cases with psychiatry and carefully review the treatment options considering resistance to treatment and potential side effects.

To conclude, when treating patients with DWM, a strong consultation-liaison approach between psychiatrists and neurologists is required. Relatives of DWM patients should be educated on the risk of psychiatric comorbidities in these patients. The current case emphasizes the importance of clinicians being aware of neuropsychiatric complications, such as intermittent explosive disorder, that may manifest in patients with DWM.

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Ethical approval

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Informed consent

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

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