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Congenital Malformation of the Posterior Fossa, Dandy-Walker Type: A Case Report

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Abstract

The Dandy-Walker syndrome (DWS) encompasses a group of anatomical midline cerebellar disorders with potential shared embryological origins, including the classic Dandy-Walker malformation, Blake's pouch cyst, and mega cisterna magna. Genetic factors, chromosomal abnormalities, and environmental influences contribute to its etiology. Clinical manifestations vary, with symptoms appearing from neonatal to adult stages. Diagnosis is performed through neuroimaging, evaluating the posterior fossa and associated anomalies. Management involves treating hydrocephalus, addressing associated anomalies, and providing neurological follow-up with a multidisciplinary team. Prognosis hinges on associated malformations and their severity, impacting long-term outcomes. We report a clinical case of a Dandy-Walker congenital malformation in a young patient in the only neurology department in Bangui, Central African Republic. Through this observation, and in a challenging medical practice context, we draw the attention of healthcare providers to the importance of medical imaging (cerebral computed tomography) in the face of neurological deficits, signs of intracranial hypertension, and/or hydrocephalus in young patients and children.

Keywords

Congenital Malformation, Dandy-Walker, Central African Republic

1. Introduction

Dandy-Walker syndrome is a rare congenital malformation of the posterior cerebral fossa of unknown etiology. The first autopsy description was reported in 1887 by Sutton [1] and it was not until 1914 when Dandy and Blackfan [2] studied

an association between hydrocephalus and cystic dilatation of the fourth ventricular. The malformation was further characterized by Dandy in [3] 1921 and by Taggart and Walker in [4] 1942 as being related to congenital atresia of the foramen of the fourth ventricle.but several studies indicate that there is a causal relationship with various types of chromosomal abnormalities and malformation syndrome. Its incidence is of the order of 1 case per 100,000 live births and it is more common in boys (1.24 per 100,000) than in girls (0.78 per 1000).

2. Medical Observation

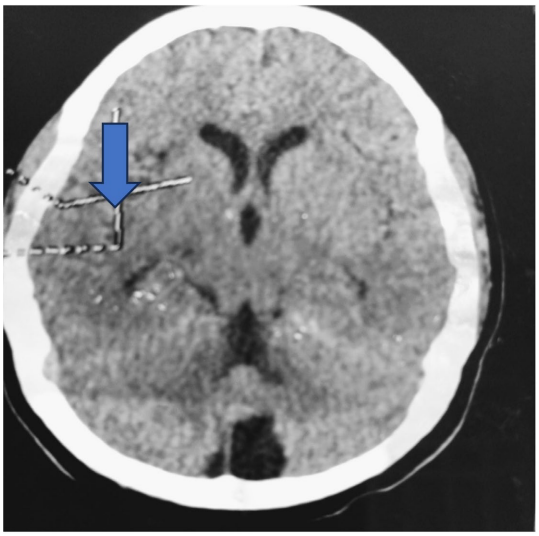

Figure 1. Source National Center for Medical Imaging 2022 (Central African Republic).

This was the patient G. D, aged 21, student, hospitalized on November 20, 2022 in the neurology department of the Sino Central African Friendship University Hospital for: headaches, motor deficit of the left hemibody, disorder of vigilance and vesico sphincter type of urinary incontinence and anal retention. The start of the symptoms dates back to the morning of November 2, 2022 with the rapidly progressive onset of headaches of progressively increasing intensity. In the evening, the headaches became boring and insomnia. Faced with these signs, self-medication was carried out with paracetamol tablets 1g every eight hours, leading to partial remission of the headaches after five days. On the morning of November 20, 2022, upon waking up, the patient presented with left hemibody heaviness, urinary incontinence, anal retention followed during the day by impaired vigilance, thus motivating an emergency consultation in neurology for better investigation and management charge. His pathological history was unremarkable. The neurological examination showed: obtundation with Glasgow score of 13/15, BP was 140/100 mmHg, rotatory nystagmus in extreme gaze on both sides, divergent strabismus of the left eye, left hemiparesis not proportional with FM = 0/5 at the MS and 2/5 at the MI, the bicipital and patellar ROT are abolished on the ipsilateral hemibody, the Babinski sign is indifferent on the left. The remainder of

the examination was unremarkable. The para-clinical examination showed: leukocytosis at 14100/mm^3, ESR at 130 mm at the 1st hour, SLV negative, BW negative, hepatic cytolysis with AST at 215 IU/l and ALT at 404 IU/l, Gamma GT at 91 IU/l, Klebsiella oxytoca cystitis;

• Brain CT was in favor of a supratentorial arteriovenous malformation with associated bleeding; at the infratentorial level: a vermian cerebellar malformation and a cystic one leading to tentorial elevation (Figures 1-3).

• The clinical course was marked by the occurrence of anemia requiring a transfusion of two bags of 450 ml of whole blood iso rhesus group on 11/20/2022 and a death occurring on 11/25/2022 in relation with intracerebral hemorrhage due to probable arteriovenous rupture, one day after the result of the brain CT was available, allowing the postmortem diagnosis to be established.

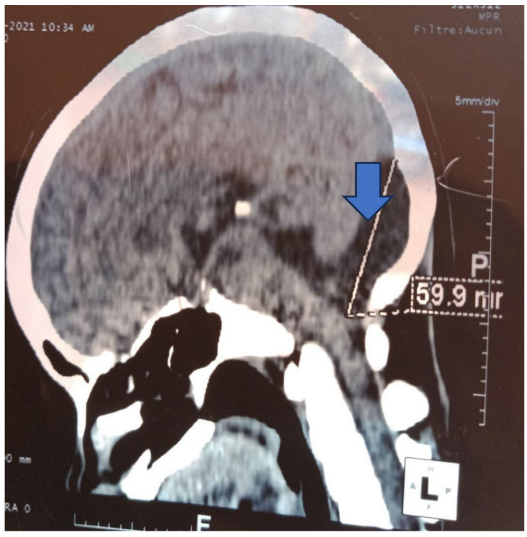

Figure 2. Source National Center for Medical Imaging 2022 (Central African Republic).

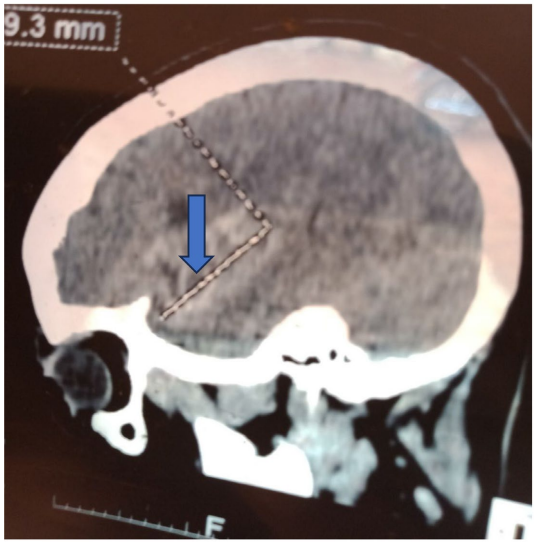

Figure 3. Source National Center for Medical Imaging 2022 (Central African Republic).

3. Discussion

Epidemiology

Dandy-Walker syndrome (DWS) is a heterogeneous group of anatomical disorders, potentially stemming from a shared embryological etiology, characterized by abnormalities in the development of midline cerebellar anatomy. These anomalies range from hypoplasia to agenesis, or even early cerebellar atrophy associated with a large cyst in the posterior fossa, typically in communication with the fourth ventricle [5]. Within this spectrum of pathologies are the classic Dandy-Walker malformation (DWM), Blake's pouch cyst (BPC), and mega cisterna magna (MCM) [6]. It is important to acknowledge the historical use of the term "Dandy-Walker variant", which has been discouraged in recent years due to its lack of specificity and its inability to adequately differentiate between distinct pathologies with similar imaging findings, such as inferior vermian hypoplasia (IVH) and other neurogenetic conditions that result in an enlarged retrocerebellar cerebrospinal fluid (CSF) space [7]. The first post-mortem description of this condition was made by Sutton in 1887 [1] [8]. Later, in 1914, Dandy and Blackfan reported the presence of hydrocephalus, an enlarged posterior fossa, agenesis of the cerebellar vermis, and cystic dilation of the fourth ventricle in a 13-month-old infant, and these findings were subsequently further characterized by Walker in 1921 [2]-[4]. The term "Dandy-Walker Malformation" was first introduced by Benda in 1954, who described six cases exhibiting the features initially reported by Dandy [8]. Benda hypothesized that abnormalities in the normal regression of the posterior medullary velum led to the formation of cystic dilations in the fourth ventricle [8]. The estimated annual incidence is 1 in every 25,000 to 35,000 live births, with a male-to-female ratio of 1:3. Hydrocephalus occurs in approximately 80% of DWS cases and accounts for 4 to 12% of pediatric hydrocephalus cases [9]. The aim of this article is to provide an updated literature review of the embryology, pathogenesis, clinical manifestations, diagnosis, and treatment of the DWS.

Clinical manifestations

The signs and symptoms present in DWS depend on the type of structural anomaly, the severity of the condition, associated malformations, and the age at diagnosis. In numerous cases, the diagnosis is already established through prenatal studies. However, in milder cases, the diagnosis may not be made until late adolescence [2]. Approximately 80% of DWM patients have hydrocephalus. Although hydrocephalus is an associated complication rather than a component of the malformation itself, it is often diagnosed before 3 months of age. Mega cisterna magna is often asymptomatic and typically detected incidentally [10]-[12]. The clinical characteristics resulting from increased intracranial pressure are influenced by both the patient's age and the severity of hydrocephalus [13].

Associated malformations

DWM is frequently associated with multiple intracranial and extracranial anomalies, which significantly impact the disorder's outcome and the severity of clinical symptoms. The severity of these disorders can vary widely, from severe

clinical presentations to almost asymptomatic cases. These malformations may be categorized as neurologic, systemic, or genetic and often coexist. Among the most common central nervous system (CNS) anomalies linked to DWM are ventriculomegaly, holoprosencephaly, encephalocele, and agenesis of the corpus callosum [9]. Typically, the initial symptoms of DWM are related to the CNS, such as hydrocephalus. However, the diagnosis of DWM may also occur due to the presence of systemic symptoms, including cardiovascular anomalies like transposition of the great arteries and congenital pulmonary stenosis; urogenital conditions such as hydrocele and horseshoe kidney; intestinal abnormalities like duodenal atresia, megarectum, and megasigmoid; and craniofacial anomalies including cleft palate, strabismus, and facial angiomas. Additionally, limb malformations and syndactyly of the fingers or toes have also been reported in association with DWM [14]. Recent studies suggest a potential association between posterior fossa abnormalities, such as DWS and psychiatric symptoms. However, the existence of a causal relationship remains unclear [15]. Peripheral malformations associated with Dandy-Walker syndrome are often described, the frequency of which varies greatly and can reach 76% [16]. In adults, progressive and slow decompensation can manifest itself as: headaches, especially chronic headaches, balance difficulties, walking disorders, unilateral sensory disorders, syncope, a progressive decrease in cognitive and intellectual abilities suggesting a picture of dementia, multiple psychiatric disorders [17].

In our case, the patient had symptomatic Dandy-Walker syndrome.

- The headaches were certainly related to intracranial hypertension;
- The vigilance disorder could be linked to acute decompensation due to probable damage to the brainstem;
- Nystagmus could reflect either a visual disorder, therefore in relation to divergent strabismus of the left eye or in relation to cerebellar damage, therefore in direct relation to Dandy Walker syndrome;
- Non-proportional left hemiparesis, which could be explained by damage to the subcortical motor pathways.

In our case, such peripheral damage was also present in the form of divergent strabismus of the left eye, which affects 10.5% of Dandy-Walker syndrome carriers [16].

The reasons for this asymptomatic period, as well as the reasons for progressive decompensation of the syndrome, remain uncertain [8]. For some authors, the reasons seem to lie in the existing imbalance between the production of cerebrospinal fluid and its absorption, which occurs late in adults [17]. Acute decompensation is manifested by: nausea, vomiting, headache, nystagmus, papilledema, etc. It is often linked to a sudden increase in intracranial pressure [18]. At the central nervous system level, DW syndrome is associated with other malformations in 48% [19] of cases respectively. In our case, we note as central nervous system malformations, a supratentorial arteriovenous malformation with bleeding and intracranial hypertension.

Treatment

The therapeutic objectives for DWM are not aimed at correcting the primary CNS malformation, but rather at managing hydrocephalus and the posterior fossa cyst. Treatment options vary according to the severity of symptoms and the presence of associated anomalies [20]. The main strategies include surgical procedures such as cyst membrane fenestration, cystoperitoneal shunting, ventriculoperitoneal shunting, cyst-ventriculoperitoneal shunting, and endoscopic third ventriculostomy with or without choroid plexus cauterization [21] [22].

Prognosis

In recent years, research has underscored the crucial impact of accurate fetal diagnosis, the detection of associated abnormalities, on the long-term prognosis of patients with DWS. According to the existing literature, infants born with congenital defects affecting two or more organ systems tend to exhibit the poorest survival rates [21]. Nevertheless, despite numerous contemporary studies, there remains a lack of consensus regarding the long-term prognosis of individuals with DWS [9]. Prognosis is intricately linked to the presence or absence of associated malformations, whether they are genetic, systemic, or neurological in nature, and the severity of each of these coexisting conditions. Therefore, the presence of the number of affected organs is associated with significantly higher neurological morbidity and neonatal mortality [22]. The literature review exposed that there has delineated two distinct prognostic categories within the spectrum of DWM. The first category pertains to cases where the vermis exhibits partial agenesis, while the rest of the brain's architecture remains intact, often resulting in a life that closely resembles normalcy. Conversely, in cases characterized by significant malformations, encompassing severe dysplasia of the vermis and substantial midline brain anomalies, the prognosis tends to be associated with adverse intellectual and neurological outcomes.

4. Conclusion

In conclusion, DWS encompasses a range of cerebellar abnormalities, likely stemming from shared embryological causes, characterized by a large cyst in the posterior fossa along with hydrocephalus and often associated with other CNS anomalies. Treatment primarily addresses hydrocephalus, often requiring surgical interventions like ventriculoperitoneal shunting or endoscopic third ventriculostomy, with each case requiring a tailored approach based on severity and associated anomalies. Prognosis for DWS varies, heavily influenced by the presence of additional anomalies, with children having concurrent CNS or systemic malformations generally facing worse outcomes. Early diagnosis and intervention are crucial for effective management and improved prognosis. This observation should draw the attention of health providers to the importance of brain CT in the face of neurological deficit, signs of intracranial hypertension and/or hydrocephalus in young subjects and children.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Sutton, J.B. (1886) The Lateral Recesses of the Fourth Ventricle; Their Relation to Certain Cysts and Tumours of the Cerebellum, and to Occipital Meningocele. *Brain*, **9**, 352-361. https://doi.org/10.1093/brain/9.3.352

[2] Dandy, W.E. and Blackfan, K.D. (1914) Internal Hydrocephalus. An Experimental, Clinical and Pathological Study. *The American Journal of Diseases of Children*, **8**, 406-482.

[3] Dandy, W.E. (1921) The Diagnosis and Treatment of Hydrocephalus Due to Occlusions of the Foramina of Magendie and Luschka. *Surgery, Gynecology and Obstetrics Archives*, **32**, 113-124.

[4] Taggart, J.K. (1942) Congenital Atresia of the Foramens of Luschka and Magendie. *Archives of Neurology and Psychiatry*, **48**, 583-612. https://doi.org/10.1001/archneurpsyc.1942.02290100083008

[5] Haddadi, K., Zare, A. and Asadian, L. (2018) Dandy-Walker Syndrome: A Review of New Diagnosis and Management in Children. *Journal of Pediatrics Review*, **6**, e63486. https://doi.org/10.5812/jpr.63486

[6] Robinson, A.J. and Ederies, M.A. (2016) Diagnostic Imaging of Posterior Fossa Anomalies in the Fetus. *Seminars in Fetal and Neonatal Medicine*, **21**, 312-320. https://doi.org/10.1016/j.siny.2016.04.007

[7] Wee, T., Gupta, N., Miller, E. and Pauranik, A. (2024) Not Dandy Walker Variant: A Review of Prominent Retrocerebellar CSF Space in Children. *Clinical Radiology*, **79**, 330-337. https://doi.org/10.1016/j.crad.2024.01.025

[8] Benda, C.E. (1954) The Dandy-Walker Syndrome or the So-Called Atresia of the Foramen Magendie. *Journal of Neuropathology & Experimental Neurology*, **13**, 14-29. https://doi.org/10.1093/jnen/13.1.14

[9] Monteagudo, A. (2020) Dandy-Walker Malformation. *American Journal of Obstetrics and Gynecology*, **223**, B38-B41. https://doi.org/10.1016/j.ajog.2020.08.184

[10] Cotes, C., Bonfante, E., Lazor, J., Jadhav, S., Caldas, M., Swischuk, L., *et al.* (2015) Congenital Basis of Posterior Fossa Anomalies. *The Neuroradiology Journal*, **28**, 238-253. https://doi.org/10.1177/1971400915576665

[11] Moosavi, A. and Kanekar, S. (2022) Congenital Malformations of Cerebellum. *Clinics in Perinatology*, **49**, 603-621. https://doi.org/10.1016/j.clp.2022.04.003

[12] Di Nora, A., Costanza, G., Pizzo, F., Di Mari, A., Sapuppo, A., Basile, A., *et al.* (2022) Dandy-Walker Malformation and Variants: Clinical Features and Associated Anomalies in 28 Affected Children—A Single Retrospective Study and a Review of the Literature. *Acta Neurologica Belgica*, **123**, 903-909. https://doi.org/10.1007/s13760-022-02059-z

[13] Reddy, P., Vannemreddy, P.S., Grier, L., *et al.* (2000) Dandy-Walker Syndrome Presentation of the Congenital Malformation in an Older Patient. *The Journal of the Louisiana State Medical Society*, **152**, 31-34.

[14] Tréhout, M., Zhang, N., Blouet, M., Borha, A. and Dollfus, S. (2018) Dandy-Walker Malformation-Like Condition Revealed by Refractory Schizophrenia: A Case Report and Literature Review. *Neuropsychobiology*, **77**, 59-66.

[15] Pascual-Castroviejo, I., Velez, A., Pascual-Pascual, S.I., Roche, M.C. and Villarejo, F.

(1991) Dandy-Walker Malformation: Analysis of 38 Cases. *Child's Nervous System*, **7**, 88-97. https://doi.org/10.1007/bf00247863

[16] Russ, P.D., Pretorius, D.H. and Johnson, M.J. (1989) Dandy-Walker Syndrome: A Review of Fifteen Cases Evaluated by Prenatal Sonography. *American Journal of Obstetrics and Gynecology*, **161**, 401-406. https://doi.org/10.1016/0002-9378(89)90531-0

[17] Hirsch, J., Pierre-Kahn, A., Renier, D., Sainte-Rose, C. and Hoppe-Hirsch, E. (1984) The Dandy-Walker Malformation: A Review of 40 Cases. *Journal of Neurosurgery*, **61**, 515-522. https://doi.org/10.3171/jns.1984.61.3.0515

[18] Hart, M.N., Malamud, N. and Ellis, W.G. (1972) The Dandy-Walker Syndrome: A Clinicopathological Study Based on 28 Cases. *Neurology*, **22**, 771-780. https://doi.org/10.1212/wnl.22.8.771

[19] Mallucci, C. and Parks, C. (2016) The Dandy-Walker Malformation. In: Cohen, A.R., Ed., *Pediatric Neurosurgery*: *Tricks of the Trade*, Thieme, 330-334.

[20] Wüest, A., Surbek, D., Wiest, R., Weisstanner, C., Bonel, H., Steinlin, M., *et al.* (2017) Enlarged Posterior Fossa on Prenatal Imaging: Differential Diagnosis, Associated Anomalies and Postnatal Outcome. *Acta Obstetricia et Gynecologica Scandinavica*, **96**, 837-843. https://doi.org/10.1111/aogs.13131

[21] Patel, S.K. and Mangano, F.T. (2023) Dandy-Walker Syndrome. In: Winn, H.R., Ed., *Youmans and Winn Neurological Surgery*, Elsevier, 1636-1641.

[22] Klein, O., Pierre-Kahn, A., Boddaert, N., Parisot, D. and Brunelle, F. (2003) Dandy-walker Malformation: Prenatal Diagnosis and Prognosis. *Child's Nervous System*, **19**, 484-489. https://doi.org/10.1007/s00381-003-0782-5